

Headache Classification Subcommittee of the International Headache Society (IHS)

**The International Classification of  
Headache Disorders  
2<sup>nd</sup> Edition**

**1<sup>st</sup> revision (May, 2005)**

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The headache classification subcommittee and its working groups have received valuable suggestions and critiques from a great number of colleagues around the world. They cannot all be acknowledged individually but we wish to mention two specifically. Robert Spitzer's immense experience in psychiatric disease classification inspired the reconstituted headache classification subcommittee at its first meeting. Michael B First is a psychiatrist and an expert in disease classification. He has been invaluable as an outside expert, early on helping the many new members who were unfamiliar with disease classification and, later, often guiding our steps in this difficult art. Despite the fact that headache is remote to his speciality he graciously accepted to chair a newly-formed working group on headache attributed to psychiatric disease.

Peter Goadsby, Richard B Lipton, Jes Olesen and Stephen D Silberstein have organised the practical aspects of our meetings.

Kirsten Hjelm has done most of the administrative and secretarial work for the subcommittee. Rosemary Chilcott has managed the finances. We thank both for their never-failing support.

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## **Preface to the First Edition (1988)**

The present document represents a major effort. The work has been going on for almost three years, and has involved not only the committee members, but also the many members of the 12 subcommittees. The work in the committee and subcommittees has been open, so that all interim documents have been available to anybody expressing an interest. We have had a two-day meeting on headache classification in March 1987 open to everybody interested. At the end of the Third International Headache Congress in Florence September 1987 we had a public meeting where the classification was presented and discussed. A final public meeting was held in San Diego, USA February 20 and 21, 1988 as a combined working session for the committee and the audience.

Despite all effort, mistakes have inevitably been made. They will appear when the classification is being used and will have to be corrected in future editions. It should also be pointed out that many parts of the document are based on the experience of the experts of the committees in the absence of sufficient published evidence. It is expected however that the existence of the operational diagnostic criteria published in this book will generate increased nosographic and epidemiologic research activity in the years to come.

We ask all scientists who study headache to take an active part in the testing and further development of the classification. Please send opinions, arguments and reprints to the chairman of the classification committee. It is planned to publish the second edition of the classification in 1993. Hopefully the revision will be based on new evidence.

The International Headache Society considers classification and diagnostic criteria for headache disorders to be a very important issue. Although the document needs further testing and modification, it is recommended to put it into immediate use in scientific studies. This pertains not only to drug trials, but also to biochemical and physiological studies.

**James W Lance**  
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## Preface to the Second Edition

Disease classifications and their diagnostic criteria are often received with scepticism by the medical community and may not be put to extensive use. It has therefore been a pleasant surprise to see how well the first edition of *The International Classification of Headache Disorders* was received. It was accepted virtually immediately throughout the world for scientific purposes. Thus, the big wave of triptan studies were all performed in patients diagnosed according to this classification. Slowly but surely the principles of the classification have also altered clinical practice. Many questions not needed in order to classify primary headaches are no longer being asked in clinical interviews and, conversely, a new criterion such as aggravation by physical activity is gradually being put to use in daily practice. The classification has been translated into more than 20 different languages and is thus available to the majority of doctors throughout the world.

When we published the first edition, we thought that a second edition of *The International Classification of Headache Disorders* would follow within five years because large parts of it were based on the opinion of experts rather than on published evidence. It took, however, 15 years until we now present the second edition and there are many good reasons for that. Relatively little criticism has prompted a revision. Nosographic research providing a better description of the clinical features of the different headache disorders has appeared only slowly and remains insufficient to allow a totally evidence-based classification. The world-wide dissemination of the English version of the first edition and the translation into more than 20 different languages has also taken much longer than we expected. Slowly, however, good suggestions for a revision accumulated and the epidemiological and nosographic knowledge increased to the extent where it became meaningful to start work on a second edition.

As for the first edition, I have also for this edition had the chairman's privilege of appointing the subcommittee members. Although the first committee did a fabulous job, it was my feeling that we should have a major replacement of membership in order to secure that the next generation of headache researchers be sufficiently represented. Consequently, the only members of the first committee who have also been members of the second are Giuseppe Nappi, James W Lance and I. We have been responsible for continuity. In appointing new members I have primarily paid attention to personal qualifications. Geographical representation and a wish to include persons with a well-argued critique against the first edition have also been taken into account. I am pleased to say that recruitment according to these principles proved successful. Every member has been vividly interested, outspoken and well-argued. Respecting the huge workload carried out by the first classification committee, the second committee openly debated any aspect of headache classification. Because of the meticulous work and the many fruitful discussions the second edition took much longer to do than any of us had expected. Every single set of criteria, every number and every word have been weighed carefully and tremendous effort and thought have been invested in the present publication. All the views of every member could not be taken into account, but every member has had considerable impact on the classification.

It is important for any field of medicine to have a generally-accepted classification that is used throughout the world. This is particularly true for headache as a young and developing field and because there is so much prejudice against headache disorders. Therefore, it is extremely important that the headache community at large and headache researchers in particular support the use of *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*. No journal should publish papers related to headache that are not using or examining this classification and the associated diagnostic criteria. On the other hand, our intention is not to lock headache research into a rigid frame and we therefore issue a strong plea to the world's headache researchers to examine this second edition

scientifically. In order to stimulate such studies, we have included an appendix which describes a number of orphan disorders that need validation. We also present a few alternative criteria that can be tested against the official ones.

I sincerely hope that this second edition of *The International Classification of Headache Disorders* will be received favourably by the headache community throughout the world and that it will be translated into even more languages than the first edition. Also, I hope that it will become a basis for world-wide teaching in headache classification and headache diagnosis and thereby benefit patient management. The International Headache Society works to improve the diagnosis, treatment and care of headache throughout the world. It also works to destigmatise headache sufferers and to gain recognition for these disorders as neurobiological conditions inflicting a very high burden on the sufferers and their relatives as well on society. It is imperative for the success of these efforts that researchers and clinicians as well as patients use the same diagnostic system and that this system is as precise as possible. This process was taken a long way by the first edition of *The International Classification of Headache Disorders*. The second edition will hopefully further promote unity in the way we classify, diagnose and treat headache patients throughout the world.

**Jes Olesen**  
**Chairman**  
**Headache Classification Subcommittee**  
**International Headache Society**

## **Preface to the First Revision**

The need for revision of the criteria for 8.2 *Medication-overuse headache* and its subforms became apparent soon after publication of *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*. Fred Sheftell first drew attention to potential problems with these criteria, and constructive criticism at the International Headache Research Seminar in Copenhagen in March, 2004, which discussed the classification in considerable detail, confirmed that some re-writing was necessary. This task was undertaken by Stephen Silberstein, chairman of the Working group on Headache Attributed to a Substance or its Withdrawal, together with the other members of the IHS Classification Subcommittee.

There are two major resultant changes. The first is the elimination of headache characteristics for the subforms of medication-overuse headache since it was agreed that none could be regarded as typical. The second is the introduction of a new subform, 8.2.6 *Medication-overuse headache attributed to combination of acute medications*, that recognises patients overusing medications of different classes whilst not meeting the criteria for overuse of any single class. This change required the re-numbering of subforms 8.2.7 (previously 8.2.6) *Headache attributed to other medication overuse* and 8.2.8 (previously 8.2.7) *Probable medication-overuse headache*.

## Introduction to the Classification

This second edition of *The International Classification of Headache Disorders*, like the first, is intended equally for research and for clinical practice. No research studies are likely to be accepted in international journals without adhering to this classification, but the classification is equally important for clinicians. The great majority of evidence-based treatments for headache have been developed using the first edition of *The International Classification of Headache Disorders*. This second edition has not changed the major principles of the classification and diagnosis of primary headache disorders. Therefore, the existing body of evidence gained using the first edition remains valid for most diagnoses made using the second edition. When you look for patients who will respond to a triptan, you must diagnose your patient according to the diagnostic criteria for migraine with aura and migraine without aura of this classification.

*The International Classification of Headache Disorders, 2<sup>nd</sup> edition*, is perhaps the single most important document to read for doctors taking an interest in the diagnosis and management of headache patients. There is often a huge gap between researchers and clinicians. Many have suggested that there should be two classifications, one for research and one for clinical use. However, if there were two classifications, all new information would be gathered using the research classification and transfer of results from research studies to clinical practice would be difficult. Therefore, the universally-accepted view of experts in disease classification is that there must be one classification only, but constructed so that it can be used at different levels of specialisation.

The answer to the problem is hierarchical classification and this system was already adopted by the first edition and remains unchanged in *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*. All headache disorders are classified into major groups and each group is then subdivided one, two or three times into headache types, subtypes and subforms. For example, 1. *Migraine* is a group consisting of one headache type (migraine) and the subtypes of migraine such as 1.2 *Migraine with aura* constitute the next level (second digit). Migraine with aura is again divided into subforms, for example 1.2.1 *Typical aura with migraine headache*. The practising family physician may only need to diagnose at the first level – migraine – in order to select acute treatment. However, when there is a problem of differential diagnosis, for example because headache is absent, it becomes necessary to distinguish between migraine with aura and other disorders that may mimic it, and thus to code at the second or third levels. Practising neurologists and headache specialists would normally diagnose the precise subform of migraine with aura at the third level. This system has proven utility at the different levels of healthcare systems throughout the world.

Classification means deciding on which kinds of diagnostic entities should be recognised and how to order them in a meaningful fashion. In doing so, one should draw upon all kinds of available evidence: clinical description, longitudinal studies of cohorts of patients, epidemiological studies, treatment results, genetics, neuroimaging and pathophysiology. This was done for the first edition and it has been repeated for the second edition of *The International Classification of Headache Disorders*. Fortunately, big changes have not been necessary, but a fairly large number of small but important changes have been made in the light of new evidence. Thus, we have introduced 1.5.1 *Chronic migraine* as a new diagnosis for those rare patients who fulfil the diagnostic criteria for migraine for 15 or more days a month without overusing medication. All secondary headaches are now described as “attributed to” another disorder while the first edition used the less-precise term “associated with”. The causal link between the underlying disorder and the headache is in most cases well-established, and we have therefore been able to strengthen the terminology.

With regard to psychiatric disorders, there is no reason to treat them differently from all other disorders that can cause secondary headaches. Therefore, we have included a new chapter 12. *Headache attributed to psychiatric disorder*. The problem is that research elucidating this field is extremely scarce so the chapter is very brief. The corresponding section in the appendix is more comprehensive and will hopefully greatly increase research into the relationship between psychiatric disorders and headache.

All headaches caused by infection are now placed in the same chapter 9. *Headache attributed to infection* whilst, previously, intracranial infections were placed in the chapter on intracranial disorders. A new chapter 10. *Headache attributed to disorder of homeostasis* has been added. Some new entities such as 4.5 *Hypnic headache*, 4.6 *Primary thunderclap headache* and 4.7 *Hemicrania continua* have also been added while 13.17 *Ophthalmoplegic "migraine"* has been moved from chapter 1. *Migraine* to chapter 12. *Cranial neuralgias and central causes of facial pain*.

As a major change in code-numbering, the tabulation below now includes WHO ICD-10NA codes (in parentheses) because these are the codes used in daily practice. In many places *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*, is more detailed than the WHO Classification. This means that some headache subtypes are not uniquely coded under the ICD-10NA system but the most appropriate ICD-10NA code has in each case been attached to the ICHD-II code.

The basic construct of each chapter in the second edition of *The International Classification of Headache Disorders* is the same as for the first edition. For each chapter the classification for that chapter is shown. Then there is an introduction and the different headaches are presented one by one in the order of the classification. For each major disease we give previously used terms and specify disorders that are related but coded elsewhere and we present short descriptions that in words try to define the disease. After that we present explicit diagnostic criteria. Finally, we provide written comments and a selected bibliography at the end of each chapter.

The explicit diagnostic criteria need a comment. Previously they were called *operational diagnostic criteria*, but the meaning of "operational" is not generally known. "Explicit" means "unambiguous, precise and with as little room for interpretation as possible". In other words, the aim is to write criteria so clearly and have such clear requirements that different doctors in different parts of the world are able to use them in the same way. Terms that are open to interpretation such as "sometimes", "often" or "usual" are largely avoided. Patients must fulfil all criteria listed as A, B, C, D, etc. For each criterion there are specific requirements such as "two of the following four characteristics", etc. The same system was used in the first edition where it proved to be reliable and reproducible. It has also been shown that the first edition was applicable in all settings spanning from epidemiological studies in the general population to tertiary headache referral centres. A validation of the first edition and its explicit diagnostic criteria was provided by the triptan studies, in which the rates of success were equal in different countries indicating that case ascertainment had been the same. Furthermore, the high success rate of injectable sumatriptan demonstrated that, at least from pathophysiological and pharmacological points of view, the diagnostic criteria for migraine with and without aura had delineated a fairly homogeneous entity. For this and many other reasons we have only made small changes in the diagnostic criteria for migraine.

Classification and diagnostic criteria can be aetiological or descriptive and the latter can be syndromic or symptom-based. Both first and second editions of *The International Classification of Headache Disorders* are aetiological for the secondary headaches and symptom-based for the primary headaches. If the course or the evolution of headache syndromes should be taken into account, there would have to be so much information available that the diagnosis of migraine, for

example, would enable the prediction of a particular course for that patient. The fact is that the evolution of primary headache syndromes cannot be predicted. Some patients will worsen and their symptoms become chronic, others will be relieved of their primary headache and yet others will stay the same for decades.

It is an important task for the future to provide prognostic factors and other characteristics that may make it possible to classify subtypes of migraine and tension-type headache. For some time it looked as if 3.1.2 *Chronic cluster headache* could be subdivided into *chronic from onset* and *evolving from episodic*, but then it was shown that a number of patient with chronic cluster headache reverted to 3.1.1 *Episodic cluster headache*. Thus, many different evolutionary patterns seem to be crossing each other. The same is true for migraine according to the longitudinal studies of Bille and others. For these reasons the evolutionary history cannot be classified until much bigger and better studies of the evolution of migraine patients become available.

Like the first, this second edition of *The International Classification of Headache Disorders* classifies patients according to the phenomenology of their headache(s). For clinical use, drug trials and pathophysiological studies this would normally mean that the patient must have had that type of headache within the last year and is likely to have further attacks. For other uses, particularly genetic studies, we are more concerned with the lifetime history of the patient. Thus, if the patient has had migraine attacks twenty years ago, but no attacks after that time, the patient still has the phenotype of migraine in a genetic study. These principles make it possible for a patient to have one diagnosis at one time and another diagnosis a few years later. It also makes it possible and necessary to give some patients more than one headache diagnosis and even two or more migraine diagnoses.

So far, only two migraine genes have been identified. They account for only half of patients with the rare disorder 1.2.4 *Familial hemiplegic migraine*. Thus, genetics have not had significant impact on *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*. However, it is expected that within the next ten years migraine genetics will be elucidated. This will undoubtedly lead to major alterations in the way we classify headaches, but it is not possible at present to say exactly how such changes will be. Some monogenic entities will probably be identified and it will become obvious that our clinically-defined phenotypes are heterogeneous. On the other hand, mutations in the same gene may cause quite different phenotypes as recently demonstrated by studies of familial hemiplegic migraine. Thus, the genetics of migraine may simply prove to be so complex that, in daily practice and perhaps to some extent in research, we shall continue with clinically-defined diagnoses.

A classification and its diagnostic criteria should be reliable, valid and exhaustive. Fortunately, as partly discussed above, the first edition of *The International Classification of Headache Disorders* has been shown to have fairly high degrees of reliability and validity. It has also proven to be exhaustive in several studies spanning from population-based studies to studies in headache clinics. We believe that the second edition is even more reliable, valid and exhaustive, but only future research can prove or disprove this belief.

## How to Use This Classification

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

1. This classification is hierarchical and you must decide how detailed you want to make your diagnosis. This can range from the first-digit level to the fourth. First one gets a rough idea about which group the patient belongs to. Is it for example 1. *Migraine* or 2. *Tension-type headache* or 3. *Cluster headache and other 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 whilst in specialist practice and headache centres a diagnosis at the third- or fourth-digit levels is appropriate.
2. Patients receive a diagnosis according to the headache phenotypes that they currently present or that they have presented within the last year. For genetic and some other uses, occurrence during the whole lifetime is used.
3. Each distinct type of headache that the patient has must be separately diagnosed and coded. Thus, a severely affected patient in a headache centre may receive three diagnoses and codes: 1.1 *Migraine without aura*, 2.2 *Frequent episodic tension-type headache* 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. If one type of headache in a particular patient fulfils two different sets of explicit diagnostic criteria, then all other available information should be used to decide which of the alternatives is the correct or more likely diagnosis. This could include the longitudinal headache history (how did the headache start?), the family history, the effect of drugs, menstrual relationship, age, gender and a range of other features. Fulfilment of the diagnostic criteria for 1. *Migraine*, 2. *Tension-type headache* or 3. *Cluster headache and other trigeminal autonomic cephalalgias*, or any of their subtypes, always trumps fulfilment of criteria for the *probable* diagnostic categories of each, which are last-described in the respective groups. In other words, a patient whose headache fulfils criteria for both 1.6 *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 whilst other attacks meet another set. In such cases, two diagnoses exist and both should be coded.
6. To receive a particular headache diagnosis the patient must, in many cases, experience a minimum number of attacks of (or days with) that headache. This number is specified in the explicit 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 explicit diagnostic criteria is provided for some headache disorders only at the first- and second-digit levels. Diagnostic criteria at the third- and fourth-digit levels then demand, as criterion A, fulfilment of the criteria for levels one and/or two and, in criterion B and onwards, specify the further specific criteria to be fulfilled.
8. The frequency of primary headache disorders varies from attacks every 1-2 years to attacks daily. The severity of attacks also varies. *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*, 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*: If a new headache occurs for the first time in close temporal relation to another disorder that is a known cause of headache, this headache is coded according to the causative disorder as a secondary headache. This remains true even when the headache has the characteristics of migraine, tension-type headache, cluster headache or one of the other trigeminal autonomic cephalalgias.

When a *pre-existing* primary headache is made worse in close temporal relation to another disorder that is a known cause of headache, there are two possibilities and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both the primary headache diagnosis and a secondary headache diagnosis according to the other disorder. Factors that support adding the secondary headache diagnosis are: a very close temporal relation to the causative disorder, a marked worsening of the primary headache, very good evidence that the causative disorder can aggravate the primary headache in the manner observed and, finally, improvement or disappearance of the headache after relief from the presumed causative disorder.

10. Many patients with headache attacks fulfilling one set of explicit diagnostic criteria also have attacks that, whilst 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.
11. When a patient is suspected of having more than one headache type 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.
12. In each chapter on the secondary headaches the most well-known and well-established causes are mentioned and criteria for these are given. However, in many chapters, for example 9. *Headache attributed to infection*, there are an almost endless number of possible causes. In order to avoid a very long list, only the most important are mentioned. In the example, rarer causes are assigned to 9.2.3 *Headache attributed to other infection*. The same system is used in the other chapters on secondary headaches.

13. The last criterion for most of the secondary headaches requires that the headache greatly improves or resolves within a specified period after relief from the causative disorder (through treatment or spontaneous remission). In such cases, fulfilment of this criterion is an essential part of the evidence for a causal relationship. Very often, there is a need to code patients before this disorder is treated or before the result of treatment is known. In such cases the diagnosis should be *Headache probably attributed to [the disorder]*. Once the treatment results are known, the diagnosis becomes *Headache attributed to [the disorder]*, or is changed if the criterion is not fulfilled.
14. In a few cases, post-traumatic headache being a good example, chronic headache subforms are recognised to occur. In such cases, the initially acute headache may persist, and causation is neither proved nor disproved by the duration of the headache in relation to onset of or relief from the causative disorder. The last criterion instead distinguishes between acute and chronic subforms, specifying resolution of headache within (for the acute subform) or persistence of headache beyond (for the chronic subform) a period of 3 months after occurrence, remission or cure of the causative disorder. In the course of the disorder, the diagnosis may therefore change after 3 months to *Chronic headache attributed to [the disorder]*. In the example, 5.1 *Acute post-traumatic headache* changes to 5.2 *Chronic post-traumatic 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.

## Classification

<b>IHS ICHD-II code</b>	<b>WHO ICD-10NA code</b>	<b>Diagnosis</b> [and aetiological ICD-10 code for secondary headache disorders]
<b>1.</b>	<b>[G43]</b>	<b>Migraine</b>
1.1	[G43.0]	Migraine without aura
1.2	[G43.1]	Migraine with aura
1.2.1	[G43.10]	Typical aura with migraine headache
1.2.2	[G43.10]	Typical aura with non-migraine headache
1.2.3	[G43.104]	Typical aura without headache
1.2.4	[G43.105]	Familial hemiplegic migraine (FHM)
1.2.5	[G43.105]	Sporadic hemiplegic migraine
1.2.6	[G43.103]	Basilar-type migraine
1.3	[G43.82]	Childhood periodic syndromes that are commonly precursors of migraine
1.3.1	[G43.82]	Cyclical vomiting
1.3.2	[G43.820]	Abdominal migraine
1.3.3	[G43.821]	Benign paroxysmal vertigo of childhood
1.4	[G43.81]	Retinal migraine
1.5	[G43.3]	Complications of migraine
1.5.1	[G43.3]	Chronic migraine
1.5.2	[G43.2]	Status migrainosus
1.5.3	[G43.3]	Persistent aura without infarction
1.5.4	[G43.3]	Migrainous infarction
1.5.5	[G43.3] + [G40.x or G41.x] <sup>1</sup>	Migraine-triggered seizures
1.6	[G43.83]	Probable migraine
1.6.1	[G43.83]	Probable migraine without aura
1.6.2	[G43.83]	Probable migraine with aura
1.6.5	[G43.83]	Probable chronic migraine
<b>2.</b>	<b>[G44.2]</b>	<b>Tension-type headache (TTH)</b>
2.1	[G44.2]	Infrequent episodic tension-type headache
2.1.1	[G44.20]	Infrequent episodic tension-type headache associated with pericranial tenderness
2.1.2	[G44.21]	Infrequent episodic tension-type headache not associated with pericranial tenderness
2.2	[G44.2]	Frequent episodic tension-type headache
2.2.1	[G44.20]	Frequent episodic tension-type headache associated with pericranial tenderness

<sup>1</sup> The additional code specifies the type of seizure.

2.2.2	[G44.21]	Frequent episodic tension-type headache not associated with pericranial tenderness
2.3	[G44.2]	Chronic tension-type headache
2.3.1	[G44.22]	Chronic tension-type headache associated with pericranial tenderness
2.3.2	[G44.23]	Chronic tension-type headache not associated with pericranial tenderness
2.4	[G44.28]	Probable tension-type headache
2.4.1	[G44.28]	Probable infrequent episodic tension-type headache
2.4.2	[G44.28]	Probable frequent episodic tension-type headache
2.4.3	[G44.28]	Probable chronic tension-type headache
<b>3.</b>	<b>[G44.0]</b>	<b>Cluster headache and other trigeminal autonomic cephalgias</b>
3.1	[G44.0]	Cluster headache
3.1.1	[G44.01]	Episodic cluster headache
3.1.2	[G44.02]	Chronic cluster headache
3.2	[G44.03]	Paroxysmal hemicrania
3.2.1	[G44.03]	Episodic paroxysmal hemicrania
3.2.2	[G44.03]	Chronic paroxysmal hemicrania (CPH)
3.3	[G44.08]	Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
3.4	[G44.08]	Probable trigeminal autonomic cephalgia
3.4.1	[G44.08]	Probable cluster headache
3.4.2	[G44.08]	Probable paroxysmal hemicrania
3.4.3	[G44.08]	Probable SUNCT
<b>4.</b>	<b>[G44.80]</b>	<b>Other primary headaches</b>
4.1	[G44.800]	Primary stabbing headache
4.2	[G44.803]	Primary cough headache
4.3	[G44.804]	Primary exertional headache
4.4	[G44.805]	Primary headache associated with sexual activity
4.4.1	[G44.805]	Preorgasmic headache
4.4.2	[G44.805]	Orgasmic headache
4.5	[G44.80]	Hypnic headache
4.6	[G44.80]	Primary thunderclap headache
4.7	[G44.80]	Hemicrania continua
4.8	[G44.2]	New daily-persistent headache (NDPH)
<b>5.</b>	<b>[G44.88]</b>	<b>Headache attributed to head and/or neck trauma</b>
5.1	[G44.880]	Acute post-traumatic headache
5.1.1	[G44.880]	Acute post-traumatic headache attributed to moderate or severe head injury [S06]
5.1.2	[G44.880]	Acute post-traumatic headache attributed to mild head injury [S09.9]
5.2	[G44.3]	Chronic post-traumatic headache
5.2.1	[G44.30]	Chronic post-traumatic headache attributed to moderate or severe

		head injury [S06]
5.2.2	[G44.31]	Chronic post-traumatic headache attributed to mild head injury [S09.9]
5.3	[G44.841]	Acute headache attributed to whiplash injury [S13.4]
5.4	[G44.841]	Chronic headache attributed to whiplash injury [S13.4]
5.5	[G44.88]	Headache attributed to traumatic intracranial haematoma
5.5.1	[G44.88]	Headache attributed to epidural haematoma [S06.4]
5.5.2	[G44.88]	Headache attributed to subdural haematoma [S06.5]
5.6	[G44.88]	Headache attributed to other head and/or neck trauma [S06]
5.6.1	[G44.88]	Acute headache attributed to other head and/or neck trauma [S06]
5.6.2	[G44.88]	Chronic headache attributed to other head and/or neck trauma [S06]
5.7	[G44.88]	Post-craniotomy headache
5.7.1	[G44.880]	Acute post-craniotomy headache
5.7.2	[G44.30]	Chronic post-craniotomy headache
<b>6.</b>	<b>[G44.81]</b>	<b>Headache attributed to cranial or cervical vascular disorder</b>
6.1	[G44.810]	Headache attributed to ischaemic stroke or transient ischaemic attack
6.1.1	[G44.810]	Headache attributed to ischaemic stroke (cerebral infarction) [I63]
6.1.2	[G44.810]	Headache attributed to transient ischaemic attack (TIA) [G45]
6.2	[G44.810]	Headache attributed to non-traumatic intracranial haemorrhage [I62]
6.2.1	[G44.810]	Headache attributed to intracerebral haemorrhage [I61]
6.2.2	[G44.810]	Headache attributed to subarachnoid haemorrhage (SAH) [I60]
6.3	[G44.811]	Headache attributed to unruptured vascular malformation [Q28]
6.3.1	[G44.811]	Headache attributed to saccular aneurysm [Q28.3]
6.3.2	[G44.811]	Headache attributed to arteriovenous malformation (AVM) [Q28.2]
6.3.3	[G44.811]	Headache attributed to dural arteriovenous fistula [I67.1]
6.3.4	[G44.811]	Headache attributed to cavernous angioma [D18.0]
6.3.5	[G44.811]	Headache attributed to encephalotrigeminal or leptomeningeal angiomatosis (Sturge Weber syndrome) [Q85.8]
6.4	[G44.812]	Headache attributed to arteritis [M31]
6.4.1	[G44.812]	Headache attributed to giant cell arteritis (GCA) [M31.6]
6.4.2	[G44.812]	Headache attributed to primary central nervous system (CNS) angiitis [I67.7]
6.4.3	[G44.812]	Headache attributed to secondary central nervous system (CNS) angiitis [I68.2]
6.5	[G44.810]	Carotid or vertebral artery pain [I63.0, I63.2, I65.0, I65.2 or I67.0]
6.5.1	[G44.810]	Headache or facial or neck pain attributed to arterial dissection [I67.0]
6.5.2	[G44.814]	Post-endarterectomy headache [I97.8]
6.5.3	[G44.810]	Carotid angioplasty headache
6.5.4	[G44.810]	Headache attributed to intracranial endovascular procedures
6.5.5	[G44.810]	Angiography headache
6.6	[G44.810]	Headache attributed to cerebral venous thrombosis (CVT) [I63.6]
6.7	[G44.81]	Headache attributed to other intracranial vascular disorder
6.7.1	[G44.81]	Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL) [I67.8]

6.7.2	[G44.81]	Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS) [G31.81]
6.7.3	[G44.81]	Headache attributed to benign angiopathy of the central nervous system [I99]
6.7.4	[G44.81]	Headache attributed to pituitary apoplexy [E23.6]
<b>7.</b>	<b>[G44.82]</b>	<b>Headache attributed to non-vascular intracranial disorder</b>
7.1	[G44.820]	Headache attributed to high cerebrospinal fluid pressure
7.1.1	[G44.820]	Headache attributed to idiopathic intracranial hypertension (IIH) [G93.2]
7.1.2	[G44.820]	Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes
7.1.3	[G44.820]	Headache attributed to intracranial hypertension secondary to hydrocephalus [G91.8]
7.2	[G44.820]	Headache attributed to low cerebrospinal fluid pressure
7.2.1	[G44.820]	Post-dural puncture headache [G97.0]
7.2.2	[G44.820]	CSF fistula headache [G96.0]
7.2.3	[G44.820]	Headache attributed to spontaneous (or idiopathic) low CSF pressure
7.3	[G44.82]	Headache attributed to non-infectious inflammatory disease
7.3.1	[G44.823]	Headache attributed to neurosarcoidosis [D86.8]
7.3.2	[G44.823]	Headache attributed to aseptic (non-infectious) meningitis [code to specify aetiology]
7.3.3	[G44.823]	Headache attributed to other non-infectious inflammatory disease [code to specify aetiology]
7.3.4	[G44.82]	Headache attributed to lymphocytic hypophysitis [E23.6]
7.4	[G44.822]	Headache attributed to intracranial neoplasm [C00-D48]
7.4.1	[G44.822]	Headache attributed to increased intracranial pressure or hydrocephalus caused by neoplasm [code to specify neoplasm]
7.4.2	[G44.822]	Headache attributed directly to neoplasm [code to specify neoplasm]
7.4.3	[G44.822]	Headache attributed to carcinomatous meningitis [C79.3]
7.4.4	[G44.822]	Headache attributed to hypothalamic or pituitary hyper- or hyposecretion [E23.0]
7.5	[G44.824]	Headache attributed to intrathecal injection [G97.8]
7.6	[G44.82]	Headache attributed to epileptic seizure [G40.x or G41.x to specify seizure type]
7.6.1	[G44.82]	Hemicrania epileptica [G40.x or G41.x to specify seizure type]
7.6.2	[G44.82]	Post-seizure headache [G40.x or G41.x to specify seizure type]
7.7	[G44.82]	Headache attributed to Chiari malformation type I (CM1) [Q07.0]
7.8	[G44.82]	Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)
7.9	[G44.82]	Headache attributed to other non-vascular intracranial disorder

<b>8.</b>	<b>[G44.4 or G44.83]</b>	<b>Headache attributed to a substance<sup>2</sup> or its withdrawal</b>
8.1	[G44.40]	Headache induced by acute substance use or exposure
8.1.1	[G44.400]	Nitric oxide (NO) donor-induced headache [X44]
8.1.1.1	[G44.400]	Immediate NO donor-induced headache [X44]
8.1.1.2	[G44.400]	Delayed NO donor-headache [X44]
8.1.2	[G44.40]	Phosphodiesterase (PDE) inhibitor-induced headache [X44]
8.1.3	[G44.402]	Carbon monoxide-induced headache [X47]
8.1.4	[G44.83]	Alcohol-induced headache [F10]
8.1.4.1	[G44.83]	Immediate alcohol-induced headache [F10]
8.1.4.2	[G44.83]	Delayed alcohol-induced headache [F10]
8.1.5	[G44.4]	Headache induced by food components and additives
8.1.5.1	[G44.401]	Monosodium glutamate-induced headache [X44]
8.1.6	[G44.83]	Cocaine-induced headache [F14]
8.1.7	[G44.83]	Cannabis-induced headache [F12]
8.1.8	[G44.40]	Histamine-induced headache [X44]
8.1.8.1	[G44.40]	Immediate histamine-induced headache [X44]
8.1.8.2	[G44.40]	Delayed histamine-induced headache [X44]
8.1.9	[G44.40]	Calcitonin gene-related peptide (CGRP)-induced headache [X44]
8.1.9.1	[G44.40]	Immediate CGRP-induced headache [X44]
8.1.9.2	[G44.40]	Delayed CGRP-induced headache [X44]
8.1.10	[G44.41]	Headache as an acute adverse event attributed to medication used for other indications [code to specify substance]
8.1.11	[G44.4 or G44.83]	Headache attributed to other acute substance use or exposure [code to specify substance]
8.2	[G44.41 or G44.83]	Medication-overuse headache (MOH)
8.2.1	[G44.411]	Ergotamine-overuse headache [Y52.5]
8.2.2	[G44.41]	Triptan-overuse headache
8.2.3	[G44.410]	Analgesic-overuse headache [F55.2]
8.2.4	[G44.83]	Opioid-overuse headache [F11.2]
8.2.5	[G44.410]	Combination analgesic-overuse headache [F55.2]
8.2.6	[G44.41 ± G44.83]	Medication-overuse headache attributed to combination of acute medications
8.2.7	[G44.410]	Headache attributed to other medication overuse [code to specify substance]
8.2.8	[G44.41 or G44.83]	Probable medication-overuse headache [code to specify substance]
8.3	[G44.4]	Headache as an adverse event attributed to chronic medication [code to specify substance]
8.3.1	[G44.418]	Exogenous hormone-induced headache [Y42.4]

<sup>2</sup> In ICD-10 substances are classified according to the presence or absence of a dependence-producing property. Headaches associated with psychoactive substances (dependence-producing) are classified in G44.83 with an additional code to indicate the nature of the disorder related to the substance use: *eg*, intoxication (F1x.0), dependence (F1x.2), withdrawal (F1x.3), *etc*. The 3rd character can be used to indicate the specific substance involved: *eg*, F10 for alcohol, F15 for caffeine, *etc*. Abuse of non-dependence-producing substances is classified in F55, with a 4th character to indicate the substance: *eg*, F55.2 abuse of analgesics. Headaches related to non-dependence-producing substances are classified in G44.4.

- 8.4 [G44.83] Headache attributed to substance withdrawal
  - 8.4.1 [G44.83] Caffeine-withdrawal headache [F15.3]
  - 8.4.2 [G44.83] Opioid-withdrawal headache [F11.3]
  - 8.4.3 [G44.83] Oestrogen-withdrawal headache [Y42.4]
  - 8.4.4 [G44.83] Headache attributed to withdrawal from chronic use of other substances [code to specify substance]

**9. Headache attributed to infection**

- 9.1 [G44.821] Headache attributed to intracranial infection [G00-G09]
  - 9.1.1 [G44.821] Headache attributed to bacterial meningitis [G00.9]
  - 9.1.2 [G44.821] Headache attributed to lymphocytic meningitis [G03.9]
  - 9.1.3 [G44.821] Headache attributed to encephalitis [G04.9]
  - 9.1.4 [G44.821] Headache attributed to brain abscess [G06.0]
  - 9.1.5 [G44.821] Headache attributed to subdural empyema [G06.2]
- 9.2 [G44.881] Headache attributed to systemic infection [A00-B97]
  - 9.2.1 [G44.881] Headache attributed to systemic bacterial infection [code to specify aetiology]
  - 9.2.2 [G44.881] Headache attributed to systemic viral infection [code to specify aetiology]
  - 9.2.3 [G44.881] Headache attributed to other systemic infection [code to specify aetiology]
- 9.3 [G44.821] Headache attributed to HIV/AIDS [B22]
- 9.4 [G44.821 or G44.881] Chronic post-infection headache [code to specify aetiology]
  - 9.4.1 [G44.821] Chronic post-bacterial meningitis headache [G00.9]

**10. [G44.882] Headache attributed to disorder of homeostasis**

- 10.1 [G44.882] Headache attributed to hypoxia and/or hypercapnia
  - 10.1.1 [G44.882] High-altitude headache [W94]
  - 10.1.2 [G44.882] Diving headache
  - 10.1.3 [G44.882] Sleep apnoea headache [G47.3]
- 10.2 [G44.882] Dialysis headache [Y84.1]
- 10.3 [G44.813] Headache attributed to arterial hypertension [I10]
  - 10.3.1 [G44.813] Headache attributed to pheochromocytoma [D35.0 (benign) or C74.1 (malignant)]
  - 10.3.2 [G44.813] Headache attributed to hypertensive crisis without hypertensive encephalopathy [I10]
  - 10.3.3 [G44.813] Headache attributed to hypertensive encephalopathy [I67.4]
  - 10.3.4 [G44.813] Headache attributed to pre-eclampsia [O13-O14]
  - 10.3.5 [G44.813] Headache attributed to eclampsia [O15]
  - 10.3.6 [G44.813] Headache attributed to acute pressor response to an exogenous agent [code to specify aetiology]
- 10.4 [G44.882] Headache attributed to hypothyroidism [E03.9]
- 10.5 [G44.882] Headache attributed to fasting [T73.0]
- 10.6 [G44.882] Cardiac cephalalgia [code to specify aetiology]

10.7	[G44.882]	Headache attributed to other disorder of homoeostasis [code to specify aetiology]
<b>11.</b>	<b>[G44.84]</b>	<b>Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures</b>
11.1	[G44.840]	Headache attributed to disorder of cranial bone [M80-M89.8]
11.2	[G44.841]	Headache attributed to disorder of neck [M99]
11.2.1	[G44.841]	Cervicogenic headache [M99]
11.2.2	[G44.842]	Headache attributed to retropharyngeal tendonitis [M79.8]
11.2.3	[G44.841]	Headache attributed to craniocervical dystonia [G24]
11.3	[G44.843]	Headache attributed to disorder of eyes
11.3.1	[G44.843]	Headache attributed to acute glaucoma [H40]
11.3.2	[G44.843]	Headache attributed to refractive errors [H52]
11.3.3	[G44.843]	Headache attributed to heterophoria or heterotropia (latent or manifest squint) [H50.3-H50.5]
11.3.4	[G44.843]	Headache attributed to ocular inflammatory disorder [code to specify aetiology]
11.4	[G44.844]	Headache attributed to disorder of ears [H60-H95]
11.5	[G44.845]	Headache attributed to rhinosinusitis [J01]
11.6	[G44.846]	Headache attributed to disorder of teeth, jaws or related structures [K00-K14]
11.7	[G44.846]	Headache or facial pain attributed to temporomandibular joint (TMJ) disorder [K07.6]
11.8	[G44.84]	Headache attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structures [code to specify aetiology]
<b>12.</b>	<b>[R51]</b>	<b>Headache attributed to psychiatric disorder</b>
12.1	[R51]	Headache attributed to somatisation disorder [F45.0]
12.2	[R51]	Headache attributed to psychotic disorder [code to specify aetiology]
<b>13.</b>	<b>[G44.847, G44.848 or G44.85]</b>	<b>Cranial neuralgias and central causes of facial pain</b>
13.1	[G44.847]	Trigeminal neuralgia
13.1.1	[G44.847]	Classical trigeminal neuralgia [G50.00]
13.1.2	[G44.847]	Symptomatic trigeminal neuralgia [G53.80] + [code to specify aetiology]
13.2	[G44.847]	Glossopharyngeal neuralgia
13.2.1	[G44.847]	Classical glossopharyngeal neuralgia [G52.10]
13.2.2	[G44.847]	Symptomatic glossopharyngeal neuralgia [G53.830] + [code to specify aetiology]
13.3	[G44.847]	Nervus intermedius neuralgia [G51.80]
13.4	[G44.847]	Superior laryngeal neuralgia [G52.20]

13.5	[G44.847]	Nasociliary neuralgia [G52.80]
13.6	[G44.847]	Supraorbital neuralgia [G52.80]
13.7	[G44.847]	Other terminal branch neuralgias [G52.80]
13.8	[G44.847]	Occipital neuralgia [G52.80]
13.9	[G44.851]	Neck-tongue syndrome
13.10	[G44.801]	External compression headache
13.11	[G44.802]	Cold-stimulus headache
13.11.1	[G44.8020]	Headache attributed to external application of a cold stimulus
13.11.2	[G44.8021]	Headache attributed to ingestion or inhalation of a cold stimulus
13.12	[G44.848]	Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions [G53.8] + [code to specify aetiology]
13.13	[G44.848]	Optic neuritis [H46]
13.14	[G44.848]	Ocular diabetic neuropathy [E10-E14]
13.15	[G44.881 or G44.847]	Head or facial pain attributed to herpes zoster
13.15.1	[G44.881]	Head or facial pain attributed to acute herpes zoster [B02.2]
13.15.2	[G44.847]	Post-herpetic neuralgia [B02.2]
13.16	[G44.850]	Tolosa-Hunt syndrome
13.17	[G43.80]	Ophthalmoplegic “migraine”
13.18	[G44.810 or G44.847]	Central causes of facial pain
13.18.1	[G44.847]	Anaesthesia dolorosa [G52.800] + [code to specify aetiology]
13.18.2	[G44.810]	Central post-stroke pain [G46.21]
13.18.3	[G44.847]	Facial pain attributed to multiple sclerosis [G35]
13.18.4	[G44.847]	Persistent idiopathic facial pain [G50.1]
13.18.5	[G44.847]	Burning mouth syndrome [code to specify aetiology]
13.19	[G44.847]	Other cranial neuralgia or other centrally mediated facial pain [code to specify aetiology]
<b>14.</b>	<b>[R51]</b>	<b>Other headache, cranial neuralgia, central or primary facial pain</b>
14.1	[R51]	Headache not elsewhere classified
14.2	[R51]	Headache unspecified

# **PART ONE**

## **THE PRIMARY HEADACHES**

Migraine

Tension-type headache

Cluster headache and other trigeminal autonomic cephalalgias

Other primary headaches

## 1. MIGRAINE

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

*Coded elsewhere:*

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

### General comment

*Primary or secondary headache or both?*

When a headache with migraine characteristics occurs for the first time in close temporal relation to another disorder that is a known cause of headache, it is coded according to the causative disorder as a secondary headache. When pre-existing migraine is made worse in close temporal relation to another disorder that is a known cause of headache, there are two possibilities, and judgment is required. The patient can either be given only the migraine diagnosis or be given both the migraine diagnosis and a secondary headache diagnosis according to the other disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the disorder, a marked worsening of the migraine, very good evidence that the disorder can cause or aggravate migraine, and improvement or resolution of migraine after relief from the disorder.

### Introduction

Migraine is a common disabling primary headache disorder. Epidemiological studies have documented its high prevalence and high socio-economic and personal impacts. It is now ranked by the World Health Organization as number 19 among all diseases world-wide causing disability.

Migraine can be divided into two major sub-types. 1.1 *Migraine without aura* is a clinical syndrome characterised by headache with specific features and associated symptoms. 1.2 *Migraine with aura* is primarily characterised by the focal neurological symptoms that usually precede or sometimes accompany the headache. Some patients also experience a premonitory phase, occurring hours or days before the headache, and a headache resolution phase. Premonitory and resolution symptoms include hyperactivity, hypoactivity, depression, craving for particular foods, repetitive yawning and other less typical symptoms reported by some patients.

When a patient fulfils criteria for more than one subtype of migraine, all subtypes should be diagnosed and coded. For example, a patient who has frequent attacks with aura but also some attacks without aura should be coded as 1.2 *Migraine with aura* and 1.1 *Migraine without aura*.

## 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 5 attacks<sup>1</sup> fulfilling criteria B-D
- B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)<sup>2;3;4</sup>
- C. Headache has at least two of the following characteristics:
  - 1. unilateral location<sup>5;6</sup>
  - 2. pulsating quality<sup>7</sup>
  - 3. moderate or severe pain intensity
  - 4. aggravation by or causing avoidance of routine physical activity (*eg*, walking or climbing stairs)
- D. During headache at least one of the following:
  - 1. nausea and/or vomiting
  - 2. photophobia and phonophobia<sup>8</sup>
- E. Not attributed to another disorder<sup>9</sup>

*Notes:*

- 1. Differentiating between 1.1 *Migraine without aura* and 2.1 *Infrequent episodic tension-type headache* may be difficult. Therefore at least 5 attacks are required. Individuals who otherwise meet criteria for 1.1 *Migraine without aura* but have had fewer than 5 attacks should be coded 1.6.1 *Probable migraine without aura*.
- 2. When the patient falls asleep during migraine and wakes up without it, duration of the attack is reckoned until the time of awakening.
- 3. In children, attacks may last 1-72 hours (although the evidence for untreated durations of less than 2 hours in children requires corroboration by prospective diary studies).

4. When attacks occur on  $\geq 15$  days/month for  $>3$  months, code as 1.1 *Migraine without aura* and as 1.5.1 *Chronic migraine*.
5. Migraine headache is commonly bilateral in young children; an adult pattern of unilateral pain usually emerges in late adolescence or early adult life.
6. Migraine headache is usually frontotemporal. Occipital headache in *children*, whether unilateral or bilateral, is rare and calls for diagnostic caution; many cases are attributable to structural lesions.
7. *Pulsating* means throbbing or varying with the heartbeat.
8. In young children, photophobia and phonophobia may be inferred from their behaviour.
9. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

*Comments:*

1.1 *Migraine without aura* is the commonest subtype of migraine. It has a higher average attack frequency and is usually more disabling than 1.2 *Migraine with aura*.

Migraine without aura often has a strict menstrual relationship. In contrast to the first edition of *The International Classification of Headache Disorders*, this edition gives criteria for A1.1.1 *Pure menstrual migraine* and A1.1.2 *Menstrually-related migraine*, but in the appendix because of uncertainty over whether they should be regarded as separate entities.

Very frequent migraine attacks are now distinguished as 1.5.1 *Chronic migraine* provided that there is no medication overuse. Migraine without aura is the disease most prone to accelerate with frequent use of symptomatic medication, resulting in a new headache which is coded as 8.2 *Medication-overuse headache*.

Regional cerebral blood flow shows no changes suggestive of cortical spreading depression during attacks of 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 migraine with aura. In all likelihood spreading depression is therefore not involved in migraine without aura. On the other hand the messenger molecules nitric oxide (NO) and calcitonin-gene-related peptide (CGRP) are clearly involved. While the disease was previously regarded as primarily vascular, the importance of sensitisation of perivascular nerve terminals, 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 and several aspects of neurotransmission in this system have been recognised. A significant contribution has been made by the advent of the triptans, 5HT<sub>1B/D</sub> receptor agonists. These drugs have remarkable efficacy in acute attacks and, in view of their high receptor-specificity, their mechanism of action provides new insight into migraine mechanisms. It is now clear that migraine without aura is a neurobiological disorder and clinical as well as basic neuroscience currently advances our knowledge of migraine mechanisms at an increasing speed.

## 1.2 Migraine with aura

### *Previously used terms:*

Classic or classical migraine, ophthalmic, hemiparaesthetic, hemiplegic or aphasic migraine, migraine accompagnée, complicated migraine

### *Coded elsewhere:*

13.17 *Ophthalmoplegic "migraine"*.

### *Description:*

Recurrent disorder manifesting in attacks of reversible focal neurological symptoms that usually develop gradually over 5-20 minutes and last for less than 60 minutes. Headache with the features of migraine without aura usually follows the aura symptoms. Less commonly, headache lacks migrainous features or is completely absent.

### *Diagnostic criteria:*

- A. At least 2 attacks fulfilling criterion B
- B. Migraine aura fulfilling criteria B and C for one of the subforms 1.2.1-1.2.6
- C. Not attributed to another disorder<sup>1</sup>

### *Note:*

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

### *Comments:*

The aura is the complex of neurological symptoms that occurs just before or at the onset of migraine headache. Most patients with migraine have exclusively attacks without aura. Many patients who have frequent attacks with aura also have attacks without aura (code as 1.2 *Migraine with aura* and 1.1 *Migraine without aura*).

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

The majority of migraine auras are associated with headache fulfilling criteria for 1.1 *Migraine without aura*. For this reason the entity 1.2.1 *Typical aura with migraine headache* has been singled out below. Migraine aura is sometimes associated with a headache that does not fulfil criteria for migraine without aura and, in other cases, migraine aura may occur without headache. These two subforms are also now distinguished.

Aura with similar features has also been described in association with other well-defined headache types, including cluster headache; the relationships between aura and headache are not fully understood.

Before or simultaneously with the onset of aura symptoms, regional cerebral blood flow is decreased in cortex corresponding to the clinically affected area and often including an even 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 has been implicated.

Systematic studies have demonstrated that many patients with visual auras occasionally have symptoms in the extremities. Conversely patients with symptoms in the extremities virtually always also suffer visual aura symptoms. A distinction between migraine with visual aura and hemiparaesthetic migraine is probably artificial and therefore is not recognised in this classification. Patients with motor weakness are classified separately because of the dominantly inherited form, 1.2.4 *Familial hemiplegic migraine*, and because of clinical differences. The genetic relationship between migraine with aura and familial hemiplegic migraine has not been established.

The previously-defined syndromes *migraine with prolonged aura* and *migraine with acute-onset aura* have been abandoned. The great majority of patients with such attacks have other attacks that fulfil criteria for one of the subforms of 1.2 *Migraine with aura* and should be coded to that diagnosis. The rest should be coded to 1.6.2 *Probable migraine with aura*, specifying the atypical feature (prolonged aura or acute-onset aura) in parenthesis.

### 1.2.1 *Typical aura with migraine headache*

#### *Description:*

Typical aura consisting of visual and/or sensory and/or speech symptoms. Gradual development, duration no longer than one hour, a mix of positive and negative features and complete reversibility characterise the aura which is associated with a headache fulfilling criteria for 1.1 *Migraine without aura*.

#### *Diagnostic criteria:*

- A. At least 2 attacks fulfilling criteria B–D
- B. Aura consisting of at least one of the following, but no motor weakness:
  - 1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
  - 2. fully reversible sensory symptoms including positive features (*ie*, pins and needles) and/or negative features (*ie*, numbness)
  - 3. fully reversible dysphasic speech disturbance
- C. At least two of the following:
  - 1. homonymous visual symptoms<sup>1</sup> and/or unilateral sensory symptoms
  - 2. at least one aura symptom develops gradually over  $\geq 5$  minutes and/or different aura symptoms occur in succession over  $\geq 5$  minutes
  - 3. each symptom lasts  $\geq 5$  and  $\leq 60$  minutes
- D. Headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows aura within 60 minutes
- E. Not attributed to another disorder<sup>2</sup>

#### *Notes:*

- 1. Additional loss or blurring of central vision may occur.
- 2. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest

such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

*Comments:*

This is the most common migraine syndrome associated with aura. The diagnosis is usually evident after a careful history alone though there are rare secondary mimics including carotid dissection, arteriovenous malformation and seizure.

Visual aura is the most common type of aura, often presenting as a fortification spectrum, *ie*, 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 variable degrees of absolute or 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. 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 and face. Numbness may occur in its wake, but numbness may also be the only symptom. Less frequent are speech disturbances, usually dysphasic but often hard to categorise. If the aura includes motor weakness, code as 1.2.4 *Familial hemiplegic migraine* or 1.2.5 *Sporadic hemiplegic migraine*.

Symptoms usually follow one another in succession beginning with visual, then sensory symptoms and dysphasia, but the reverse and other orders have been noted. Patients often find it hard to describe their symptoms in which case they should be instructed in how to time and record them. After such prospective observation the clinical picture often becomes clearer. Common mistakes are incorrect reports of lateralisation of headache, of sudden onset when it is gradual and of monocular visual disturbances when they are homonymous, as well as incorrect duration of aura and mistaking sensory loss for weakness. After an initial consultation, use of an aura diary may clarify the diagnosis.

### **1.2.2 Typical aura with non-migraine headache**

*Description:*

Typical aura consisting of visual and/or sensory and/or speech symptoms. Gradual development, duration no longer than one hour, a mix of positive and negative features and complete reversibility characterise the aura which is associated with a headache that does not fulfil criteria for 1.1

*Migraine without aura.*

*Diagnostic criteria:*

- A. At least 2 attacks fulfilling criteria B–D
- B. Aura consisting of at least one of the following, but no motor weakness:
  - 1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
  - 2. fully reversible sensory symptoms including positive features (*ie*, pins and needles) and/or negative features (*ie*, numbness)
  - 3. fully reversible dysphasic speech disturbance
- C. At least two of the following:
  - 1. homonymous visual symptoms<sup>1</sup> and/or unilateral sensory symptoms
  - 2. at least one aura symptom develops gradually over  $\geq 5$  minutes and/or different aura symptoms occur in succession over  $\geq 5$  minutes
  - 3. each symptom lasts  $\geq 5$  and  $\leq 60$  minutes

- D. Headache that does not fulfil criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows aura within 60 minutes
- E. Not attributed to another disorder<sup>2</sup>

*Notes:*

1. Additional loss or blurring of central vision may occur.
2. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

*Comment:*

In the absence of headache fulfilling criteria for 1.1 *Migraine without aura*, precise diagnosis of aura and its distinction from mimics that may signal serious disease (*eg*, transient ischaemic attack) become much more important.

### **1.2.3 Typical aura without headache**

*Description:*

Typical aura consisting of visual and/or sensory symptoms with or without speech symptoms. Gradual development, duration no longer than one hour, a mix of positive and negative features and complete reversibility characterise the aura which is not associated with headache.

*Diagnostic criteria:*

- A. At least 2 attacks fulfilling criteria B–D
- B. Aura consisting of at least one of the following, with or without speech disturbance but no motor weakness:
  1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
  2. fully reversible sensory symptoms including positive features (*ie*, pins and needles) and/or negative features (*ie*, numbness)
- C. At least two of the following:
  1. homonymous visual symptoms<sup>1</sup> and/or unilateral sensory symptoms
  2. at least one aura symptom develops gradually over  $\geq 5$  minutes and/or different aura symptoms occur in succession over  $\geq 5$  minutes
  3. each symptom lasts  $\geq 5$  and  $\leq 60$  minutes
- D. Headache does not occur during aura nor follow aura within 60 minutes
- E. Not attributed to another disorder<sup>2</sup>

*Notes:*

1. Additional loss or blurring of central vision may occur.
2. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

*Comments:*

In some patients a typical aura is always followed by migraine headache, but many patients have, in addition, attacks with aura followed by non-migraine headache or even without headache. A small number of patients have 1.2.3 *Typical aura without headache* exclusively. More commonly, as patients with 1.2.1 *Typical aura with migraine headache* become older, their headache may lose migraine characteristics or disappear completely even though auras continue. Some individuals, primarily males, have 1.2.3 *Typical aura without headache* from onset.

In the absence of headache fulfilling criteria for 1.1 *Migraine without aura*, precise diagnosis of aura and its distinction from mimics that may signal serious disease (*eg*, transient ischaemic attack) become much more important. This distinction may require investigation. Especially if aura begins after age 40, if negative features (*eg*, hemianopia) are predominant, or if aura is prolonged or very short, other causes should be ruled out.

#### **1.2.4 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. At least 2 attacks fulfilling criteria B and C
- B. Aura consisting of fully reversible motor weakness and at least one of the following:
  - 1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
  - 2. fully reversible sensory symptoms including positive features (*ie*, pins and needles) and/or negative features (*ie*, numbness)
  - 3. fully reversible dysphasic speech disturbance
- C. At least two of the following:
  - 1. at least one aura symptom develops gradually over  $\geq 5$  minutes and/or different aura symptoms occur in succession over  $\geq 5$  minutes
  - 2. each aura symptom lasts  $\geq 5$  minutes and  $< 24$  hours
  - 3. headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows onset of aura within 60 minutes
- D. At least one first- or second-degree relative has had attacks fulfilling these criteria A-E
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

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

*Comments:*

It may be difficult to distinguish weakness from sensory loss.

New genetic data have allowed a more precise definition of FHM than previously. Specific genetic subtypes of 1.2.4 *Familial hemiplegic migraine* have been identified: in FHM1 there are mutations

in the CACNA1A gene on chromosome 19, and in FHM2 mutations occur in the ATP1A2 gene on chromosome 1. If genetic testing is done, the genetic subtype should be specified parenthetically.

It has been shown that FHM1 very often has basilar-type symptoms in addition to the typical aura symptoms and that headache is virtually always present. During FHM1 attacks, disturbances of consciousness (sometimes including coma), fever, CSF pleocytosis and confusion can occur. FHM1 attacks can be triggered by (mild) head trauma. In approximately 50% of FHM1 families, chronic progressive cerebellar ataxia occurs independently of the migraine attacks.

FHM is very often mistaken for epilepsy, and (unsuccessfully) treated as such.

### ***1.2.5 Sporadic hemiplegic migraine***

#### *Description:*

Migraine with aura including motor weakness but no first- or second-degree relative has aura including motor weakness.

#### *Diagnostic criteria:*

- A. At least 2 attacks fulfilling criteria Band C
- B. Aura consisting of fully reversible motor weakness and at least one of the following:
  - 1. fully reversible visual symptoms including positive features (*eg*, flickering lights, spots or lines) and/or negative features (*ie*, loss of vision)
  - 2. fully reversible sensory symptoms including positive features (*ie*, pins and needles) and/or negative features (*ie*, numbness)
  - 3. fully reversible dysphasic speech disturbance
- C. At least two of the following:
  - 1. at least one aura symptom develops gradually over  $\geq 5$  minutes and/or different aura symptoms occur in succession over  $\geq 5$  minutes
  - 2. each aura symptom lasts  $\geq 5$  minutes and  $< 24$  hours
  - 3. headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows onset of aura within 60 minutes
- D. No first- or second-degree relative has attacks fulfilling these criteria A-E
- E. Not attributed to another disorder<sup>1</sup>

#### *Note:*

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

#### *Comments:*

Epidemiological studies have shown that sporadic cases occur with approximately the same prevalence as familial cases. The attacks have the same clinical characteristics as those in 1.2.4 *Familial hemiplegic migraine*.

Sporadic cases always require neuroimaging and other tests to rule out other cause. A lumbar puncture is also necessary to rule out pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis. This condition is more prevalent in males and often associated with transient hemiparesis and aphasia.

### 1.2.6 *Basilar-type migraine*

#### *Previously used terms:*

Basilar artery migraine, basilar migraine

#### *Description:*

Migraine with aura symptoms clearly originating from the brainstem and/or from both hemispheres simultaneously affected, but no motor weakness.

#### *Diagnostic criteria:*

- A. At least 2 attacks fulfilling criteria B-D
- B. Aura consisting of at least two of the following fully reversible symptoms, but no motor weakness:
  - 1. dysarthria
  - 2. vertigo
  - 3. tinnitus
  - 4. hypacusia
  - 5. diplopia
  - 6. visual symptoms simultaneously in both temporal and nasal fields of both eyes
  - 7. ataxia
  - 8. decreased level of consciousness
  - 9. simultaneously bilateral paraesthesias
- C. At least one of the following:
  - 1. at least one aura symptom develops gradually over  $\geq 5$  minutes and/or different aura symptoms occur in succession over  $\geq 5$  minutes
  - 2. each aura symptom lasts  $\geq 5$  and  $\leq 60$  minutes
- D. Headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the aura or follows aura within 60 minutes
- E. Not attributed to another disorder<sup>1</sup>

#### *Note:*

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

#### *Comments:*

Basilar-type attacks are mostly seen in young adults. Many patients who have basilar-type attacks also report attacks with typical aura (code for both disorders).

If motor weakness is present, code as 1.2.4 *Familial hemiplegic migraine* or 1.2.5 *Sporadic hemiplegic migraine*. Patients with 1.2.4 *Familial hemiplegic migraine* have basilar-type symptoms in 60% of cases. Therefore, 1.2.6 *Basilar-type migraine* should be diagnosed only when no motor weakness occurs.

Many of the symptoms listed under criterion B are subject to misinterpretation as they may occur with anxiety and hyperventilation.

Originally the terms *basilar artery migraine* or *basilar migraine* were used but, since involvement of the basilar artery territory is uncertain (*ie*, the disturbance may be bihemispheric), the term *basilar-type migraine* is preferred.

### 1.3 Childhood periodic syndromes that are commonly precursors of migraine

#### 1.3.1 Cyclical vomiting

##### *Description:*

Recurrent episodic attacks, usually stereotypical in the individual patient, of vomiting and intense nausea. Attacks are associated with pallor and lethargy. There is complete resolution of symptoms between attacks.

##### *Diagnostic criteria:*

- A. At least 5 attacks fulfilling criteria Band C
- B. Episodic attacks, stereotypical in the individual patient, of intense nausea and vomiting lasting from 1 hour to 5 days
- C. Vomiting during attacks occurs at least 4 times/hour for at least 1 hour
- D. Symptom-free 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.

##### *Comment:*

Cyclical vomiting is a self-limiting episodic condition of childhood, with periods of complete normality between episodes. This disorder was not included as a childhood periodic syndrome in the first edition of *The International Classification of Headache Disorders*. 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 cyclical vomiting is a condition related to migraine.

#### 1.3.2 Abdominal migraine

##### *Description:*

An idiopathic recurrent disorder seen mainly in children and characterised by episodic midline abdominal pain manifesting in attacks lasting 1-72 hours with normality between episodes. The pain is of moderate to severe intensity and associated with vasomotor symptoms, nausea and vomiting.

##### *Diagnostic criteria:*

- A. At least 5 attacks fulfilling criteria B-D
- B. Attacks of abdominal pain lasting 1-72 hours (untreated or unsuccessfully treated)
- C. Abdominal pain has all of the following characteristics:
  1. midline location, periumbilical or poorly localised
  2. dull or “just sore” quality
  3. moderate or severe intensity
- D. During abdominal pain at least 2 of the following:
  1. anorexia

2. nausea
  3. vomiting
  4. pallor
- E. 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 is severe enough to interfere with normal daily activities.

Children may find it difficult to distinguish anorexia from nausea. The 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.

### **1.3.3 Benign paroxysmal vertigo of childhood**

*Description:*

This probably heterogeneous disorder is characterised by recurrent brief episodic attacks of vertigo occurring without warning and resolving spontaneously in otherwise healthy children.

*Diagnostic criteria:*

- A. At least 5 attacks fulfilling criterion B
- B. Multiple episodes of severe vertigo<sup>1</sup>, occurring without warning and resolving spontaneously after minutes to hours
- C. Normal neurological examination; audiometric and vestibular functions between attacks
- D. Normal electroencephalogram

*Note:*

1. Often associated with nystagmus or vomiting; unilateral throbbing headache may occur in some attacks.

## **1.4 Retinal migraine**

*Description:*

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

*Diagnostic criteria:*

- A. At least 2 attacks fulfilling criteria B and C
- B. Fully reversible monocular positive and/or negative visual phenomena (*eg*, scintillations, scotomata or blindness) confirmed by examination during an attack or (after proper instruction) by the patient's drawing of a monocular field defect during an attack

- C. Headache fulfilling criteria B-D for 1.1 *Migraine without aura* begins during the visual symptoms or follows them within 60 minutes
- D. Normal ophthalmological examination between attacks
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

1. Appropriate investigations exclude other causes of transient monocular blindness.

*Comment:*

Some patients who complain of monocular visual disturbance in fact have hemianopia. Some cases without headache have been reported, but their migrainous nature cannot be ascertained. Other causes of transient monocular blindness (*amaurosis fugax*), such as optic neuropathy or carotid dissection, must be excluded.

## **1.5 Complications of migraine**

*Comment:*

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

### **1.5.1 Chronic migraine**

*Description:*

Migraine headache occurring on 15 or more days per month for more than 3 months in the absence of medication overuse.

*Diagnostic criteria:*

- A. Headache fulfilling criteria C and D for 1.1 *Migraine without aura* on  $\geq 15$  days/month for  $>3$  months
- B. Not attributed to another disorder<sup>1;2</sup>

*Note:*

1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.
2. When medication overuse is present and fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache*, it is uncertain whether this criterion B is fulfilled until 2 months after medication has been withdrawn without improvement (see *Comments*).

*Comments:*

Most cases of chronic migraine start as 1.1 *Migraine without aura*. Therefore, chronicity may be regarded as a complication of episodic migraine.

As chronicity develops, headache tends to lose its attack-wise (episodic) presentation although it has not been clearly demonstrated that this is always so.

When medication overuse is present (*ie*, fulfilling criterion B for any of the subforms of 8.2 *Medication-overuse headache*), this is the most likely cause of chronic symptoms. Therefore, the default rule is to code such patients according to the antecedent migraine subtype (usually 1.1 *Migraine without aura*) plus 1.6.5 *Probable chronic migraine* plus 8.2.8 *Probable medication-overuse headache*. When these criteria are still fulfilled 2 months after medication overuse has ceased, 1.5.1 *Chronic migraine* plus the antecedent migraine subtype should be diagnosed, and 8.2.8 *Probable medication-overuse headache* discarded. If at any time sooner they are no longer fulfilled, because improvement has occurred, code for 8.2 *Medication-overuse headache* plus the antecedent migraine subtype and discard 1.6.5 *Probable chronic migraine*.

These criteria require further study.

### **1.5.2 Status migrainosus**

#### *Description:*

A debilitating migraine attack lasting for more than 72 hours.

#### *Diagnostic criteria:*

- A. The present attack in a patient with 1.1 *Migraine without aura* is typical of previous attacks except for its duration
- B. Headache has both of the following features:
  - 1 unremitting for >72 hours
  - 2 severe intensity
- C. Not attributed to another disorder

#### *Comment:*

Interruption during sleep is disregarded. Short-lasting relief due to medication is also disregarded. Status may often be caused by medication overuse and should be coded accordingly. Non-debilitating attacks lasting >72 hours but otherwise meeting these criteria are coded as 1.6.1 *Probable migraine without aura*.

### **1.5.3 Persistent aura without infarction**

#### *Description:*

Aura symptoms persist for more than 1 week without radiographic evidence of infarction.

#### *Diagnostic criteria:*

- A. The present attack in a patient with 1.2 *Migraine with aura* is typical of previous attacks except that one or more aura symptoms persists for >1 week
- B. Not attributed to another disorder

#### *Comments:*

Persisting aura symptoms are rare but well documented. They are often bilateral and may last for months or years. Reliably effective treatment is not known though acetazolamide and valproic acid have helped in a few cases.

Exclude posterior leukoencephalopathy by diffusion MRI among other things. Exclude 1.5.4 *Migrainous infarction* by MRI.

#### **1.5.4 Migrainous infarction**

##### *Description:*

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

##### *Diagnostic criteria:*

- A. The present attack in a patient with 1.2 *Migraine with aura* is typical of previous attacks except that one or more aura symptoms persists for >60 minutes
- B. Neuroimaging demonstrates ischaemic infarction in a relevant area
- C. Not attributed to another disorder

##### *Comments:*

Ischaemic stroke in a migraine sufferer may be categorised as cerebral infarction of other cause coexisting with migraine, cerebral infarction of other cause presenting with symptoms resembling migraine with aura, or cerebral infarction occurring during the course of a typical migraine with aura attack. Only the last fulfils criteria for 1.5.4 *Migrainous infarction*.

Increased risk for stroke in migraine patients has been demonstrated in women under age 45 in several studies. Evidence for an association between migraine and stroke in older women and in men is inconsistent.

#### **1.5.5 Migraine-triggered seizure**

##### *Description:*

A seizure triggered by a migraine aura.

##### *Diagnostic criteria:*

- A. Migraine fulfilling criteria for 1.2 *Migraine with aura*
- B. A seizure fulfilling diagnostic criteria for one type of epileptic attack occurs during or within 1 hour after a migraine aura

##### *Comment:*

Migraine and epilepsy are prototypical examples of paroxysmal brain disorders. While migraine-like headaches are quite frequently seen in the postictal period, sometimes a seizure occurs during or following a migraine attack. This phenomenon, sometimes referred to as *migralepsy*, has been described in patients with migraine with aura.

### **1.6 Probable migraine**

##### *Previously used terms:*

Migrainous disorder

*Coded elsewhere:*

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

*Description:*

Attacks and/or headache missing one of the features needed to fulfil all criteria for a disorder coded above (1.6.3 *Probable childhood periodic syndromes that are commonly precursors of migraine* and 1.6.4 *Probable retinal migraine* are not currently recognised).

### **1.6.1 Probable migraine without aura**

*Diagnostic criteria:*

- A. Attacks fulfilling all but one of criteria A-D for 1.1 *Migraine without aura*
- B. Not attributed to another disorder

*Comment:*

Do not code as 1.6.1 *Probable migraine without aura* if the patient fulfils the criteria for 1.5.1 *Chronic migraine* or 1.5.2 *Status migrainosus*.

### **1.6.2 Probable migraine with aura**

*Diagnostic criteria:*

- A. Attacks fulfilling all but one of criteria A-D for 1.2 *Migraine with aura* or any of its subforms
- B. Not attributed to another disorder

### **1.6.5 Probable chronic migraine**

*Diagnostic criteria:*

- A. Headache fulfilling criteria C and D for 1.1 *Migraine without aura* on  $\geq 15$  days/month for  $>3$  months
- B. Not attributed to another disorder<sup>1</sup> but there is, or has been within the last 2 months, medication overuse fulfilling criterion B for any of the subforms of 8.2 *Medication-overuse headache*

*Note:*

1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12 (other than 8.2 *Medication-overuse headache*), or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.

## Aggravating factors

Migraine may be aggravated by a number of factors. That is, in a person who already meets criteria for migraine, particular factors may be associated with a relatively long-term (usually weeks to months) increase in the severity or frequency of attacks. Examples of commonly-reported aggravating factors include: psychosocial stress, frequent intake of alcoholic beverages, other environmental factors.

## Trigger factors (precipitating factors)

Trigger factors increase the probability of a migraine attack in the short term (usually <48 hours) in a person with migraine. Though some trigger factors have been reasonably well studied epidemiologically (*eg*, menstruation) or in clinical trials (*eg*, chocolate, aspartame), causal attribution in individual patients may be difficult.

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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 and psychogenic headache

### *Coded elsewhere:*

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

## **General comment**

### *Primary or secondary headache or both?*

When a headache with tension-type characteristics occurs for the first time in close temporal relation to another disorder that is a known cause of headache, it is coded according to the causative disorder as a secondary headache. When pre-existing tension-type headache is made worse in close temporal relation to another disorder that is a known cause of headache, there are two possibilities, and judgment is required. The patient can either be given only the tension-type headache diagnosis or be given both the tension-type headache diagnosis and a secondary headache diagnosis according to the other disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the disorder, a marked worsening of the tension-type headache, very good evidence that the disorder can cause or aggravate tension-type headache and, finally, improvement or resolution of tension-type headache after relief from the disorder.

## **Introduction**

This is the most common type of primary headache: its lifetime prevalence in the general population ranges in different studies from 30 to 78%. At the same time, it is the least studied of the primary headache disorders, despite the fact that it has the highest socio-economic impact.

Whilst this type of headache was previously considered to be primarily psychogenic, a number of studies have appeared after the first edition of *The International Classification of Headache*

*Disorders* that strongly suggest a neurobiological basis, at least for the more severe subtypes of tension-type headache.

The division into *episodic* and *chronic* subtypes that was introduced in the first edition of the classification has proved extremely useful. The chronic subtype is a serious disease causing greatly decreased quality of life and high disability. In the present edition we have decided to subdivide episodic tension-type headache further, into an *infrequent* subtype with headache episodes less than once per month and a *frequent* subtype. The infrequent subtype has very little impact on the individual and does not deserve much attention from the medical profession. However, frequent sufferers can encounter considerable disability that sometimes warrants expensive drugs and prophylactic medication. The chronic subtype is of course always associated with disability and high personal and socio-economic costs.

The first edition arbitrarily separated patients with and without disorder of the pericranial muscles. This has proved to be a valid subdivision but the only really useful distinguishing feature is tenderness on manual palpation and not, as suggested in the first edition, evidence from surface EMG or pressure algometry. Therefore, we now use only manual palpation, preferably as pressure-controlled palpation, to subdivide all three subtypes of tension-type headache.

The exact mechanisms of 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*. The classification subcommittee encourages further research into the pathophysiological mechanisms and treatment of tension-type headache.

There are some reasons to believe that, with the diagnostic criteria set out in the first edition, patients coded for episodic tension-type headache included some who had a mild form of migraine without aura and patients coded for chronic tension-type headache included some who had chronic migraine. Clinical experience favours this suspicion, especially in patients who also have migraine attacks, and some patients may display pathophysiological features typical of migraine (Schoenen *et al*, 1987). Within the classification subcommittee there was an attempt to tighten the diagnostic criteria for tension-type headache for the second edition, with the hope to exclude migraine patients whose headache phenotypically resembles tension-type headache. However, this would have compromised the sensitivity of the criteria and there was no evidence to show the beneficial effects of such a change. Therefore a consensus was not reached, but a proposal for new, stricter diagnostic criteria is published under A2 *Tension-type headache* in the appendix. The classification subcommittee recommends comparisons between patients diagnosed according to the explicit criteria and others diagnosed according to the appendix criteria. This pertains not only to the clinical features but also to pathophysiological mechanisms and response to treatments.

## **2.1 Infrequent episodic tension-type headache**

### *Description:*

Infrequent episodes of headache lasting minutes to days. The pain is typically bilateral, pressing or tightening in quality and of mild to moderate intensity, and it does not worsen with routine physical activity. There is no nausea but photophobia or phonophobia may be present.

### *Diagnostic criteria:*

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

- B. Headache lasting from 30 minutes to 7 days
- C. Headache has at least two of the following 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. Both of the following:
  - 1. no nausea or vomiting (anorexia may occur)
  - 2. no more than one of photophobia or phonophobia
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

- 1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.

### ***2.1.1 Infrequent episodic tension-type headache associated with pericranial tenderness***

*Diagnostic criteria:*

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

*Comments:*

Increased pericranial tenderness recorded by manual palpation is the most significant abnormal finding in patients with tension-type headache. The tenderness increases with the intensity and frequency of headache and is further increased during actual headache. The diagnostic value of EMG and pressure algometry is limited and these recordings are therefore omitted from the second edition. Pericranial tenderness is easily recorded by manual palpation by small rotating movements and a firm pressure (preferably aided by use of a palpometer) with the second and third finger on the frontal, temporal, masseter, pterygoid, sternocleidomastoid, splenius and trapezius muscles. A local tenderness score from 0-3 on each muscle can be summated to yield a total tenderness score for each individual. It has been demonstrated that, using a pressure sensitive device that allows palpation with a controlled pressure, this clinical examination becomes more valid and reproducible. However, such equipment is not generally available to clinicians and it is advised that clinicians simply perform the manual palpation as a traditional clinical examination.

Palpation is a useful guide for the treatment strategy. It also adds value and credibility to the explanations given to the patient.

## 2.2 Frequent episodic tension-type headache

### *Description:*

Frequent episodes of headache lasting minutes to days. The pain is typically bilateral, pressing or tightening in quality and of mild to moderate intensity, and it does not worsen with routine physical activity. There is no nausea but photophobia or phonophobia may be present.

### *Diagnostic criteria:*

- A. At least 10 episodes occurring on  $\geq 1$  but  $< 15$  days per month for at least 3 months ( $\geq 12$  and  $< 180$  days per year) and fulfilling criteria B-D
- B. Headache lasting from 30 minutes to 7 days
- C. Headache has at least two of the following 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. Both of the following:
  - 1. no nausea or vomiting (anorexia may occur)
  - 2. no more than one of photophobia or phonophobia
- E. Not attributed to another disorder<sup>1</sup>

### *Note:*

- 1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.

### *Comment:*

Frequent tension-type headache often coexists with migraine without aura. Coexisting tension-type headache in migraineurs should preferably be identified by a diagnostic headache diary. The treatment of migraine differs considerably from that of tension-type headache and it is important to educate patients to differentiate between these types of headaches in order to select the right treatment and to prevent medication-overuse headache.

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

#### *Diagnostic criteria:*

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

## 2.3 Chronic tension-type headache

*Coded elsewhere:*

### 4.8 New daily-persistent headache

*Description:*

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

*Diagnostic criteria:*

- A. Headache occurring on  $\geq 15$  days per month on average for  $>3$  months ( $\geq 180$  days per year)<sup>1</sup> and fulfilling criteria B-D
- B. Headache lasts hours or may be continuous
- C. Headache has at least two of the following 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. 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 attributed to another disorder<sup>2;3</sup>

*Notes:*

- 1 2.3 *Chronic tension-type headache* evolves over time from episodic tension-type headache; when these criteria A-E are fulfilled by headache that, unambiguously, is daily and unremitting within 3 days of its first onset, code as 4.8 *New daily-persistent headache*. When the manner of onset is not remembered or is otherwise uncertain, code as 2.3 *Chronic tension-type headache*.
- 2 History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.
- 3. When medication overuse is present and fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache*, it is uncertain whether this criterion E is fulfilled until 2 months after medication has been withdrawn without improvement (see *Comments*).

*Comments:*

The introduction of 1.5.1 *Chronic migraine* into *The International Classification of Headache Disorders, 2<sup>nd</sup> edition*, creates a problem in relation to the differential diagnosis between this and 2.3 *Chronic tension-type headache*. Both diagnoses require headache (meeting the criteria for migraine or tension-type headache respectively) on at least 15 days a month. Therefore it is possible theoretically that a patient can have both these diagnoses. A very small group of patients have 15 or more headaches per month fulfilling the diagnostic criteria for both 1.5.1 *Chronic migraine* and 2.3 *Chronic tension-type headache*. This is possible when two (and only two) of the four pain

characteristics are present and headaches are associated with mild nausea. In these rare cases, other clinical evidence that is not part of the explicit diagnostic criteria should be taken into account and the clinician should base thereon the best possible choice of diagnosis. When it is uncertain how many attacks fulfil one or other set of criteria it is strongly recommended to use a diagnostic headache diary prospectively.

In many uncertain cases there is overuse of medication. When this fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache*, the default rule is to code for 2.4.3 *Probable chronic tension-type headache* plus 8.2.8 *Probable medication-overuse headache*. When these criteria are still fulfilled 2 months after medication overuse has ceased, 2.3 *Chronic tension-type headache* should be diagnosed and 8.2.8 *Probable medication-overuse headache* discarded. If at any time sooner they are no longer fulfilled, because improvement has occurred, 8.2 *Medication-overuse headache* should be diagnosed and 2.4.3 *Probable chronic tension-type headache* discarded.

It should be remembered that some patients with chronic tension-type headache develop migraine-like features if they have severe pain and , conversely, some migraine patients develop increasingly frequent tension-type-like interval headaches, the nature of which remains unclear.

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

*Diagnostic criteria:*

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

## **2.4 *Probable tension-type headache***

*Comment:*

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

### **2.4.1 *Probable infrequent episodic tension-type headache***

*Diagnostic criteria:*

- A. Episodes fulfilling all but one of criteria A-D for 2.1 *Infrequent episodic tension-type headache*
- B. Episodes do not fulfil criteria for 1.1 *Migraine without aura*
- C. Not attributed to another disorder

#### 2.4.2 Probable frequent episodic tension-type headache

*Diagnostic criteria:*

- A. Episodes fulfilling all but one of criteria A-D for 2.2 *Frequent episodic tension-type headache*
- B. Episodes do not fulfil criteria for 1.1 *Migraine without aura*
- C. Not attributed to another disorder

#### 2.4.3 Probable chronic tension-type headache

*Diagnostic criteria:*

- A. Headache occurring on  $\geq 15$  days per month on average for  $>3$  months ( $\geq 180$  days per year) and fulfilling criteria B-D
- B. Headache lasts hours or may be continuous
- C. Headache has at least two of the following 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. 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 attributed to another disorder but there is, or has been within the last 2 months, medication overuse fulfilling criterion B for any of the subforms of 8.2 *Medication-overuse headache*

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### **3. CLUSTER HEADACHE AND OTHER TRIGEMINAL AUTONOMIC CEPHALALGIAS**

- 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 (CPH)
- 3.3 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
- 3.4 Probable trigeminal autonomic cephalalgia
  - 3.4.1 Probable cluster headache
  - 3.4.2 Probable paroxysmal hemicrania
  - 3.4.3 Probable SUNCT

*Coded elsewhere:*

4.7 *Hemicrania continua*

#### **General comment**

*Primary or secondary headache or both?*

When a headache with the characteristics of a trigeminal autonomic cephalalgia (TAC) occurs for the first time in close temporal relation to another disorder that is a known cause of headache, it is coded according to the causative disorder as a secondary headache. When a pre-existing TAC is made worse in close temporal relation to another disorder that is a known cause of headache, there are two possibilities, and judgment is required. The patient can either be given only the TAC diagnosis or be given both the TAC diagnosis and a secondary headache diagnosis according to the other disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the disorder, a marked worsening of the TAC, very good evidence that the disorder can cause or aggravate the TAC and, finally, improvement or resolution of the TAC after relief from the disorder.

#### **Introduction**

The trigeminal autonomic cephalalgias share the clinical features of headache and prominent cranial parasympathetic autonomic features. Experimental and human functional imaging suggests that these syndromes activate a normal human trigeminal-parasympathetic reflex with clinical signs of cranial sympathetic dysfunction being secondary.

*Hemicrania continua*, whose cranial autonomic features are less constant, is to be found under 4. *Other primary headaches.*

#### **3.1 Cluster headache**

*Previously used terms:*

Ciliary neuralgia, erythro-melalgia of the head, erythroprosopalgia of Bing, hemicrania angioparalytica, hemicrania neuralgiformis chronica, histaminic cephalalgia, Horton's headache,

Harris-Horton's disease, migrainous neuralgia (of Harris), petrosal neuralgia (of Gardner), Sluder's neuralgia, sphenopalatine neuralgia, vidian neuralgia

*Coded elsewhere:*

Symptomatic cluster headache is coded to the underlying causative 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 8 times a day. The attacks are associated with one or more of the following, all of which are ipsilateral: conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis, eyelid oedema. Most patients are restless or agitated during an attack.

*Diagnostic criteria:*

- A. At least 5 attacks fulfilling criteria B-D
- B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes if untreated<sup>1</sup>
- C. Headache is accompanied by at least one of the following:
  - 1. ipsilateral conjunctival injection and/or lacrimation
  - 2. ipsilateral nasal congestion and/or rhinorrhoea
  - 3. ipsilateral eyelid oedema
  - 4. ipsilateral forehead and facial sweating
  - 5. ipsilateral miosis and/or ptosis
  - 6. a sense of restlessness or agitation
- D. Attacks have a frequency from one every other day to 8 per day<sup>2</sup>
- E. Not attributed to another disorder<sup>3</sup>

*Notes:*

- 1. During part (but less than half) of the time-course of cluster headache, attacks may be less severe and/or of shorter or longer duration.
- 2. During part (but less than half) of the time-course of cluster headache, attacks may be less frequent.
- 3. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

*Comments:*

Acute attacks involve activation of the posterior hypothalamic grey matter. Cluster headache may be inherited (autosomal dominant) in about 5% of cases.

Attacks usually occur in series (*cluster periods*) lasting for weeks or months separated by remission periods usually lasting months or years. However, about 10-15% of patients have chronic symptoms without remissions.

In a large series with good follow-up, 27% of patients had only a single cluster period. These should be coded as 3.1 *Cluster headache*.

During a cluster period, and in the chronic subtype, attacks occur regularly and may be provoked by alcohol, histamine or nitroglycerine. Pain is maximal orbitally, supraorbitally, temporally or in any combination of these sites, but may spread to other regions of the head. Pain almost invariably recurs on the same side during an individual cluster period. During the worst attacks, the intensity of pain is excruciating. Patients are usually unable to lie down and characteristically pace the floor.

Age at onset is usually 20-40 years. For unknown reasons prevalence is 3-4 times higher in men than in women.

*Cluster headache with coexistent trigeminal neuralgia (cluster-tic syndrome):*

Some patients have been described who have both 3.1 *Cluster headache* and 13.1 *Trigeminal neuralgia*. They should receive both diagnoses. The importance of this observation is that both conditions must be treated for the patient to be headache free.

### **3.1.1 Episodic cluster headache**

*Description:*

Cluster headache attacks occurring in periods lasting 7 days to 1 year separated by pain-free periods lasting 1 month or longer.

*Diagnostic criteria:*

- A. Attacks fulfilling criteria A-E for 3.1 *Cluster headache*
- B. At least two cluster periods lasting 7-365 days<sup>1</sup> and separated by pain-free remission periods of  $\geq 1$  month

*Note:*

1. Cluster periods usually last between 2 weeks and 3 months.

*Comment:*

The duration of the remission period has been increased in this second edition to a minimum of 1 month.

### **3.1.2 Chronic cluster headache**

*Description:*

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

*Diagnostic criteria:*

- A. Attacks fulfilling criteria A-E for 3.1 *Cluster headache*
- B. Attacks recur over  $>1$  year without remission periods or with remission periods lasting  $<1$  month

*Comments:*

Chronic cluster headache may arise *de novo* (previously referred to as *primary chronic cluster headache*) or evolve from the episodic subtype (previously referred to as *secondary chronic cluster headache*). Some patients may switch from chronic to episodic cluster headache.

### 3.2 Paroxysmal hemicrania

#### *Description:*

Attacks with similar characteristics of pain and associated symptoms and signs to those of cluster headache, but they are shorter-lasting, more frequent, occur more commonly in females and respond absolutely to indomethacin.

#### *Diagnostic criteria:*

- A. At least 20 attacks fulfilling criteria B-D
- B. Attacks of severe unilateral orbital, supraorbital or temporal pain lasting 2-30 minutes
- C. Headache is accompanied by at least one of the following:
  - 1. ipsilateral conjunctival injection and/or lacrimation
  - 2. ipsilateral nasal congestion and/or rhinorrhoea
  - 3. ipsilateral eyelid oedema
  - 4. ipsilateral forehead and facial sweating
  - 5. ipsilateral miosis and/or ptosis
- D. Attacks have a frequency above 5 per day for more than half of the time, although periods with lower frequency may occur
- E. Attacks are prevented completely by therapeutic doses of indomethacin<sup>1</sup>
- F. Not attributed to another disorder<sup>2</sup>

#### *Notes:*

- 1. In order to rule out incomplete response, indomethacin should be used in a dose of  $\geq 150$  mg daily orally or rectally, or  $\geq 100$  mg by injection, but for maintenance smaller doses are often sufficient.
- 2. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

#### *Comments:*

There is no male predominance. Onset is usually in adulthood, although childhood cases are reported.

In the first edition all paroxysmal hemicranias were referred to as *chronic paroxysmal hemicrania*. Sufficient clinical evidence for the episodic subtype has accumulated to separate it in a manner analogous to cluster headache.

#### *Paroxysmal hemicrania with coexistent trigeminal neuralgia (CPH-tic syndrome):*

Patients who fulfil criteria for both 3.2 *Paroxysmal hemicrania* and 13.1 *Trigeminal neuralgia* should receive both diagnoses. The importance of this observation is that both conditions require treatment. The pathophysiological significance of the association is not yet clear.

### **3.2.1 Episodic paroxysmal hemicrania**

*Description:*

Attacks of paroxysmal hemicrania occurring in periods lasting 7 days to 1 year separated by pain-free periods lasting  $\geq 1$  month.

*Diagnostic criteria:*

- A. Attacks fulfilling criteria A-F for 3.2 *Paroxysmal hemicrania*
- B. At least two attack periods lasting 7-365 days and separated by pain-free remission periods of  $\geq 1$  month

### **3.2.2 Chronic paroxysmal hemicrania (CPH)**

*Description:*

Attacks of paroxysmal hemicrania occurring for  $>1$  year without remission or with remissions lasting  $<1$  month.

*Diagnostic criteria:*

- A. Attacks fulfilling criteria A-F for 3.2 *Paroxysmal hemicrania*
- B. Attacks recur over  $>1$  year without remission periods or with remission periods lasting  $<1$  month

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

*Description:*

This syndrome is characterised by short-lasting attacks of unilateral pain that are much briefer than those seen in any other TAC and very often accompanied by prominent lacrimation and redness of the ipsilateral eye.

*Diagnostic criteria:*

- A. At least 20 attacks fulfilling criteria B-D
- B. Attacks of unilateral orbital, supraorbital or temporal stabbing or pulsating pain lasting 5-240 seconds
- C. Pain is accompanied by ipsilateral conjunctival injection and lacrimation
- D. Attacks occur with a frequency from 3 to 200 per day
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

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

*Comments:*

This syndrome was described after the publication of the first edition of *The International Classification of Headache Disorders* and has become well recognised in the last decade.

Patients may be seen with only one of conjunctival injection or tearing, or other cranial autonomic symptoms such as nasal congestion, rhinorrhoea or eyelid oedema may be seen. 3.3 *SUNCT* may be a subform of A3.3 *Short-lasting Unilateral Neuralgiform headache attacks with cranial Autonomic symptoms* (SUNA), described in the appendix.

The literature suggests that the most common mimics of 3.3 *SUNCT* are lesions in the posterior fossa or involving the pituitary gland.

*SUNCT with coexistent trigeminal neuralgia:* Patients have been described in whom there is an overlap between 3.3 *SUNCT* and 13.1 *Trigeminal neuralgia*. Such patients should receive both diagnoses. This differentiation is clinically difficult.

### **3.4 Probable trigeminal autonomic cephalalgia**

*Description:*

Headache attacks that are believed to be a subtype of trigeminal autonomic cephalalgia but which do not quite meet the diagnostic criteria for any of the subtypes described above.

*Diagnostic criteria:*

- A. Attacks fulfilling all but one of the specific criteria for one of the subtypes of trigeminal autonomic cephalalgia
- B. Not attributed to another disorder

*Comment:*

Patients coded as 3.4 *Probable trigeminal autonomic cephalalgia* or one of its subforms either have had an insufficient number of typical attacks or fail to fulfil one of the other criteria.

#### **3.4.1 Probable cluster headache**

*Diagnostic criteria:*

- A. Attacks fulfilling all but one of criteria A-D for 3.1 *Cluster headache*
- B. Not attributed to another disorder

#### **3.4.2 Probable paroxysmal hemicrania**

*Diagnostic criteria:*

- A. Attacks fulfilling all but one of criteria A-E for 3.2 *Paroxysmal hemicrania*
- B. Not attributed to another disorder

### 3.4.3 *Probable short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing*

#### *Diagnostic criteria:*

- A. Attacks fulfilling all but one of criteria A-D for 3.3 *Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)*
- B. Not attributed to another disorder

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## 4. OTHER PRIMARY HEADACHES

- 4.1 Primary stabbing headache
- 4.2 Primary cough headache
- 4.3 Primary exertional headache
- 4.4 Primary headache associated with sexual activity
  - 4.4.1 Preorgasmic headache
  - 4.4.2 Orgasmic headache
- 4.5 Hypnic headache
- 4.6 Primary thunderclap headache
- 4.7 Hemicrania continua
- 4.8 New daily-persistent headache (NDPH)

### General comment

#### *Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to another disorder that is a known cause of headache, this headache is coded according to the causative disorder as a secondary headache. This is also true if the headache has the characteristics of migraine or other primary headache. When a pre-existing primary headache is made worse in close temporal relation to another disorder that is a known cause of headache, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and a secondary headache diagnosis according to the other disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the disorder, a marked worsening of the pre-existing headache, very good evidence that the disorder can cause or aggravate the primary headache and, finally, improvement or resolution of the primary headache after relief from the disorder.

### Introduction

This chapter includes headaches that are clinically heterogeneous. The pathogenesis of these types of headache is still poorly understood, and their treatment is suggested on the basis of anecdotal reports or uncontrolled trials.

Several headache disorders included in this chapter can be symptomatic and need careful evaluation by imaging and/or other appropriate tests.

The onset of some of these headaches, 4.6 *Primary thunderclap headache* especially, can be acute and affected patients are usually assessed in Emergency Departments. Appropriate and full investigation (neuroimaging, in particular) is mandatory in these cases.

The chapter also includes some clinical entities, such as 4.1 *Primary stabbing headache* and 4.5 *Hypnic headache* (this latter recently described), that are primary in most cases.

### 4.1 Primary stabbing headache

#### *Previously used terms:*

Ice-pick pains, jabs and jolts, ophthalmodynia periodica

*Description:*

Transient and localised 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 as a single stab or a series of stabs and fulfilling criteria B-D
- B. Exclusively or predominantly felt in the distribution of the first division of the trigeminal nerve (orbit, temple and parietal area)
- C. Stabs last for up to a few seconds and recur with irregular frequency ranging from one to many per day
- D. No accompanying symptoms
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but pain does not occur for the first time in close temporal relation to the disorder.

*Comments:*

In a single published descriptive study, 80% of stabs lasted 3 seconds or less. In rare cases, stabs occur repetitively over days, and there has been one description of *status* lasting one week.

Stabs may move from one area to another in either the same or the opposite hemicranium. When they are strictly localised to one area, structural changes at this site and in the distribution of the affected cranial nerve must be excluded.

Stabbing pains are more commonly experienced by people subject to migraine (about 40%) or cluster headache (about 30%), in which cases they are felt in the site habitually affected by these headaches.

A positive response to indomethacin has been reported in some uncontrolled studies, whilst others have observed partial or no responses.

## **4.2 Primary cough headache**

*Previously used terms:*

Benign cough headache, Valsalva-manoeuvre headache

*Description:*

Headache precipitated by coughing or straining in the absence of any intracranial disorder.

*Diagnostic criteria:*

- A. Headache fulfilling criteria B and C
- B. Sudden onset, lasting from one second to 30 minutes
- C. Brought on by and occurring only in association with coughing, straining and/or Valsalva manoeuvre
- D. Not attributed to another disorder<sup>1</sup>

*Note:*

1. Cough headache is symptomatic in about 40% of cases and the large majority of these present Arnold-Chiari malformation type I. Other reported causes of symptomatic cough headache include carotid or vertebrobasilar diseases and cerebral aneurysms. Diagnostic neuroimaging plays an important role in differentiating secondary cough headache from 4.2 *Primary cough headache*.

*Comment:*

Primary cough headache is usually bilateral and predominantly affects patients older than 40 years of age. Whilst indomethacin is usually effective in the treatment of primary cough headache, a positive response to this medication has also been reported in some symptomatic cases.

### **4.3 Primary exertional headache**

*Previously used terms:*

Benign exertional headache

*Coded elsewhere:*

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

*Description:*

Headache precipitated by any form of exercise. Subforms such as “weight-lifters’ headache” are recognised.

*Diagnostic criteria:*

- A. Pulsating headache fulfilling criteria B and C
- B. Lasting from 5 minutes to 48 hours
- C. Brought on by and occurring only during or after physical exertion
- D. Not attributed to another disorder<sup>1</sup>

*Note:*

1. On first occurrence of this headache type it is mandatory to exclude subarachnoid haemorrhage and arterial dissection.

*Comments:*

Primary exertional headache occurs particularly in hot weather or at high altitude. There are reports of prevention in some patients by the ingestion of ergotamine tartrate. Indomethacin has been found effective in the majority of the cases.

Headache described in weight-lifters has been considered a subform of 4.3 *Primary exertional headache*; because of its sudden onset and presumed mechanism it may have more similarities to 4.2 *Primary cough headache*.

## 4.4 Primary headache associated with sexual activity

*Previously used terms:*

Benign sex headache, coital cephalalgia, benign vascular sexual headache, sexual headache

*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.

### 4.4.1 Preorgasmic headache

*Diagnostic criteria:*

- A. Dull ache in the head and neck associated with awareness of neck and/or jaw muscle contraction and meeting criterion B
- B. Occurs during sexual activity and increases with sexual excitement
- C. Not attributed to another disorder

### 4.4.2 Orgasmic headache

*Coded elsewhere:*

Postural headache resembling that of low CSF pressure has been reported to develop after coitus. Such headache should be coded as 7.2.3 *Headache attributed to spontaneous (or idiopathic) low CSF pressure* because it is due to CSF leakage.

*Diagnostic criteria:*

- A. Sudden severe (“explosive”) headache meeting criterion B
- B. Occurs at orgasm
- C. Not attributed to another disorder<sup>1</sup>

*Note:*

1. On first onset of orgasmic headache it is mandatory to exclude conditions such as subarachnoid haemorrhage and arterial dissection.

*Comments:*

An association between 4.4 *Primary headache associated with sexual activity*, 4.3 *Primary exertional headache* and migraine is reported in approximately 50% of cases.

Two subtypes (*dull type* and *explosive type headache*) were included in the first edition of *The International Classification of Headache Disorders*. No specific investigation has been undertaken since then to clarify whether they are separate entities. In most published reports of headache with sexual activity, only explosive (“vascular type”) headache has been reported. The dull type may be a subtype of tension-type headache, but no evidence supports this hypothesis.

No firm data are available on the duration of primary headache associated with sexual activity, but it is usually considered to last from 1 minute to 3 hours.

## 4.5 Hypnic headache

*Previously used terms:*

Hypnic headache syndrome, “alarm clock” headache

*Description:*

Attacks of dull headache that always awaken the patient from asleep.

*Diagnostic criteria:*

- A. Dull headache fulfilling criteria B-D
- B. Develops only during sleep, and awakens patient
- C. At least two of the following characteristics:
  - 1. occurs >15 times per month
  - 2. lasts ≥15 minutes after waking
  - 3. first occurs after age of 50 years
- D. No autonomic symptoms and no more than one of nausea, photophobia or phonophobia
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

- 1. Intracranial disorders must be excluded. Distinction from one of the trigeminal autonomic cephalalgias is necessary for effective management.

*Comments:*

The pain of hypnic headache is usually mild to moderate, but severe pain is reported by approximately 20% of patients. Pain is bilateral in about two-thirds of cases. The attack usually lasts from 15 to 180 minutes, but longer durations have been described.

Caffeine and lithium have been effective treatments in several reported cases.

## 4.6 Primary thunderclap headache

*Previously used terms:*

Benign thunderclap headache

*Coded elsewhere:*

4.2 *Primary cough headache*, 4.3 *Primary exertional headache* and 4.4 *Primary headache associated with sexual activity* can all present as thunderclap headache but should be coded as those headache types, not as 4.6 *Primary thunderclap headache*.

*Description:*

High-intensity headache of abrupt onset mimicking that of ruptured cerebral aneurysm.

*Diagnostic criteria:*

- A. Severe head pain fulfilling criteria B and C
- B. Both of the following characteristics:
  - 1. sudden onset, reaching maximum intensity in <1 minute
  - 2. lasting from 1 hour to 10 days

- C. Does not recur regularly over subsequent weeks or months<sup>1</sup>
- D. Not attributed to another disorder<sup>2</sup>

*Notes:*

1. Headache may recur within the first week after onset.
2. Normal CSF and normal brain imaging are required.

*Comment:*

Evidence that thunderclap headache exists as a primary condition is poor: the search for an underlying cause should be expedient and exhaustive. Thunderclap headache is frequently associated with serious vascular intracranial disorders, particularly subarachnoid haemorrhage: it is mandatory to exclude this and a range of other such conditions including intracerebral haemorrhage, cerebral venous thrombosis, unruptured vascular malformation (mostly aneurysm), arterial dissection (intra- and extracranial), CNS angiitis, reversible benign CNS angiopathy and pituitary apoplexy. Other organic causes of thunderclap headache are colloid cyst of the third ventricle, CSF hypotension and acute sinusitis (particularly with barotrauma). 4.6 *Primary thunderclap headache* should be the diagnosis only when all organic causes have been excluded.

#### **4.7 Hemicrania continua**

*Description:*

Persistent strictly unilateral headache responsive to indomethacin.

*Diagnostic criteria:*

- A. Headache for >3 months fulfilling criteria B-D
- B. All of the following characteristics:
  1. unilateral pain without side-shift
  2. daily and continuous, without pain-free periods
  3. moderate intensity, but with exacerbations of severe pain
- C. At least one of the following autonomic features occurs during exacerbations and ipsilateral to the side of pain:
  1. conjunctival injection and/or lacrimation
  2. nasal congestion and/or rhinorrhoea
  3. ptosis and/or miosis
- D. Complete response to therapeutic doses of indomethacin
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.

*Comment:*

Hemicrania continua is usually unremitting, but rare cases of remission are reported. Whether this headache type can be subdivided according to length of history and persistence is yet to be determined.

## 4.8 New daily-persistent headache (NDPH)

*Previously used terms:*

*De novo* chronic headache; chronic headache with acute onset

*Description:*

Headache that is daily and unremitting from very soon after onset (within 3 days at most). The pain is typically bilateral, pressing or tightening in quality and of mild to moderate intensity. There may be photophobia, phonophobia or mild nausea.

*Diagnostic criteria:*

- A. Headache that, within 3 days of onset<sup>1</sup>, fulfils criteria B-D
- B. Headache is present daily, and is unremitting, for >3 months
- C. At least two of the following pain 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
- 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 attributed to another disorder<sup>2</sup>

*Note:*

- 1. Headache may be unremitting from the moment of onset or very rapidly build up to continuous and unremitting pain. Such onset or rapid development must be clearly recalled and unambiguously described by the patient. Otherwise code as 2.3 *Chronic tension-type headache*.
- 2. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12 (including 8.2 *Medication-overuse headache* and its subforms), or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.

*Comments:*

This second edition of the classification recognises 4.8 *New daily-persistent headache* as a separate entity from 2.3 *Chronic tension-type headache*. Although it has many similarities to tension-type headache, NDPH is unique in that headache is daily and unremitting from or almost from the moment of onset, typically in individuals without a prior headache history. A clear recall of such an onset is necessary for the diagnosis of 4.8 *New daily-persistent headache*.

The headache of NDPH can have associated features suggestive of either migraine or tension-type headache. Secondary headaches such as low CSF volume headache, raised CSF pressure headache, post-traumatic headache and headache attributed to infection (particularly viral) should be ruled out by appropriate investigations.

If there is or has been within the last 2 months medication overuse fulfilling criterion B for any of the subforms of 8.2 *Medication-overuse headache*, the rule is to code for any pre-existing primary

headache plus 8.2.8 *Probable medication-overuse headache* but not for 4.8 *New daily-persistent headache*.

NDPH may take either of two subforms: a self-limiting subform which typically resolves without therapy within several months and a refractory subform which is resistant to aggressive treatment programmes. The subcommittee aims to stimulate further clinical characterisation and pathophysiological research of this syndrome, especially studies comparing 4.8 *New daily-persistent headache* with 2.3 *Chronic tension-type headache*.

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# PART TWO

## THE SECONDARY HEADACHES

Headache attributed to head and neck trauma

Headache attributed to cranial or cervical vascular disorder

Headache attributed to non-vascular intracranial disorder and other causes

Headache attributed to substances or their withdrawal

Headache attributed to infection

Headache attributed to disturbance of homeostasis

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

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 neoplasm* – even if the headache phenomenologically is migraine, tension-type headache or cluster headache. In other words, a *de novo* headache occurring with another disorder recognised 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. In the first edition of *The International Classification of Headache Disorders* we concluded after many discussions that only a new headache could be regarded as secondary. During the work with the second edition it has become obvious, however, that this results in some unacceptable situations. What about a patient who throughout her life has had ten migraine attacks but who, after a head trauma, begins to have migraine attacks twice a week and becomes disabled by these headaches? According to the system of the first edition this patient could only receive the diagnosis of migraine. Another example is a patient who has had tension-type headache which becomes worse, whilst retaining the same characteristics, in association with a brain tumour. The diagnosis of 7.4 *Headache attributed to intracranial neoplasm* could not previously be given. Finally, nothing in the past could be diagnosed as medication-overuse headache because this is always an aggravation of a primary headache, usually migraine, which would remain the only diagnosis.

For these reasons, we introduce a new way of diagnosing and coding primary headaches that are made significantly worse in close temporal relation to another disorder known from good scientific studies to be able to cause headache. Such patients can now receive two diagnoses: the primary headache diagnosis and the secondary headache diagnosis. In theory the new system is more open to interpretation than the old but, in fact, the old system has never been followed when it led to unreasonable diagnoses. The problem with the new system is to decide, in patients whose primary headache worsens in relation to another disorder, whether to use only the primary diagnosis or whether to add a secondary headache diagnosis also. The following factors support the use of two diagnoses: a very close temporal relation, marked worsening of the primary headache, the existence of other evidence that the other disorder can aggravate primary headache in the manner observed, and remission of the headache after cure or remission of the other disorder.

In the first edition of *The International Classification of Headache Disorders* the diagnostic criteria for secondary headaches varied a great deal and were often uninformative about headache characteristics. For this second edition it has been decided to standardise the format and give more headache characteristics whenever possible. The diagnostic criteria therefore have the following disposition:

*Diagnostic criteria for secondary headaches:*

- A. Headache with one (or more) of the following [listed] characteristics<sup>1;2</sup> and fulfilling criteria C and D
- B. Another disorder known to be able to cause headache has been demonstrated
- C. Headache occurs in close temporal relation to the other disorder and/or there is other evidence of a causal relationship
- D. Headache is greatly reduced or resolves within 3 months (this may be shorter for some disorders) after successful treatment or spontaneous remission of the causative disorder<sup>3</sup>

*Notes:*

1. For most secondary headaches the characteristics of the headache itself are poorly described in the scientific literature. Even for those where it is well described, there are usually few diagnostically important features. Therefore, diagnostic criterion A in the standard set of criteria is usually not very contributory to establishing causation. However, criteria B, C and D usually effectively establish causation. This makes it possible to use criterion A not only as a defining feature but also to tell as much about the headache as possible or to show how little we know of it. This is why the formulation of criterion A now allows mention of a number of features. Hopefully, this will stimulate more research into the characteristics of secondary headaches so that, eventually, criterion A for most of these headaches can become much more clearly defined.
2. If nothing is known about the headache, it is stated “no typical characteristics known”.
3. Criterion D cannot always be ascertained and some presumed causative disorders cannot be treated or do not remit. In such cases criterion D may be replaced by: “Other causes ruled out by appropriate investigations”.

In many cases sufficient follow-up is not available or a diagnosis has to be made before the expected time needed for remission. In most such cases the headache should be coded as *Headache probably attributed to [the disorder]*: a definite relationship can only be established with full confidence once criterion D is fulfilled. This is especially so in situations where a pre-existing primary headache has been made worse by another disorder. For example, the great majority of patients otherwise fulfilling the criteria for 1.5.1 *Chronic migraine* are overusing medication and will improve after this overuse ceases. The default rule in this case, pending withdrawal of the overused medication, is to code according to the antecedent migraine subtype (usually 1.1 *Migraine without aura*) plus 1.6.5 *Probable chronic migraine* plus 8.2.8 *Probable medication-overuse headache*. Following withdrawal, criterion D for 8.2 *Medication-overuse headache* is not fulfilled if a patient does not improve within 2 months and this diagnosis must then be discarded in favour of 1.5.1 *Chronic migraine*. A similar rule applies to patients overusing medication but otherwise fulfilling the criteria for 2.3 *Chronic tension-type headache*.

In most cases criterion D has a time-limit for improvement of the headache after cure or spontaneous remission or removal of the presumed cause. Usually this is 3 months but it is shorter for some secondary headaches. If headache persists after 3 months (or a shorter limit) it should be questioned whether it was actually secondary to the presumed cause. Secondary headaches persisting after 3 months have often been observed but most have not been of scientifically-proven aetiology. Such cases have been included in the appendix as *Chronic headache attributed to [a specified disorder]*.

## 5. HEADACHE ATTRIBUTED TO HEAD AND/OR NECK TRAUMA

- 5.1 Acute post-traumatic headache
  - 5.1.1 Acute post-traumatic headache attributed to moderate or severe head injury
  - 5.1.2 Acute post-traumatic headache attributed to mild head injury
- 5.2 Chronic post-traumatic headache
  - 5.2.1 Chronic post-traumatic headache attributed to moderate or severe head injury
  - 5.2.2 Chronic post-traumatic headache attributed to mild head injury
- 5.3 Acute headache attributed to whiplash injury
- 5.4 Chronic headache attributed to whiplash injury
- 5.5 Headache attributed to traumatic intracranial haematoma
  - 5.5.1 Headache attributed to epidural haematoma
  - 5.5.2 Headache attributed to subdural haematoma
- 5.6 Headache attributed to other head and/or neck trauma
  - 5.6.1 Acute headache attributed to other head and/or neck trauma
  - 5.6.2 Chronic headache attributed to other head and/or neck trauma
- 5.7 Post-craniotomy headache
  - 5.7.1 Acute post-craniotomy headache
  - 5.7.2 Chronic post-craniotomy headache

### General comment

#### *Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to a known trauma, it is coded as a secondary headache attributed to the trauma. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to a trauma, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the trauma. Factors that support adding the latter diagnosis are: a very close temporal relation to the trauma, a marked worsening of the pre-existing headache, very good evidence that the particular kind of trauma can aggravate the primary headache and, finally, improvement of the headache after recovery from the trauma.

#### *Definite, probable or chronic?*

In many cases of secondary headache, the diagnosis is definite only when headache resolves or greatly improves within a specified time after effective treatment or spontaneous remission of the causative disorder. In such cases this temporal relation is an essential part of the evidence of causation. This is not so in the case of trauma: causation is established by onset in close temporal relation to trauma, whilst it is well recognised that headache after trauma often persists. When this occurs, for example after head trauma, 5.2 *Chronic post-traumatic headache* is diagnosed. Until sufficient time for recovery has elapsed, the diagnosis of 5.1 *Acute post-traumatic headache* is definite if the criteria are fulfilled. The same applies after whiplash injury. There is no option for a diagnosis of *Headache probably attributed to head and/or neck trauma*.

### Introduction

Headache is a symptom that may occur after injury to the head, neck or brain. Frequently, headache that results from head trauma is accompanied by other symptoms such as dizziness, difficulty in

concentration, nervousness, personality changes and insomnia. This constellation of symptoms is known as the post-traumatic syndrome; amongst them, headache is usually the most prominent.

A variety of pain patterns may develop after head injury, and may closely resemble primary headache disorders – most frequently tension-type headache, in more than 80% of patients. In some cases, typical migraine with or without aura may be triggered, and a cluster-like syndrome has been described in a few patients.

It is easy to establish the relationship between a headache and head or neck trauma when the headache develops immediately or in the first days after trauma has occurred. On the other hand it is very difficult when a headache develops weeks or even months after trauma, especially when the majority of these headaches have the pattern of tension-type headache and the prevalence of this type of headache in the population is very high. Such late-onset post-traumatic headaches have been described in anecdotal reports but not in case-control studies.

There are recognised risk factors for a poor outcome after head injury or whiplash injury. Women have a higher risk for post-traumatic headache, and increasing age is associated with less-rapid and less-complete recovery. Mechanical factors such as the position of the head at impact – rotated or inclined – increase the risk of headache after the trauma. The relationship between severity of the injury and severity of the post-traumatic syndrome has not been conclusively established. Although there are some controversial data, most studies suggest that post-traumatic headache is *less* frequent when the head injury is *more* severe. However, the causal relationship between head and/or neck trauma and headache is difficult to establish in some cases with very mild trauma.

The role of litigation in the persistence of headache is still discussed, and some studies show a reduction of headache in countries where the accident victims do not receive compensation. 5.2 *Chronic post-traumatic headache* and 5.4 *Chronic post-whiplash injury headache* are often part of the post-traumatic syndrome in which the complex inter-relationship between organic and psychosocial factors is difficult to assess.

## 5.1 Acute post-traumatic headache

### 5.1.1 Acute post-traumatic headache attributed to moderate or severe head injury

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Head trauma with at least one of the following:
  - 1. loss of consciousness for >30 minutes
  - 2. Glasgow Coma Scale (GCS) <13
  - 3. post-traumatic amnesia for >48 hours
  - 4. imaging demonstration of a traumatic brain lesion (cerebral haematoma, intracerebral and/or subarachnoid haemorrhage, brain contusion and/or skull fracture)
- C. Headache develops within 7 days after head trauma or after regaining consciousness following head trauma
- D. One or other of the following:
  - 1. headache resolves within 3 months after head trauma
  - 2. headache persists but 3 months have not yet passed since head trauma

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

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Head trauma with all the following:
  - 1. either no loss of consciousness, or loss of consciousness of <30 minutes' duration
  - 2. Glasgow Coma Scale (GCS)  $\geq$ 13
  - 3. symptoms and/or signs diagnostic of concussion
- C. Headache develops within 7 days after head trauma
- D: One or other of the following:
  - 1. headache resolves within 3 months after head trauma
  - 2. headache persists but 3 months have not yet passed since head trauma

#### *Comment:*

Mild head injury may give rise to a symptom complex of cognitive, behavioural and consciousness abnormalities and a GCS of  $\geq$ 13. It can occur with or without abnormalities in the neurological examination, neuroimaging (CT scan, MRI), EEG, evoked potentials, CSF examination, vestibular function tests and neuropsychological testing. There is no evidence that an abnormality in any of these changes the prognosis or contributes to treatment. These studies should not be considered routine for patients with ongoing post-traumatic headache. They may be considered on a case-by-case basis, or for research purposes.

## 5.2 Chronic post-traumatic headache

#### *Comment:*

Chronic post-traumatic headache is often part of the post-traumatic syndrome which includes a variety of symptoms such as equilibrium disturbance, poor concentration, decreased work ability, irritability, depressive mood, sleep disturbances, *etc.* The relationship between legal settlements and the temporal profile of chronic post-traumatic headache is not clearly established, but it is important to assess patients carefully who may be malingering and/or seeking enhanced compensation.

### 5.2.1 Chronic post-traumatic headache attributed to moderate or severe head injury

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Head trauma with at least one of the following:
  - 1. loss of consciousness for >30 minutes
  - 2. Glasgow Coma Scale (GCS) <13
  - 3. post-traumatic amnesia for >48 hours
  - 4. imaging demonstration of a traumatic brain lesion (cerebral haematoma, intracerebral and/or subarachnoid haemorrhage, brain contusion and/or skull fracture)
- C. Headache develops within 7 days after head trauma or after regaining consciousness following head trauma
- D. Headache persists for >3 months after head trauma

### 5.2.2 *Chronic post-traumatic headache attributed to mild head injury*

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Head trauma with all the following:
  - 1. either no loss of consciousness, or loss of consciousness of <30 minutes' duration
  - 2. Glasgow Coma Scale (GCS)  $\geq 13$
  - 3. symptoms and/or signs diagnostic of concussion
- C. Headache develops within 7 days after head trauma
- D. Headache persists for >3 months after head trauma

#### *Comment:*

Mild head injury may give rise to a symptom complex of cognitive, behavioural and consciousness abnormalities and a GCS of  $\geq 13$ . It can occur with or without abnormalities in the neurological examination, neuroimaging (CT scan, MRI), EEG, evoked potentials, CSF examination, vestibular function tests and neuropsychological testing. There is no evidence that an abnormality in any of these changes the prognosis or contributes to treatment. These studies should not be considered routine for patients with ongoing post-traumatic headache. They may be considered on a case-by-case basis, or for research purposes.

### 5.3 **Acute headache attributed to whiplash injury**

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. History of whiplash (sudden and significant acceleration/deceleration movement of the neck) associated at the time with neck pain
- C. Headache develops within 7 days after whiplash injury
- D. One or other of the following:
  - 1. headache resolves within 3 months after whiplash injury
  - 2. headache persists but 3 months have not yet passed since whiplash injury

#### *Comments:*

The term *whiplash* commonly refers to a sudden acceleration and/or deceleration of the neck (in the majority of cases due to a road accident). The clinical manifestations include symptoms and signs that relate to the neck, as well as somatic extracervical, neurosensory, behavioural, cognitive and affective disorders whose appearance and modes of expression and evolution can vary widely over time. Headache is very common in this post-whiplash syndrome. The Quebec Task Force on Whiplash-Associated Disorders has proposed a classification in five categories that may be useful in prospective studies.

There are important differences in the incidence of post-whiplash syndrome in different countries, perhaps related to expectations for compensation.

### 5.4 **Chronic headache attributed to whiplash injury**

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D

- B. History of whiplash (sudden and significant acceleration/deceleration movement of the neck) associated at the time with neck pain
- C. Headache develops within 7 days after whiplash injury
- D. Headache persists for >3 months after whiplash injury

*Comment:*

Chronic post-whiplash injury headache is often part of the post-traumatic syndrome. There is no good evidence that ongoing litigation, with settlement pending, is associated with prolongation of headache. It is important to assess patients carefully who may be malingering and/or seeking enhanced compensation.

## **5.5 Headache attributed to traumatic intracranial haematoma**

*Coded elsewhere:*

Headache attributed to traumatic intracerebral and/or subarachnoid haemorrhage or to traumatic intracerebral haematoma is coded as 5.1.1 *Acute post-traumatic headache attributed to moderate or severe head injury* or 5.2.1 *Chronic post-traumatic headache attributed to moderate or severe head injury*.

### **5.5.1 Headache attributed to epidural haematoma**

*Diagnostic criteria:*

- A. Acute-onset headache, no other typical characteristics known, fulfilling criteria C and D
- B. Neuroimaging evidence of epidural haematoma
- C. Headache develops within minutes to 24 hours after development of the haematoma
- D. One or other of the following:
  1. headache resolves within 3 months after evacuation of the haematoma
  2. headache persists but 3 months have not yet passed since evacuation of the haematoma

*Comment:*

Epidural haematoma occurs within hours of head trauma which may be moderate. It is always associated with focal signs and disorders of consciousness. Emergency surgery is required.

### **5.5.2 Headache attributed to subdural haematoma**

*Diagnostic criteria:*

- A. Acute or progressive headache, no other typical characteristics known, fulfilling criteria C and D
- B. Neuroimaging evidence of subdural haematoma
- C. Headache develops within 24-72 hours after development of the haematoma
- D. One or other of the following:
  1. headache resolves within 3 months after evacuation of the haematoma
  2. headache persists but 3 months have not yet passed since evacuation of the haematoma

*Comments:*

Different types of subdural haematomas should be differentiated according to their temporal profile. In acute and subacute haematomas, which usually occur after obvious head trauma, headache is frequent (11-53% of cases) but commonly overshadowed by focal signs and disorders of consciousness. In chronic subdural haematomas, headache is more frequent still (up to 81%) and, though moderate, can be the leading symptom. The diagnosis can be difficult, because the causative head trauma is often trivial and may have been forgotten by the patient. Chronic subdural haematoma should always be considered in an elderly patient with a progressive headache particularly if there is some cognitive impairment and/or mild focal signs.

Bilateral subdural haematomas may be a complication of CSF hypotension. Headache attributed to these is coded here. In such cases, the headache is initially postural and may either remain predominantly postural or become continuous.

## **5.6 Headache attributed to other head and/or neck trauma**

### **5.6.1 Acute headache attributed to other head and/or neck trauma**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Evidence of head and/or neck trauma of a type not described above
- C. Headache develops in close temporal relation to, and/or other evidence exists to establish a causal relationship with, the head and/or neck trauma
- D. One or other of the following:
  - 1. headache resolves within 3 months after the head and/or neck trauma
  - 2. headache persists but 3 months have not yet passed since the head and/or neck trauma

### **5.6.2 Chronic headache attributed to other head and/or neck trauma**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Evidence of head and/or neck trauma of a type not described above
- C. Headache develops in close temporal relation to, and/or other evidence exists to establish a causal relationship with, the head and/or neck trauma
- D. Headache persists for >3 months after the head and/or neck trauma

## **5.7 Post-craniotomy headache**

### **5.7.1 Acute post-craniotomy headache**

*Diagnostic criteria:*

- A. Headache of variable intensity, maximal in the area of the craniotomy, fulfilling criteria C and D
- B. Craniotomy performed for a reason other than head trauma<sup>1</sup>
- C. Headache develops within 7 days after craniotomy

- D. One or other of the following:
1. headache resolves within 3 months after craniotomy
  2. headache persists but 3 months have not yet passed since craniotomy

*Note:*

1. When the craniotomy was for head trauma, code as 5.1.1 *Acute post-traumatic headache attributed to moderate or severe head injury*.

### **5.7.2 Chronic post-craniotomy headache**

*Diagnostic criteria:*

- A. Headache of variable intensity, maximal in the area of the craniotomy, fulfilling criteria C and D
- B. Craniotomy performed for a reason other than head trauma<sup>1</sup>
- C. Headache develops within 7 days after craniotomy
- D. Headache persists for >3 months after craniotomy

*Note:*

1. When the craniotomy was for head trauma, code as 5.2.1 *Chronic post-traumatic headache attributed to moderate or severe head injury*.

*Comments:*

Immediate post-operative headache may occur in up to 80% of patients after craniotomy but resolves in most patients within 7 days. Fewer than one-quarter develop persistent (>3 months) headache related to the surgical procedure. Posterior fossa procedures, especially suboccipital craniotomies performed for acoustic neuromas, are more likely to be associated with post-craniotomy headache.

The pathogenesis of chronic headache after craniotomy is unclear but may involve meningeal inflammation, nerve entrapment, adhesion of muscle to dura or other mechanisms. Modifications in the operative procedure, including the use of osteoplastic cranioplasty, may lead to a reduction in the incidence of post-craniotomy headache by preventing adhesion of muscle and fascia to the underlying dura.

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## 6. HEADACHE ATTRIBUTED TO CRANIAL OR CERVICAL VASCULAR DISORDER

- 6.1 Headache attributed to ischaemic stroke or transient ischaemic attack
  - 6.1.1 Headache attributed to ischaemic stroke (cerebral infarction)
  - 6.1.2 Headache attributed to transient ischaemic attack (TIA)
- 6.2 Headache attributed to non-traumatic intracranial haemorrhage
  - 6.2.1 Headache attributed to intracerebral haemorrhage
  - 6.2.2 Headache attributed to subarachnoid haemorrhage (SAH)
- 6.3 Headache attributed to unruptured vascular malformation
  - 6.3.1 Headache attributed to saccular aneurysm
  - 6.3.2 Headache attributed to arteriovenous malformation (AVM)
  - 6.3.3 Headache attributed to dural arteriovenous fistula
  - 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 central nervous system (CNS) angiitis
  - 6.4.3 Headache attributed to secondary central nervous system (CNS) angiitis
- 6.5 Carotid or vertebral artery pain
  - 6.5.1 Headache or facial or neck pain attributed to arterial dissection
  - 6.5.2 Post-endarterectomy headache
  - 6.5.3 Carotid angioplasty headache
  - 6.5.4 Headache attributed to intracranial endovascular procedures
  - 6.5.5 Angiography headache
- 6.6 Headache attributed to cerebral venous thrombosis (CVT)
- 6.7 Headache attributed to other intracranial vascular disorder
  - 6.7.1 CADASIL (Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy)
  - 6.7.2 MELAS (Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes)
  - 6.7.3 Headache attributed to benign angiopathy of the central nervous system
  - 6.7.4 Headache attributed to pituitary apoplexy

### General comment

#### *Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to a vascular disorder, it is coded as a secondary headache attributed to the vascular disorder. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to a vascular disorder, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the vascular disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the vascular disorder, a marked worsening of the pre-existing headache, very good evidence that the vascular disorder can aggravate the primary headache and, finally, improvement of the headache after the acute phase of the vascular disorder.

### *Definite, probable or chronic?*

A diagnosis of *Headache attributed to vascular disorder* usually becomes definite only when the headache resolves or greatly improves within a specified time after its onset or after the acute phase of the disorder. When this is not the case, or before the specified time has elapsed, a diagnosis of *Headache probably attributed to vascular disorder* is usually applied.

The alternative, when headache does not resolve or greatly improve after 3 months, is a diagnosis of *A6.8 Chronic post-vascular-disorder headache*. This is described only in the appendix as such headaches have been poorly documented, and research is needed to establish better criteria for causation.

## **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 recognise 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 all vascular disorders listed here, the diagnostic criteria include whenever possible:

- A. Headache with one (or more) of the stated characteristics (if any are known) and fulfilling criteria C and D
- B. Major diagnostic criteria of the vascular disorder
- C. The temporal relationship of the association with, and/or other evidence of causation by, the vascular disorder
- D. Improvement or disappearance of headache within a defined period<sup>1</sup> after its onset or after the vascular disorder has remitted or after its acute phase

### *Note:*

1. For headache attributed to some vascular disorders, criterion D is not indicated because there are not enough data to give any time limit for improvement or disappearance of the headache.

## **6.1 Headache attributed to ischaemic stroke or transient ischaemic attack**

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

#### *Diagnostic criteria:*

- A. Any new acute headache fulfilling criterion C
- B. Neurological signs and/or neuroimaging evidence of a recent ischaemic stroke
- C. Headache develops simultaneously with or in very close temporal relationship to signs or other evidence of ischaemic stroke

#### *Comments:*

The headache of ischaemic stroke is accompanied by focal neurological signs and/or alterations in consciousness usually allowing easy differentiation from the primary headaches. It is usually of moderate intensity and has no specific characteristics.

Headache accompanies ischaemic stroke in 17-34% of cases; it is more frequent in basilar- than in carotid-territory strokes. It is of little practical value in establishing stroke aetiology except that headache is very rarely associated with lacunar infarcts but extremely common in arterial dissection.

### **6.1.2 Headache attributed to transient ischaemic attack (TIA)**

#### *Diagnostic criteria:*

- A. Any new acute headache fulfilling criteria C and D
- B. Focal neurological deficit of ischaemic origin lasting <24 hours
- C. Headache develops simultaneously with onset of focal deficit
- D. Headache resolves within 24 hours

#### *Comment:*

Whilst more common with basilar- than carotid-territory TIA, headache is very rarely a prominent symptom of TIA. The differential diagnosis between TIA with headache and an attack of migraine with aura may be particularly difficult. The mode of onset is crucial: the focal deficit is typically sudden in a TIA and more frequently progressive in a migrainous aura. Furthermore, positive phenomena (eg, scintillating scotoma) are far more common in migrainous aura than in TIA whereas negative phenomena are more usual in 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 haematoma is coded as 5.1.1 *Acute post-traumatic headache attributed to moderate or severe head injury* or 5.2.1 *Chronic post-traumatic headache attributed to moderate or severe head injury*.

Headache attributed to traumatic epidural haematoma is coded as 5.5.1 *Headache attributed to epidural haematoma*; headache attributed to traumatic subdural haematoma is coded as 5.5.2 *Headache attributed to subdural haematoma*.

### **6.2.1 Headache attributed to intracerebral haemorrhage**

#### *Diagnostic criteria:*

- A. Any new acute headache fulfilling criterion C
- B. Neurological signs or neuroimaging evidence of recent non-traumatic intracerebral haemorrhage
- C. Headache develops simultaneously with or in very close temporal relation to intracerebral haemorrhage

#### *Comments:*

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

Headache is more frequent and more severe in haemorrhagic than in ischaemic stroke. It is usually overshadowed by focal deficits or coma, but it can be the prominent early feature of cerebellar haemorrhage which may require emergency surgical decompression.

6.2.1 *Headache attributed to intracerebral haemorrhage* is more often due to associated subarachnoid blood and to local compression than to intracranial hypertension. It can occasionally present as thunderclap headache.

### **6.2.2 Headache attributed to subarachnoid haemorrhage (SAH)**

#### *Diagnostic criteria:*

- A. Severe headache of sudden onset fulfilling criteria C and D
- B. Neuroimaging (CT or MRI T2 or flair) or CSF evidence of non-traumatic subarachnoid haemorrhage with or without other clinical signs
- C. Headache develops simultaneously with haemorrhage
- D. Headache resolves within 1 month

#### *Comments:*

Subarachnoid haemorrhage is by far the most common cause of intense and incapacitating headache of abrupt onset (thunderclap headache) and remains a serious condition (50% of patients die following SAH, often before arriving at hospital, and 50% of survivors are left disabled).

Excluding trauma, 80% of cases result from ruptured saccular aneurysms.

The headache of SAH is often unilateral at onset and accompanied by nausea, vomiting, disorders of consciousness and nuchal rigidity and less frequently by fever and cardiac dysrhythmia. However, it may be less severe and without associated signs. The abrupt onset is the key feature. Any patient with headache of abrupt onset or thunderclap headache should be evaluated for SAH. Diagnosis is confirmed by CT scan without contrast or MRI (flair sequences) which have a sensitivity of over 90% in the first 24 hours. If neuroimaging is negative, equivocal or technically inadequate, a lumbar puncture should be performed.

Subarachnoid haemorrhage is a neurosurgical emergency.

### **6.3 Headache attributed to unruptured vascular malformation**

*Coded elsewhere:*

Headache attributed to ruptured vascular malformation is coded as 6.2.1 *Headache attributed to intracerebral haemorrhage* or 6.2.2 *Headache attributed to subarachnoid haemorrhage*.

#### **6.3.1 Headache attributed to saccular aneurysm**

*Diagnostic criteria:*

- A. Any new acute headache including thunderclap headache and/or painful third nerve palsy fulfilling criteria C and D
- B. Neuroimaging evidence of saccular aneurysm
- C. Evidence exists of causation by the saccular aneurysm
- D. Headache resolves within 72 hours
- E. Subarachnoid haemorrhage, intracerebral haemorrhage and other causes of headache ruled out by appropriate investigations

*Comments:*

Headache is reported by approximately 18% of patients with unruptured cerebral aneurysm.

It usually has no specific features. However, thunderclap headache occurs *prior to* confirmed aneurysmal SAH in about 50% of patients. Although thunderclap headache may occur in the absence of vascular malformations, such malformations should be looked for by appropriate non-invasive investigations (MRA or CT angiography) and, in doubtful cases, by conventional angiography. A classic variety of “warning pain” (signalling impending rupture or progressive enlargement) is an acute third nerve palsy with retro-orbital pain and a dilated pupil, indicating an aneurysm of the posterior communicating cerebral artery or end of carotid artery.

#### **6.3.2 Headache attributed to arteriovenous malformation (AVM)**

*Diagnostic criteria:*

- A. Any new acute headache fulfilling criteria C and D
- B. Neuroimaging evidence of arteriovenous malformation
- C. Evidence exists of causation by the arteriovenous malformation
- D. Headache resolves within 72 hours
- E. Subarachnoid haemorrhage, intracerebral haemorrhage and other causes of headache ruled out by appropriate investigations

*Comments:*

Cases have been reported highlighting the association of AVM with a variety of headaches such as cluster headache, chronic paroxysmal hemicrania (CPH) and short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT), but these cases had atypical features. There is no good evidence of a relationship between AVM and these primary headaches when they are typical.

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 large AVM series, migraine as a presenting symptom is rare, much less common than haemorrhage, epilepsy or focal deficits.

### **6.3.3 Headache attributed to dural arterio-venous fistula**

*Diagnostic criteria:*

- A. Any new acute headache fulfilling criterion C
- B. Neuroimaging evidence of dural arteriovenous fistula
- C. Evidence exists of causation by the fistula
- D. Subarachnoid haemorrhage, intracerebral haemorrhage and other causes of headache ruled out by appropriate investigations

*Comment:*

Studies devoted to headache with dural arteriovenous fistula are lacking. A painful pulsatile tinnitus can be a presenting symptom, as well as headache with other signs of intracranial hypertension due to decrease in venous outflow and sometimes to sinus thrombosis. Carotido-cavernous fistulae may present as painful ophthalmoplegia.

### **6.3.4 Headache attributed to cavernous angioma**

*Coded elsewhere:*

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

*Diagnostic criteria:*

- A. Any new acute headache fulfilling criterion C
- B. Neuroimaging evidence of cavernous angioma
- C. Evidence exists of causation by the cavernous angioma
- D. Subarachnoid haemorrhage, intracerebral haemorrhage and other causes of headache ruled out by appropriate investigations

*Comment:*

Cavernous angiomas are increasingly recognised on MRI. There is no good study devoted to headache associated with these malformations. Headache is commonly reported as a consequence of cerebral haemorrhage or of seizures due to cavernous angioma; and should be coded to these accordingly.

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

*Diagnostic criteria:*

- A. Any new acute headache fulfilling criterion C
- B. Facial angioma, seizures and neuroimaging evidence of meningeal angioma ipsilateral to the facial angioma
- C. Evidence exists of causation by the angiomas

- D. Other causes of headache ruled out by appropriate investigations

*Comment:*

Headache is commonly reported in this condition but poorly documented. Isolated cases suggest that encephalotrigeminal or leptomenigeal angiomatosis may be a cause of symptomatic migraine, particularly of attacks with prolonged auras (possibly related to chronic oligoemia).

## **6.4 Headache attributed to arteritis**

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

*Previously used terms:*

Temporal arteritis, Horton's disease

*Diagnostic criteria:*

- A. Any new persisting headache fulfilling criteria C and D
- B. At least one of the following:
  - 1. swollen tender scalp artery with elevated erythrocyte sedimentation rate (ESR) and/or C reactive protein (CRP)
  - 2. temporal artery biopsy demonstrating giant cell arteritis
- C. Headache develops in close temporal relation to other symptoms and signs of giant cell arteritis
- D. Headache resolves or greatly improves within 3 days of high-dose steroid treatment

*Comments:*

Of all arteritides and collagen vascular diseases, giant cell arteritis is the disease most conspicuously associated with headache (which is due to inflammation of head arteries, mostly branches of the external carotid artery). The following points should be stressed:

- the variability in the characteristics of headache and other associated 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 1 week;
- there are also risks of cerebral ischaemic events and of dementia;
- on histological examination, the temporal artery may appear uninvolved in some areas (skip lesions) pointing to the necessity of serial sectioning;
- duplex scanning of the temporal arteries may visualise the thickened arterial wall (as a halo on axial sections) and may help to select the site for biopsy.

#### **6.4.2 Headache attributed to primary central nervous system (CNS) angiitis**

*Previously used terms:*

Isolated CNS angiitis, granulomatous CNS angiitis

*Diagnostic criteria:*

- A. Any new persisting headache fulfilling criteria D and E
- B. Encephalic signs of any type (*eg*, stroke, seizures, disorders of cognition or consciousness)
- C. CNS angiitis proven by cerebral or meningeal biopsy or suspected on angiographic signs in the absence of systemic arteritis
- D. Headache develops in close temporal relation to encephalic signs
- E. Headache improves within 1 month of steroid and/or immunosuppressive treatment

*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 pathogenesis of the headache is multifactorial: inflammation, stroke (ischaemic or haemorrhagic), raised intracranial pressure and/or SAH.

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 central nervous system (CNS) angiitis**

*Diagnostic criteria:*

- A. Any new persisting headache fulfilling criteria D and E
- B. Encephalic signs of any type (*eg*, stroke, seizures, disorders of cognition or consciousness)
- C. Evidence of systemic arteritis
- D. Headache develops in close temporal relation to encephalic signs
- E. Headache improves within 1 month of steroid and/or immunosuppressive treatment

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

The pathogenesis of the headache is multifactorial: inflammation, stroke (ischaemic or haemorrhagic), raised intracranial pressure and/or subarachnoid haemorrhage.

## 6.5 Carotid or vertebral artery pain

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

#### *Diagnostic criteria:*

- A. Any new headache, facial pain or neck pain of acute onset, with or without other neurological symptoms or signs and fulfilling criteria C and D
- B. Dissection demonstrated by appropriate vascular and/or neuroimaging investigations
- C. Pain develops in close temporal relation to and on the same side as the dissection
- D. Pain resolves within 1 month

#### *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 it is also the most frequent inaugural symptom (33-86% of cases).

Headache and facial and neck pain are usually unilateral (ipsilateral to the dissected artery), severe and persistent (for a mean of 4 days). However, it has no constant specific pattern and it can sometimes be very misleading, mimicking other headaches such as migraine, cluster headache, primary thunderclap headache and SAH (particularly since intracranial vertebral artery dissection can itself present with SAH). Associated signs are frequent: signs of cerebral or retinal ischaemia and local signs. A painful Horner's syndrome or a painful tinnitus of sudden onset are highly suggestive of carotid dissection.

Headache usually precedes the onset of ischaemic signs and therefore requires early diagnosis and treatment. Diagnosis is based on Duplex scanning, MRI, MRA and/or helical CT and, in doubtful cases, conventional angiography. Several of these investigations are commonly needed since any of them can be normal. There have been no randomised trials of treatment but there is a consensus in favour of heparin followed by warfarin for 3-6 months according to the quality of the arterial recovery.

### 6.5.2 Post-endarterectomy headache

#### *Diagnostic criteria:*

- A. Acute headache with one of the following sets of characteristics and fulfilling criteria C and D:
  - 1. diffuse mild pain
  - 2. unilateral cluster-like pain occurring once or twice a day in attacks lasting 2-3 hours
  - 3. unilateral pulsating severe pain
- B. Carotid endarterectomy has been performed
- C. Headache, in the absence of dissection, develops within 1 week of surgery
- D. Headache resolves within 1 month after surgery

#### *Comment:*

Three subforms of headache have been described after carotid endarterectomy. The most frequent (up to 60% of cases) is a diffuse, mild isolated headache occurring in the first few days after surgery. It is a benign self-limiting condition. The second type (reported in up to 38% of cases) is a unilateral cluster-like pain with attacks, lasting 2-3 hours, occurring once or twice a day. It resolves

in about 2 weeks. The third type is part of the rare hyperperfusion syndrome with a unilateral pulsating and severe pain occurring after an interval of 3 days after surgery. It often precedes a rise in blood pressure and the onset of seizures or neurological deficits on about the 7th day. Urgent treatment is required since these symptoms can herald cerebral haemorrhage.

### **6.5.3 Carotid angioplasty headache**

#### *Diagnostic criteria:*

- A. Any new acute headache fulfilling criteria C and D
- B. Extra- or intracranial angioplasty has been performed
- C. Headache, in the absence of dissection, develops during or within a week of angioplasty
- D. Headache resolves within 1 month

#### *Comments:*

Percutaneous transluminal angioplasty (PTA) and stenting *versus* surgery are presently undergoing randomised trials. Data on headache are still scarce and headache is not mentioned in large series of carotid PTA. In a small series of 53 patients, cervical pain occurred in 51% of patients and head pain in 33% during balloon inflation. It mostly disappeared within seconds of balloon deflation.

Headache as part of a hyperperfusion syndrome (see 6.5.2 *Post-endarterectomy headache*) has also been reported after carotid PTA.

### **6.5.4 Headache attributed to intracranial endovascular procedures**

#### *Diagnostic criteria:*

- A. Unilateral severe localised headache of abrupt onset and fulfilling criteria C and D
- B. Intracranial angioplasty or embolisation has been performed
- C. Headache develops within seconds of the procedure
- D. Headache resolves within 24 hours after the end of the procedure

#### *Comment:*

A very specific subtype of headache has been reported after balloon inflation or embolisation of an AVM or aneurysm. It is a severe pain of abrupt onset, localised in specific areas according to the artery involved, occurring within a few seconds of the procedure and disappearing rapidly.

### **6.5.5 Angiography headache**

#### *Diagnostic criteria:*

- A. Acute headache with one of the following sets of characteristics and fulfilling criteria C and D
  1. diffuse burning severe headache
  2. headache, in a patient with migraine, having the features of migraine
- B. Intra-arterial carotid or vertebral angiography has been performed
- C. Headache develops during angiography
- D. Headache resolves within 72 hours

*Comment:*

The intracarotid or intravertebral injection of contrast induces a diffuse severe headache with a burning sensation which resolves spontaneously. The injection can also trigger a migraine attack in a person who has migraine. This should be coded both under 1. *Migraine* (as the appropriate subtype) and as 6.5.5 *Angiography headache*.

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

*Diagnostic criteria:*

- A. Any new headache, with or without neurological signs, fulfilling criteria C and D
- B. Neuroimaging evidence of cerebral venous thrombosis
- C. Headache (and neurological signs if present) develops in close temporal relation to CVT
- D. Headache resolves within 1 month after appropriate treatment

*Comments:*

Headache is by far the most frequent symptom of CVT (present in 80-90% of cases) and it is also the most frequent inaugural symptom. It has no specific characteristics. Most often it is diffuse, progressive, severe and associated with other signs of intracranial hypertension. It can also be unilateral and sudden, and sometimes very misleading, mimicking migraine, primary thunderclap headache, CSF hypotension or SAH (of which it can be a cause). 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, any recent persisting headache should raise suspicion, particularly in the presence of an underlying prothrombotic condition. Diagnosis is based on neuroimaging (MRI plus MRA or CT scan plus CT angiography or intra-arterial angiography in doubtful cases). Treatment should be started as early as possible and includes symptomatic treatment, heparin followed by at least 6 months of oral anticoagulation and, whenever indicated, treatment of the underlying cause.

## **6.7 Headache attributed to other intracranial vascular disorder**

### **6.7.1 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)**

*Diagnostic criteria:*

- A. Attacks of migraine with aura, with or without other neurological signs
- B. Typical white matter changes on MRI T2WI
- C. Diagnostic confirmation from skin biopsy evidence or genetic testing (Notch 3 mutations)

*Comment:*

CADASIL is a recently identified autosomal dominant (with some sporadic cases) small artery disease of the brain characterised clinically by recurrent small deep infarcts, subcortical dementia, mood disturbances and migraine with aura.

Migraine with aura is present in one third of cases and, in such cases, is usually the first symptom of the disease, appearing at a mean age of 30, some 15 years before ischaemic strokes and 20-30 years before death. Attacks are typical of 1.2 *Migraine with aura* except for an unusual frequency of prolonged aura.

MRI is always abnormal with striking white matter changes on T2WI. The disease involves the smooth muscle cells in the media of small arteries and it is due to mutations of Notch 3 gene. The diagnosis is made on a simple skin biopsy with immunostaining of Notch 3 antibodies.

CADASIL is an excellent model to study the pathophysiology of migraine with aura and the relationships between it and ischaemic stroke.

### **6.7.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)**

*Diagnostic criteria:*

- A. Attacks of migraine with or without aura
- B. Stroke-like episodes and seizures
- C. Genetic abnormality (3243 point mitochondrial DNA mutation in the tRNA Leu gene or other DNA MELAS point mutation)

*Comment:*

Migraine attacks are frequent in MELAS and this has led to the hypothesis that mitochondrial mutations could play a role in migraine with aura but the 3243 mutation was not detected in two groups of subjects with 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.

### **6.7.3 Headache attributed to benign (or reversible) angiopathy of the central nervous system**

*Diagnostic criteria:*

- A. Diffuse, severe headache of abrupt or progressive onset, with or without focal neurological deficits and/or seizures and fulfilling criteria C and D
- B. "Strings and beads" appearance on angiography and subarachnoid haemorrhage ruled out by appropriate investigations
- C. One or both of the following:
  - 1. headache develops simultaneously with neurological deficits and/or seizures
  - 2. headache leads to angiography and discovery of "strings and beads" appearance
- D. Headache (and neurological deficits, if present) resolves spontaneously within 2 months

*Comments:*

This is a poorly understood condition characterised clinically by a severe diffuse headache of variable modes of onset: it can be abrupt, mimicking SAH, or progressive rapidly over hours or more slowly over days. It is one of the identified causes of thunderclap headache. It can be the only symptom of this condition but it is usually associated with fluctuating focal neurological deficits and sometimes seizures. Angiography is, by definition, abnormal, with alternating segments of arterial constriction and dilatation.

A number of causes have been identified: the best defined is post-partum angiopathy which has been related in some cases to use of bromocriptine. The disease is self-limiting in 1-2 months without treatment and with disappearance of the arterial abnormalities but, given the diagnostic difficulty with primary CNS angiitis, a course of steroids is sometimes given.

#### **6.7.4 Headache attributed to pituitary apoplexy**

##### *Diagnostic criteria:*

- A. Severe acute retro-orbital, frontal or diffuse headache accompanied by at least one of the following and fulfilling criteria C and D:
  - 1. nausea and vomiting
  - 2. fever
  - 3. diminished level of consciousness
  - 4. hypopituitarism
  - 5. hypotension
  - 6. ophthalmoplegia or impaired visual acuity
- B. Neuroimaging evidence of acute haemorrhagic pituitary infarction
- C. Headache develops simultaneously with acute haemorrhagic pituitary infarction
- D. Headache and other symptoms and/or signs resolve within 1 month

##### *Comment:*

This rare clinical syndrome is an acute, life-threatening condition, characterised by spontaneous haemorrhagic infarction of the pituitary gland. It is one of the causes of thunderclap headache.

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

### General comment

#### *Primary or secondary headache or both?*

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 the intracranial disorder. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to an intracranial disorder, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the intracranial disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the intracranial disorder, a marked worsening of the pre-existing headache, very good evidence that the intracranial disorder can aggravate the primary headache and, finally, improvement or resolution of the headache after relief from the intracranial disorder.

#### *Definite, probable or chronic?*

A diagnosis of *Headache attributed to non-vascular intracranial disorder* usually becomes definite only when the headache resolves or greatly improves after effective treatment or spontaneous

remission of the causative disorder. If the intracranial disorder cannot be treated effectively or does not remit spontaneously, or when there has been insufficient time for this to happen, a diagnosis of *Headache probably attributed to a non-vascular intracranial disorder* is usually applied.

The alternative, when the causative disorder is effectively treated or remits spontaneously but headache does not resolve or markedly improve after 3 months, is a diagnosis of A 7.10 *Chronic post-intracranial disorder headache*. This is described only in the appendix as such headaches have been poorly documented, and research is needed to establish better criteria for causation.

## **Introduction**

In this chapter are the headaches attributed to changes in intracranial pressure. Both increased and decreased 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 on these headache types. Controlled trials of therapy are almost non-existent.

Headache persisting for more than 1 month after successful treatment or spontaneous resolution of the intracranial disorder usually has other mechanisms. Chronic headache persisting for >3 months after treatment or remission of intracranial disorders is defined in the appendix for research purposes. Such headaches exist but have been poorly studied and the appendix entries are intended to stimulate further research into such headaches and their mechanisms.

## **7.1 Headache attributed to high cerebrospinal fluid pressure**

*Coded elsewhere:*

7.4.1 *Headache attributed to increased intracranial pressure or hydrocephalus caused by neoplasm.*

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

*Previously used terms:*

Benign intracranial hypertension (BIH), pseudotumor cerebri, meningeal hydrops, serous meningitis

*Diagnostic criteria:*

- A. Progressive headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. daily occurrence
  - 2. diffuse and/or constant (non-pulsating) pain
  - 3. aggravated by coughing or straining
- B. Intracranial hypertension fulfilling the following criteria:
  - 1. alert patient with neurological examination that either is normal or demonstrates any of the following abnormalities:
    - a) papilloedema
    - b) enlarged blind spot
    - c) visual field defect (progressive if untreated)

- d) sixth nerve palsy
- 2. increased CSF pressure (>200 mm H<sub>2</sub>O in the non-obese, >250 mm H<sub>2</sub>O in the obese) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
- 3. normal CSF chemistry (low CSF protein is acceptable) and cellularity
- 4. intracranial diseases (including venous sinus thrombosis) ruled out by appropriate investigations
- 5. no metabolic, toxic or hormonal cause of intracranial hypertension
- C. Headache develops in close temporal relation to increased intracranial pressure
- D. Headache improves after withdrawal of CSF to reduce pressure to 120-170 mm H<sub>2</sub>O and resolves within 72 hours of persistent normalisation of intracranial pressure

*Comments:*

IIH most commonly occurs in young obese women.

Although the majority of patients with IIH have papilloedema, IIH without papilloedema is observed. Other symptoms or signs of IIH include intracranial noises, tinnitus, transient visual obscurations and diplopia.

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

*Coded elsewhere:*

Headache attributed to increased intracranial pressure due to head trauma, vascular disorder or intracranial infection is coded to whichever one of those disorders is present. Headache attributed to raised intracranial pressure occurring as a side-effect of medication is coded as 8.3 *Headache as an adverse event attributed to chronic medication*.

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. daily occurrence
  - 2. diffuse and/or constant (non-pulsating) pain
  - 3. aggravated by coughing or straining
- B. Intracranial hypertension fulfilling the following criteria:
  - 1. alert patient with neurological examination that either is normal or demonstrates any of the following abnormalities:
    - a) papilloedema
    - b) enlarged blind spot
    - c) visual field defect (progressive if untreated)
    - d) sixth nerve palsy
  - 2. increased CSF pressure (>200 mm H<sub>2</sub>O in the non-obese, >250 mm H<sub>2</sub>O in the obese) measured by lumbar puncture in the recumbent position or by epidural or intraventricular pressure monitoring
  - 3. normal CSF chemistry (low CSF protein is acceptable) and cellularity
  - 4. intracranial diseases (including venous sinus thrombosis) ruled out by appropriate investigations
- C. Headache develops after weeks or months of endocrine disorder, hypervitaminosis A or intake of substances (other than medications) that can elevate CSF pressure
- D. Headache resolves within 3 months after removal of the cause

*Comments:*

Normal pressure hydrocephalus does not cause headache.

### **7.1.3 Headache attributed to intracranial hypertension secondary to hydrocephalus**

*Diagnostic criteria:*

- A. Headache with at least two of the following characteristics and fulfilling criteria C and D:
  - 1. diffuse pain
  - 2. worse in the morning
  - 3. worse with Valsalva-like manoeuvres
  - 4. accompanied by vomiting
  - 5. associated with papilloedema, 6th nerve palsy, altered level of consciousness, gait instability and/or increased head circumference (in children <5 years old)
- B. High-pressure hydrocephalus fulfilling the following criteria:
  - 1. ventricular enlargement on neuroimaging
  - 2. intracranial pressure >200 mm H<sub>2</sub>O in the non-obese or >250 mm H<sub>2</sub>O in the obese
  - 3. no other intracranial disorder causing increased CSF pressure
- C. Headache develops in close temporal relation to increased CSF pressure
- D. Headache resolves within 72 hours of normalisation of CSF pressure

## **7.2 Headache attributed to low cerebrospinal fluid pressure**

### **7.2.1 Post-dural (post-lumbar) puncture headache**

*Diagnostic criteria:*

- A. Headache that worsens within 15 minutes after sitting or standing and improves within 15 minutes after lying, with at least one of the following and fulfilling criteria C and D:
  - 1. neck stiffness
  - 2. tinnitus
  - 3. hypacusia
  - 4. photophobia
  - 5. nausea
- B. Dural puncture has been performed
- C. Headache develops within 5 days after dural puncture
- D. Headache resolves either<sup>1</sup>:
  - 1. spontaneously within 1 week
  - 2. within 48 hours after effective treatment of the spinal fluid leak (usually by epidural blood patch)

*Note:*

- 1. In 95% of cases this is so. When headache persists, causation is in doubt.

### 7.2.2 CSF fistula headache

#### *Diagnostic criteria:*

- A. Headache that worsens within 15 minutes after sitting or standing, with at least one of the following and fulfilling criteria C and D:
  - 1. neck stiffness
  - 2. tinnitus
  - 3. hypacusia
  - 4. photophobia
  - 5. nausea
- B. A known procedure or trauma has caused persistent CSF leakage with at least one of the following:
  - 1. evidence of low CSF pressure on MRI (*eg*, pachymeningeal enhancement)
  - 2. evidence of CSF leakage on conventional myelography, CT myelography or cisternography
  - 3. CSF opening pressure <60 mm H<sub>2</sub>O in sitting position
- C. Headache develops in close temporal relation to CSF leakage
- D. Headache resolves within 7 days of sealing the CSF leak

### 7.2.3 Headache attributed to spontaneous (or idiopathic) low CSF pressure

#### *Previously used terms:*

Spontaneous intracranial hypotension, primary intracranial hypotension, low CSF-volume headache, hypoliqorrhoeic headache

#### *Diagnostic criteria:*

- A. Diffuse and/or dull headache that worsens within 15 minutes after sitting or standing, with at least one of the following and fulfilling criterion D:
  - 1. neck stiffness
  - 2. tinnitus
  - 3. hypacusia
  - 4. photophobia
  - 5. nausea
- B. At least one of the following:
  - 1. evidence of low CSF pressure on MRI (*eg*, pachymeningeal enhancement)
  - 2. evidence of CSF leakage on conventional myelography, CT myelography or cisternography
  - 3. CSF opening pressure <60 mm H<sub>2</sub>O in sitting position
- C. No history of dural puncture or other cause of CSF fistula
- D. Headache resolves within 72 hours after epidural blood patching

#### *Comments:*

The underlying disorder may be low CSF volume. A history of trivial increase in intracranial pressure (*eg*, on vigorous coughing) is often elicited. In other cases a sudden drop in atmospheric pressure has occurred.

Postural headache resembling that of low CSF pressure has been reported after coitus. Such headache should be coded here because it is due to CSF leakage.

Many patients with spontaneous low CSF pressure headache respond to epidural blood patching, epidural saline infusion or pharmacological therapies such as intravenous caffeine or conventional analgesics. Some have spontaneous resolution of their headache, while others relapse after initial successful treatment. Cases of dural sleeve herniation, particularly in the thoracic area, have been reported and have been successfully treated surgically.

Dural puncture should be avoided in patients with positive MRI signs such as meningeal enhancement with contrast.

### **7.3 Headache attributed to non-infectious inflammatory disease**

#### **7.3.1 Headache attributed to neurosarcoidosis**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Evidence of neurosarcoidosis<sup>1</sup>
- C. Headache develops in temporal relation to neurosarcoidosis
- D. Headache resolves within 3 months after successful treatment of neurosarcoidosis

*Note:*

1. Evidence of neurosarcoidosis includes cranial nerve lesions, intracranial space-occupying lesion on MRI, aseptic meningitis and/or periventricular inflammatory focal lesions and homogeneously enhancing mass lesions that are confirmed on biopsy as non-caseating granulomas.

#### **7.3.2 Headache attributed to aseptic (non-infectious) meningitis**

*Diagnostic criteria:*

- A. Diffuse headache fulfilling criterion D
- B. Examination of CSF shows lymphocytic pleocytosis, mildly elevated protein and normal glucose in the absence of infectious organisms
- C. Use of one of the following: ibuprofen, immunoglobulins, penicillin or trimethoprim, intrathecal injections, or insufflations
- D. Headache resolves within 3 months after withdrawal of the offending substance

#### **7.3.3 Headache attributed to other non-infectious inflammatory disease**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Evidence of one of the inflammatory diseases known to be associated with headache<sup>1</sup>
- C. Headache develops in close temporal relation to the inflammatory disorder
- D. Headache resolves within 3 months after successful treatment of the inflammatory disorder

*Note:*

1. Headache can be associated with but is not usually a presenting or dominant symptom of acute demyelinating encephalomyelitis (ADEM), systemic lupus erythematosus (SLE), Behçet's syndrome, anti-phospholipid antibody syndrome, Vogt-Koyanagi-Harada syndrome.

#### **7.3.4 Headache attributed to lymphocytic hypophysitis**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criterion C
- B. Hypopituitarism fulfilling the following criteria:
  1. MRI demonstrates symmetrical pituitary enlargement with homogeneous contrast-enhancement
  2. biopsy confirmation of lymphocytic hypophysitis
- C. Headache develops in close temporal relation to hypopituitarism

*Comments:*

Lymphocytic hypophysitis is often accompanied by hyperprolactinaemia (50% of cases) or autoantibodies against hypophysial cytosol protein (20%).

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

### **7.4 Headache attributed to intracranial neoplasm**

#### **7.4.1 Headache attributed to increased intracranial pressure or hydrocephalus caused by neoplasm**

*Diagnostic criteria:*

- A. Diffuse non-pulsating headache with at least one of the following characteristics and fulfilling criteria C and D:
  1. associated with nausea and/or vomiting
  2. worsened by physical activity and/or manoeuvres known to increase intracranial pressure (such as Valsalva manoeuvre, coughing or sneezing)
  3. occurring in attack-like episodes<sup>1</sup>
- B. Space-occupying intracranial tumour demonstrated by CT or MRI and causing hydrocephalus<sup>2</sup>
- C. Headache develops and/or deteriorates in close temporal relation to the hydrocephalus
- D. Headache improves within 7 days after surgical removal or volume-reduction of tumour

*Notes:*

1. Onset of headache can be sudden (thunderclap headache) and, in such cases, associated with loss of consciousness.
2. For example, colloid cyst of the IIIrd ventricle.

#### **7.4.2 Headache attributed directly to neoplasm**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. progressive
  - 2. localised
  - 3. worse in the morning
  - 4. aggravated by coughing or bending forward
- B. Intracranial neoplasm shown by imaging
- C. Headache develops in temporal (and usually spatial) relation to the neoplasm
- D. Headache resolves within 7 days after surgical removal or volume-reduction of neoplasm or treatment with corticosteroids

#### **7.4.3 Headache attributed to carcinomatous meningitis**

*Diagnostic criteria:*

- A. Diffuse or localised headache fulfilling criterion C
- B. Carcinomatous meningitis proven by (repeated) CSF examination and/or dural enhancement on MRI
- C. Headache develops and/or deteriorates with advancing disease

*Comment:*

Headache may improve temporarily with intrathecal chemotherapy or prednisone (prednisolone).

#### **7.4.4 Headache attributed to hypothalamic or pituitary hyper- or hyposecretion**

*Diagnostic criteria:*

- A. Bilateral, frontotemporal and/or retro-orbital headache fulfilling criteria C and D
- B. At least one of the following:
  - 1. prolactin, growth hormone (GH) and adrenocorticotrophic hormone (ACTH) hypersecretion associated with microadenomas <10 mm in diameter
  - 2. disorder of temperature regulation, abnormal emotional state, altered thirst and appetite and change in level of consciousness associated with hypothalamic tumour
- C. Headache develops during endocrine abnormality
- D. Headache resolves within 3 months after surgical resection or specific and effective medical therapy

#### **7.5 Headache attributed to intrathecal injection**

*Diagnostic criteria:*

- A. Diffuse headache remaining present in the recumbent position and fulfilling criteria C and D
- B. Intrathecal injection has been given
- C. Headache develops within 4 hours after intrathecal injection
- D. Headache resolves within 14 days<sup>1</sup>

*Note:*

1. If headache persists beyond 14 days, the likely diagnosis is 7.2.2 *CSF fistula headache*.

## **7.6 Headache attributed to epileptic seizure**

*Comment:*

The association between migraine and epilepsy is complex and bi-directional. It may be related to genetic and/or environmental risk factors that increase neuronal excitability or decrease the threshold to both types of attacks. Migraine and epilepsy may coexist without either being a contributing risk factor for the other. Migraine and epilepsy may be co-morbid as certain brain disorders (eg, MELAS) predispose patients to both epilepsy and migraine occurring remotely from each other. There appears also to be a high incidence of migraine in certain forms of epilepsy such as benign occipital epilepsy, benign rolandic epilepsy and corticoreticular epilepsy with absence seizures. Furthermore, structural lesions such as arteriovenous malformations may present with clinical features of migraine with aura along with seizures, usually accompanied by headache. Finally, seizures have been reported to occur during or immediately following a migraine aura. The term *migralsepsy* has been used to denote epileptic seizures occurring between the migrainous aura and the headache phase of migraine. There should be no reason why epileptic seizures, so vulnerable to extrinsic and intrinsic precipitating factors, could not be susceptible to cortical changes induced by migraine. However, this is so extremely rare that only a few case reports have been published despite that migraine and epilepsy are among the commonest brain diseases. According to a recent review, most of these are genuine occipital seizures imitating migraine aura. For example, two of the three “migralsepsy” patients of Lennox and Lennox (1960) seemed to have symptomatic and idiopathic occipital epilepsy with visual hallucinations.

### **7.6.1 Hemicrania epileptica**

*Diagnostic criteria:*

- A. Headache lasting seconds to minutes, with features of migraine, fulfilling criteria C and D
- B. The patient is having a partial epileptic seizure
- C. Headache develops synchronously with the seizure and is ipsilateral to the ictal discharge
- D. Headache resolves immediately after the seizure

*Comment:*

Synchronous ipsilateral headache with migrainous features occurring as an ictal manifestation of the seizure discharge is recognised, albeit rare. Diagnosis requires the simultaneous onset of headache with electroencephalographically-demonstrated ictal discharge.

### **7.6.2 Post-ictal headache**

*Diagnostic criteria:*

- A. Headache with features of tension-type headache or, in a patient with migraine, of migraine headache and fulfilling criteria C and D
- B. The patient has had a partial or generalised epileptic seizure
- C. Headache develops within 3 hours following the seizure
- D. Headache resolves within 72 hours after the seizure

*Comments:*

Post-ictal headache with migrainous features is a well-recognised consequence of a seizure discharge. Post-ictal headache is often indistinguishable from migraine headache and associated with nausea and vomiting. It is equally common in those with or without a family history of migraine. Other similarities with migraine headache are that, in some patients, post-ictal headache develops 3-15 minutes after the end of visual hallucinations (and it is longer and more severe after visual seizures of longer duration). Similar post-ictal headache has been reported in patients with symptomatic epilepsy but it is mainly emphasised in idiopathic occipital seizures. It may be that the seizure discharges in the occipital lobes trigger a genuine migraine headache through trigeminovascular or brainstem mechanisms.

In a study of 100 patients with epilepsy, post-ictal headache occurred in 51 and most commonly lasted 6-72 hours. Major seizures were more often associated with post-ictal headache than were minor attacks. Nine patients in this series also had migraine: in eight, a typical albeit mild migraine attack was provoked by seizures. Post-ictal headache in the 43 who did not develop migraine was accompanied by vomiting in 11 cases, photophobia in 14 cases and vomiting with photophobia in 4 cases. Furthermore, post-ictal headache was accentuated by coughing, bending and sudden head movements and relieved by sleep. It is, therefore, clear that seizures provoke a syndrome similar to the headache phase of migraine in 50% of epileptics.

## **7.7 Headache attributed to Chiari malformation type I (CM1)**

*Diagnostic criteria:*

- A. Headache characterised by at least one of the following and fulfilling criterion D:
  - 1. precipitated by cough and/or Valsalva manoeuvre
  - 2. protracted (hours to days) occipital and/or sub-occipital headache
  - 3. associated with symptoms and/or signs of brainstem, cerebellar and/or cervical cord dysfunction
- B. Cerebellar tonsillar herniation as defined by one of the following on craniocervical MRI:
  - 1.  $\geq 5$  mm caudal descent of the cerebellar tonsils
  - 2.  $\geq 3$  mm caudal descent of the cerebellar tonsils plus at least one of the following indicators of crowding of the subarachnoid space in the area of the craniocervical junction:
    - a) compression of the CSF spaces posterior and lateral to the cerebellum
    - b) reduced height of the supraocciput
    - c) increased slope of the tentorium
    - d) kinking of the medulla oblongata
- C. Evidence of posterior fossa dysfunction, based on at least two of the following:
  - 1. otoneurological symptoms and/or signs (*eg*, dizziness, dysequilibrium, sensations of alteration in ear pressure, hypacusia or hyperacusia, vertigo, down-beat nystagmus, oscillopsia)
  - 2. transient visual symptoms (spark photopsias, visual blurring, diplopia or transient visual field deficits)
  - 3. demonstration of clinical signs relevant to cervical cord, brainstem or lower cranial nerves or of ataxia or dysmetria
- D. Headache resolves within 3 months after successful treatment of the Chiari malformation

*Comments:*

Headache is often descriptively similar to primary cough headache with the exception of possibly longer duration (minutes rather than seconds).

Headache is the most common symptom of Chiari malformation type I (CM1), but patients may also have localised vestibulo-ocular (74% of cases), lower cranial nerve, brainstem, cerebellar (50%) and/or spinal cord dysfunction suggestive of syringomyelia (66%). Although no specific criteria currently exist to characterise headache attributed to CM1, rigid adherence to the clinical and radiological criteria described above is recommended prior to surgical intervention. However, these criteria require validation and will inevitably be altered in future revisions of *The International Classification of Headache Disorders*. Prospective studies with long-term surgical outcome are needed.

### **7.8 Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis (HaNDL)**

*Previously used terms:*

Migraine with cerebrospinal pleocytosis; pseudomigraine with lymphocytic pleocytosis

*Diagnostic criteria:*

- A. Episodes of moderate or severe headache lasting hours before resolving fully and fulfilling criteria C and D
- B. Cerebrospinal fluid pleocytosis with lymphocytic predominance (>15 cells/ $\mu$ l) and normal neuroimaging, CSF culture and other tests for aetiology
- C. Episodes of headache are accompanied by or shortly follow transient neurological deficits and commence in close temporal relation to the development of CSF pleocytosis
- D. Episodes of headache and neurological deficits recur over <3 months

*Comments:*

This syndrome, first clearly delineated by Bartleson *et al* (1981), has also been referred to in the literature as a migrainous syndrome with cerebrospinal pleocytosis and as pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis. The clinical picture is of one to >20 discrete episodes of neurological deficits accompanied or followed by moderate to severe headache. Most of the episodes last hours. The neurological manifestations, involving either cerebral hemisphere and/or the brainstem/cerebellum, are most commonly sensory symptoms (78% of reported cases), aphasia (66%) and motor deficits (56%). Migraine-aura-like visual symptoms are relatively uncommon (18%). Some individuals report a “march” of symptoms similar to that reported in typical migraine aura. Patients are asymptomatic between episodes.

In addition to CSF lymphocytosis (10-760 cells/ $\mu$ l), there are elevations of CSF total protein (20-250 mg/dl) in >90% of cases and of the CSF opening pressure (100-400 mm H<sub>2</sub>O] in >50% of cases. Papilloedema is occasionally present. Routine CT and MRI scans (with or without intravenous contrast) and angiography are virtually always normal. Microbiological studies have been uniformly normal. EEG and SPECT scans may show focally abnormal areas consistent with the focal neurological deficits.

The CSF pleocytosis eventually normalises on repeat sampling. Although no large systematic long-term follow-up studies have been reported, it appears that some patients with this syndrome may experience recurrence of it.

Most patients with this syndrome have no prior history of migraine. The clinician must consider other diagnoses that may share some of its clinical features, including familial hemiplegic migraine, neuroborreliosis, neurosyphilis, neurobrucellosis, mycoplasma, meningitis, granulomatous and neoplastic arachnoiditis, encephalitis and CNS vasculitis.

## **7.9 Headache attributed to other non-vascular intracranial disorder**

### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. daily occurrence
  - 2. diffuse pain
  - 3. aggravated by Valsalva manoeuvre
- B. Evidence of an intracranial disorder other than those described above
- C. Headache develops in close temporal relation to the intracranial disorder
- D. Headache resolves within 3 months after cure or spontaneous remission of the intracranial disorder

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### *7.8 Syndrome of transient headache and neurologic deficits with cerebrospinal fluid lymphocytosis*

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## 8. HEADACHE ATTRIBUTED TO A SUBSTANCE OR ITS WITHDRAWAL

- 8.1 Headache induced by acute substance use or exposure
  - 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-headache
  - 8.1.2 Phosphodiesterase (PDE) inhibitor-induced headache
  - 8.1.3 Carbon monoxide-induced headache
  - 8.1.4 Alcohol-induced headache.
    - 8.1.4.1 Immediate alcohol-induced headache
    - 8.1.4.2 Delayed alcohol-induced headache
  - 8.1.5 Headache induced by food components and additives
    - 8.1.5.1 Monosodium glutamate-induced headache
  - 8.1.6 Cocaine-induced headache
  - 8.1.7 Cannabis-induced headache
  - 8.1.8 Histamine-induced headache
    - 8.1.8.1 Immediate histamine-induced headache
    - 8.1.8.2 Delayed histamine-induced headache
  - 8.1.9 Calcitonin gene-related peptide (CGRP)-induced headache
    - 8.1.9.1 Immediate CGRP-induced headache
    - 8.1.9.2 Delayed CGRP-induced headache
  - 8.1.10 Headache as an acute adverse event attributed to medication used for other indications
  - 8.1.11 Headache attributed to other acute substance use or exposure
- 8.2 Medication-overuse headache (MOH)
  - 8.2.1 Ergotamine-overuse headache
  - 8.2.2 Triptan-overuse headache
  - 8.2.3 Analgesic-overuse headache
  - 8.2.4 Opioid-overuse headache
  - 8.2.5 Combination analgesic-overuse headache
  - 8.2.6 Medication-overuse headache attributed to combination of acute medications
  - 8.2.7 Headache attributed to other medication overuse
  - 8.2.8 Probable medication-overuse headache
- 8.3 Headache as an adverse event attributed to chronic medication
  - 8.3.1 Exogenous hormone-induced headache
- 8.4 Headache attributed to substance withdrawal
  - 8.4.1 Caffeine-withdrawal headache
  - 8.4.2 Opioid-withdrawal headache
  - 8.4.3 Oestrogen-withdrawal headache
  - 8.4.4 Headache attributed to withdrawal from chronic use of other substances

*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, 10.3.6 Headache attributed to acute pressor response to an exogenous agent.*

## General comment

### *Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to substance exposure, it is coded as a secondary headache attributed to the substance. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to substance exposure, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the substance. Factors that support adding the latter diagnosis are: a very close temporal relation to the substance exposure, a marked worsening of the pre-existing headache, very good evidence that the substance can aggravate the primary headache and, finally, improvement or resolution of the headache after termination of effect of the substance.

### *Definite, probable or chronic?*

A diagnosis of *Headache attributed to a substance* usually becomes definite only when the headache resolves or greatly improves after termination of exposure to the substance. When exposure to a substance ceases but headache does not resolve or markedly improve after 3 months, a diagnosis of A8.5 *Chronic post-substance exposure headache*, described in the appendix, may be considered. However, such headaches have not been documented and the criteria are proposed only for research purposes.

In the particular case of 8.2 *Medication-overuse headache*, a period of 2 months after cessation of overuse is stipulated in which improvement (resolution of headache, or reversion to its previous pattern) must occur if the diagnosis is to be definite. Prior to cessation, or pending improvement within 2 months after cessation, the diagnosis 8.2.8 *Probable medication-overuse headache* should be applied. If such improvement does not then occur within the 2 months, this diagnosis must be discarded.

## Introduction

Migraineurs are physiologically and perhaps psychologically hyper-responsive to a variety of internal and external stimuli. Alcohol, food and food additives and chemical and drug ingestion and withdrawal have all been reported to provoke or activate migraine in susceptible individuals. The association is often based on anecdotal data and reports of adverse drug reactions.

The fact that these stimuli are associated with headache does not prove causation or eliminate the need to consider other aetiologies. Because common events happen commonly, the association between a headache and an exposure to a substance may be mere coincidence. Headache can occur just on the basis of chance. Headache can be a symptom of a systemic disease, and drugs given to treat such a condition will be associated with headache. In acute migraine drug trials, headache, as well as associated symptoms, is listed as an adverse drug reaction despite that it is a symptom of the treated disorder and not the result of treatment. Some disorders may predispose to substance related headache. Alone, neither the drug nor the condition would produce headache. A nonsteroidal anti-inflammatory drug may produce headache by inducing aseptic meningitis in susceptible individuals.

Finally, some acute or chronic substance exposures have been proven to be causally related to headache.

## 8.1 Headache induced by acute substance use or exposure

*Coded elsewhere:*

10.3.6 Headache attributed to acute pressor response to an exogenous agent.

### **Introduction**

This group of headache disorders can be caused 1) by an unwanted effect of a toxic substance, 2) by an unwanted effect of a substance in normal therapeutic use and 3) in experimental studies.

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

Headache as a side effect has been recorded with many drugs, often as just a reflection of the very high prevalence of headache. Only when it occurs more often after 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 NO donors, such studies have led to a deeper understanding of the involvement of neurotransmitter mechanisms in primary headaches. A number of substances such as NO donors and histamine induce an immediate headache in normal volunteers and in migraineurs. However, it is now clear that sufferers of primary headache also develop a delayed headache one to several hours after the inducing substance has been cleared from the blood.

Knowing the potential headache-inducing effects of substances in clinical use is important in order to label these substances appropriately. In general, migraine sufferers are much more susceptible to such headaches than other individuals and the same may be true for sufferers of chronic tension-type headache, episodic tension-type headache and cluster headache during cluster periods.

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

Combinations such as alcohol and disulfiram may cause headache when individual agents might not.

### **8.1.1 Nitric oxide (NO) donor-induced headache**

#### **8.1.1.1 Immediate NO donor-induced headache**

*Previously used terms:*

Nitroglycerine headache, dynamite headache, hot dog headache

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. pulsating quality
  - 4. aggravated by physical activity
- B. Absorption of a NO donor
- C. Headache develops within 10 minutes after absorption of NO donor
- D. Headache resolves within 1 hour after release of NO has ended

### 8.1.1.2 Delayed NO donor-induced headache

#### Diagnostic criteria:

- A. Headache, in a person who suffers from primary headache, with the characteristics of that primary headache type<sup>1</sup> and fulfilling criteria C and D
- B. Absorption of a NO donor
- C. Headache develops after NO is cleared from the blood<sup>2</sup>
- D. Headache resolves within 72 hours after single exposure

#### Notes:

1. Normal subjects rarely develop delayed NO donor-induced headache whilst migraineurs develop an attack of migraine without aura, tension-type headache sufferers develop a tension-type headache and cluster headache sufferers develop a cluster headache attack.
2. Migraine and tension-type headache develop after a mean of 5-6 hours, cluster headache typically after 1-2 hours.

#### Comments:

The headache is typically bilateral, pulsating and frontotemporal in location.

All NO donors (*eg*, amyl nitrate, erythrityl tetranitrate, glyceryl trinitrate [GTN], isosorbide mono- or dinitrate, sodium nitroprusside, mannitol hexanitrate, pentaerythrityl tetranitrate) can cause headache of this subtype particularly in persons with migraine. GTN is the best studied substance. It reliably induces headache in most normal individuals and migraine sufferers develop a more severe immediate headache than non-migraine sufferers. GTN can also cause a delayed headache in migraine sufferers which fulfils the diagnostic criteria for 1.1 *Migraine without aura*, even in patients whose spontaneous migraine attacks are with aura. In people with chronic tension-type headache, GTN has been shown to induce a delayed headache which has the characteristics of tension-type headache. It is not known if it has the same effect in sufferers of episodic tension-type headache. Cluster headache sufferers do not develop delayed headache outside cluster periods but, during a cluster period, GTN fairly reliably induces a cluster headache attack usually occurring 1-2 hours after intake. The delayed headache in those with migraine or tension-type headache occurs at variable times but on average 5-6 hours after exposure.

Headache is well known as a side effect of therapeutic use of nitroglycerine and other NO donors. With chronic use tolerance develops within a week, and GTN-induced headache disappears in most patients within that time. With intermittent use headache continues, and may be severe enough to compromise the use of NO donors for angina. Most heart patients are, however, male and beyond middle age, which probably explains why the problem is not of greater magnitude.

Other NO donors have been much less studied but available evidence suggests that they too may 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.

### 8.1.2 Phosphodiesterase (PDE) inhibitor-induced headache

#### Diagnostic criteria:

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  1. bilateral

2. frontotemporal location
  3. pulsating quality
  4. aggravated by physical activity
- B. A single dose of a phosphodiesterase inhibitor has been given
  - C. Headache develops within 5 hours of PDE inhibitor intake
  - D. Headache resolves within 72 hours

*Comment:*

PDEs are a large family of enzymes that break down cyclic nucleotides cGMP and cAMP. When PDEs are inhibited, the levels of cGMP and/or cAMP therefore increase. PDE-5 inhibitors sildenafil and dipyridamole are the only formally studied compounds in this group. The headache, unlike GTN-induced headache, is monophasic. In normal volunteers it has the characteristics of tension-type headache but in migraine sufferers it has the characteristics of migraine without aura. Headache has been noted as a side effect of sildenafil in clinical trials but only recent experimental studies have shown that, in young persons – especially females – the side effect occurs in a majority of subjects and in migraine patients sildenafil usually induces a migraine attack. Migraine sufferers should be warned of this side effect.

### **8.1.3 Carbon monoxide-induced headache**

*Previously used terms:*

Warehouse workers' headache

*Diagnostic criteria:*

- A. Bilateral and/or continuous headache, with quality and intensity that may be related to the severity of carbon monoxide intoxication<sup>1</sup>, fulfilling criteria C and D
- B. Exposure to carbon monoxide (CO)
- C. Headache develops within 12 hours of exposure
- D. Headache resolves within 72 hours after elimination of carbon monoxide

*Note:*

1. Typically: mild headache without gastrointestinal or neurological symptoms with carboxyhaemoglobin levels in the range 10-20%; moderate pulsating headache and irritability with levels of 20-30%; severe headache with nausea, vomiting and blurred vision with levels of 30-40%.

*Comments:*

With higher carboxyhaemoglobin levels (>40%) headache is not usually a complaint because of changes in consciousness.

There are no good studies of the long-term effects of CO intoxication on headache. Casuistic evidence suggests the possibility of chronic post-intoxication headache.

## 8.1.4 Alcohol-induced headache

### 8.1.4.1 Immediate alcohol-induced headache

*Previously used terms:*

Cocktail headache

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. pulsating quality
  - 4. aggravated by physical activity
- B. Ingestion of beverage containing alcohol<sup>1</sup>
- C. Headache develops within 3 hours after ingestion of alcoholic beverage
- D. Headache resolves within 72 hours

*Note:*

- 1. The effective dose has not been determined.

*Comment:*

A few subjects develop headache due to a direct effect of alcohol or alcoholic beverages. This is much rarer than delayed alcohol-induced headache.

### 8.1.4.2 Delayed alcohol-induced headache

*Previously used terms:*

Hangover headache

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. pulsating quality
  - 4. aggravated by physical activity
- B. Ingestion of a modest amount of alcoholic beverage by a migraine sufferer or an intoxicating amount by a non-migraine sufferer
- C. Headache develops after blood alcohol level declines or reduces to zero
- D. Headache resolves within 72 hours

*Comment:*

This is one of the commonest types of headache. It remains unclear whether, in addition to alcohol, other components of alcoholic beverages play a role. It also remains uncertain whether the mechanism is a delayed response to toxic effects or whether mechanisms similar to those responsible for delayed NO donor-induced headache may be involved.

The susceptibility to hangover headache of well-diagnosed headache patients compared with non-headache sufferers has not been determined. In migraine sufferers a migraine attack can be induced the next day after modest intake of alcoholic beverages, while non-migraineurs usually need a high intake of alcoholic beverages in order to develop 8.1.4.2 *Delayed alcohol-induced headache*.

### **8.1.5 Headache induced by food components and additives**

*Previously used terms:*

Dietary headache

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. pulsating quality
  - 4. aggravated by physical activity
- B. Ingestion of a minimum dose of food component or additive<sup>1</sup>
- C. Headache develops within 12 hours after substance intake
- D. Headache resolves within 72 hours after single intake

*Note:*

- 1. Phenylethylamine, tyramine and aspartame have been incriminated but their headache-inducing potential is not sufficiently validated.

#### **8.1.5.1 Monosodium glutamate-induced headache**

*Previously used terms:*

Chinese restaurant syndrome

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. aggravated by physical activity
- B. Ingestion of monosodium glutamate (MSG)
- C. Headache develops within 1 hour after MSG ingestion
- D. Headache resolves within 72 hours after single intake

*Comment:*

MSG-induced headache is typically dull or burning and non-pulsating, but may be pulsating in migraine sufferers. It is commonly associated with other symptoms of this syndrome including pressure in the chest, pressure and/or tightness in the face, burning sensations in the chest, neck or shoulders, flushing of face, dizziness and abdominal discomfort.

### **8.1.6 Cocaine-induced headache**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. pulsating quality
  - 4. aggravated by physical activity
- B. Use of cocaine

- C. Headache develops within 1 hour after cocaine use
- D. Headache resolves within 72 hours after single use

*Comment:*

Headache is a reported side effect of cocaine use. It is frequent, develops immediately or within one hour after use and is not associated with other symptoms unless there is concomitant stroke or TIA.

### **8.1.7 Cannabis-induced headache**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. stabbing or pulsating quality
  - 3. feeling of pressure in the head
- B. Use of cannabis
- C. Headache develops within 12 hours after cannabis use
- D. Headache resolves within 72 hours after single use

*Comment:*

Cannabis use is reported to cause headache associated with dryness of the mouth, paraesthesias, feelings of warmth and suffusion of the conjunctivae.

### **8.1.8 Histamine-induced headache**

*Comment:*

Histamine has been shown to cause an immediate headache in non-headache sufferers and an immediate as well as a delayed headache in migraine sufferers. The latter fulfils criteria for 1.1 *Migraine without aura*. The headache-inducing property of histamine has been studied after intravenous administration, after cutaneous administration and after inhalation: all routes of administration have the same effect. The mechanism is primarily mediated via the H1 receptor because it is almost completely blocked by mepyramine.

#### **8.1.8.1 Immediate histamine-induced headache**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontotemporal location
  - 3. pulsating quality
  - 4. aggravated by physical activity
- B. Absorption of histamine
- C. Headache develops within 10 minutes after absorption of histamine
- D. Headache resolves within 1 hour after absorption of histamine has ceased

### 8.1.8.2 *Delayed histamine-induced headache*

#### *Diagnostic criteria:*

- A. Headache, in a person who suffers from primary headache, with the characteristics of that primary headache type<sup>1</sup> and fulfilling criteria C and D
- B. Absorption of histamine
- C. Headache develops after histamine is cleared from the blood<sup>2</sup>
- D. Headache resolves within 72 hours after single exposure

#### *Notes:*

1. Normal subjects rarely develop delayed histamine-induced headache whilst migraineurs develop an attack of migraine without aura, tension-type headache sufferers develop a tension-type headache and cluster headache sufferers develop a cluster headache attack.
2. Migraine and tension-type headache develop typically after 5-6 hours, cluster headache typically after 1-2 hours.

### 8.1.9 *Calcitonin gene-related peptide (CGRP)-induced headache*

#### *Comment:*

The headache-inducing property of CGRP has been studied only in one double-blind controlled trial. There is, however, no doubt that CGRP causes an immediate headache. Delayed migraine attacks were induced in 3 out of 10 subjects. Recently, it has been shown that a CGRP antagonist is effective in the acute treatment of migraine.

#### 8.1.9.1 *Immediate CGRP-induced headache*

##### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  1. bilateral
  2. frontotemporal location
  3. pulsating quality
  4. aggravated by physical activity
- B. Absorption of CGRP
- C. Headache develops within 10 minutes after absorption of CGRP
- D. Headache resolves within 1 hour after absorption of CGRP has ceased

#### 8.1.9.2 *Delayed CGRP-induced headache*

##### *Diagnostic criteria:*

- A. Headache, in a person who suffers from primary headache, with the characteristics of that primary headache type<sup>1</sup> and fulfilling criteria C and D
- B. Absorption of CGRP
- C. Headache develops after CGRP is cleared from the blood<sup>2</sup>
- D. Headache resolves within 72 hours after infusion of CGRP

*Notes:*

1. Normal subjects rarely develop delayed CGRP-induced headache whilst migraineurs develop an attack of migraine without aura, tension-type headache sufferers develop a tension-type headache and cluster headache sufferers develop a cluster headache attack.
2. Migraine and tension-type headache develop typically after 5-6 hours, cluster headache typically after 1-2 hours.

#### **8.1.10 Headache as an acute adverse event attributed to medication used for other indications**

*Diagnostic criteria:*

- A. Headache fulfilling criteria C and D
- B. Use of a medication for a therapeutic indication other than headache
- C. Headache develops within minutes to hours after use
- D. Headache resolves within 72 hours after cessation of use

*Comments:*

Headache has been reported after use of a number of drugs. The following are the most commonly incriminated: atropine, digitalis, disulfiram, hydralazine, imipramine, nicotine, nifedipine, nimodipine. A longer list can be found in the appendix (table 1).

The headache characteristics are not very well defined in the literature but most are dull, continuous, diffuse and moderate to severe.

#### **8.1.11 Headache attributed to other acute substance use or exposure**

*Diagnostic criteria:*

- A. Headache fulfilling criteria C and D
- B. Acute use of or other acute exposure to a substance other than those described above
- C. Headache develops within 12 hours of use or exposure
- D. Headache resolves within 72 hours after single use or exposure

*Comments:*

Headache has been reported after exposure to a number of organic and inorganic substances. The following are the most commonly incriminated substances:

*Inorganic compounds:* arsenic, borate, bromate, chlorate, copper, iodine, lead, lithium, mercury, tolazoline hydrochloride.

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

The headache characteristics are not very well defined in the literature but most are dull, diffuse, continuous and moderate to severe.

## 8.2 Medication-overuse headache (MOH)

*Previously used terms:*

Rebound headache, drug-induced headache, medication-misuse headache

*Diagnostic criteria:*

- A. Headache<sup>1</sup> present on  $\geq 15$  days/month fulfilling criteria C and D
- B. Regular overuse<sup>2</sup> for  $>3$  months of one or more drugs that can be taken for acute and/or symptomatic treatment of headache<sup>3</sup>
- C. Headache has developed or markedly worsened during medication overuse
- D. Headache resolves or reverts to its previous pattern within 2 months after discontinuation of overused medication<sup>4</sup>

*Notes:*

1. The headache associated with medication overuse is variable and often has a peculiar pattern with characteristics shifting, even within the same day, from migraine-like to those of tension-type headache.
2. Overuse is defined in terms of duration and treatment days per week. What is crucial is that treatment occurs both frequently and regularly, *ie*, on 2 or more days each week. Bunching of treatment days with long periods without medication intake, practised by some patients, is much less likely to cause medication-overuse headache and does not fulfil criterion B.
3. MOH can occur in headache-prone patients when acute headache medications are taken for other indications.
4. A period of 2 months after cessation of overuse is stipulated in which improvement (resolution of headache, or reversion to its previous pattern) must occur if the diagnosis is to be definite. Prior to cessation, or pending improvement within 2 months after cessation, the diagnosis 8.2.8 *Probable medication-overuse headache* should be applied. If such improvement does not then occur within 2 months, this diagnosis must be discarded.

*Comments*

Medication-overuse headache is an interaction between a therapeutic agent used excessively and a susceptible patient. The best example is overuse of symptomatic headache drugs causing headache in the headache-prone patient. By far the most common cause of migraine-like headache occurring on  $\geq 15$  days per month and of a mixed picture of migraine-like and tension-type-like headaches on  $\geq 15$  days per month is overuse of symptomatic migraine drugs and/or analgesics. Chronic tension-type headache is less often associated with medication overuse but, especially amongst patients seen in headache centres, episodic tension-type headache has commonly become a chronic headache through overuse of analgesics.

Patients with a pre-existing primary headache who develop a new type of headache or whose migraine or tension-type headache is made markedly worse during medication overuse should be given both the diagnosis of the pre-existing headache and the diagnosis of 8.2 *Medication-overuse headache*.

The diagnosis of medication-overuse headache is clinically extremely important because patients rarely respond to preventative medications whilst overusing acute medications.

### **8.2.1 Ergotamine-overuse headache**

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Ergotamine intake on  $\geq 10$  days/month on a regular basis for  $>3$  months

#### *Comment:*

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

### **8.2.2 Triptan-overuse headache**

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Triptan intake (any formulation) on  $\geq 10$  days/month on a regular basis for  $>3$  months

#### *Comment:*

Triptan overuse may increase migraine frequency to that of chronic migraine. Evidence suggests that this occurs sooner with triptan-overuse than with ergotamine-overuse.

### **8.2.3 Analgesic-overuse headache**

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Intake of simple analgesics on  $\geq 15$  days/month<sup>1</sup> on a regular basis for  $>3$  months

#### *Note:*

1. Expert opinion rather than formal evidence suggests that use on  $\geq 15$  days/month rather than  $\geq 10$  days/month is needed to induce analgesic-overuse headache.

### **8.2.4 Opioid-overuse headache**

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Opioid intake on  $\geq 10$  days/month on a regular basis for  $>3$  months

#### *Comment:*

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

### **8.2.5 Combination analgesic-overuse headache**

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Intake of combination analgesic medications<sup>1</sup> on  $\geq 10$  days/month on a regular basis for  $>3$  months

*Note:*

1. Combinations typically implicated are those containing simple analgesics combined with opioids, butalbital and/or caffeine

### **8.2.6 Medication-overuse headache attributed to combination of acute medications**

*Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Intake of any combination of ergotamine, triptans, analgesics and/or opioids on  $\geq 10$  days/month on a regular basis for  $>3$  months without overuse of any single class alone<sup>1</sup>

*Note:*

1. The specific subform(s) 8.2.1-8.2.5 should be diagnosed if criterion B is fulfilled in respect of any one or more single class(es) of these medications

### **8.2.7 Headache attributed to other medication overuse**

*Diagnostic criteria:*

- A. Headache fulfilling criteria A, C and D for 8.2 *Medication-overuse headache*
- B. Regular overuse<sup>1</sup> for  $>3$  months of a medication other than those described above

*Note:*

1. The definition of overuse in terms of treatment days per week is likely to vary with the nature of the medication.

### **8.2.8 Probable medication-overuse headache**

*Diagnostic criteria:*

- A. Headache fulfilling criteria A and C for 8.2 *Medication-overuse headache*
- B. Medication overuse fulfilling criterion B for any one of the subforms 8.2.1 to 8.2.7
- C. One or other of the following:
  1. overused medication has not yet been withdrawn
  2. medication overuse has ceased within the last 2 months but headache has not so far resolved or reverted to its previous pattern

*Comments:*

Codable subforms of 8.2.8 *Probable medication-overuse headache* are 8.2.8.1 *Probable ergotamine-overuse headache*, 8.2.8.2 *Probable triptan-overuse headache*, 8.2.8.3 *Probable analgesic-overuse headache*, 8.2.8.4 *Probable opioid-overuse headache*, 8.2.8.5 *Probable combination analgesic-overuse headache*, 8.2.8.6 *Headache probably attributed to overuse of acute medication combinations* and 8.2.8.7 *Headache probably attributed to other medication overuse*.

Many patients fulfilling the criteria for 8.2.8 *Probable medication-overuse headache* also fulfil criteria for either 1.6.5 *Probable chronic migraine* or 2.4.3 *Probable chronic tension-type headache*. They should be coded for both until causation is established after withdrawal of the

overused medication. Patients with 1.6.5 *Probable chronic migraine* should additionally be coded for the antecedent migraine subtype (usually 1.1 *Migraine without aura*).

### **8.3 Headache as an adverse event attributed to chronic medication**

#### *Diagnostic criteria:*

- A. Headache present on >15 days/month fulfilling criteria C and D
- B. Chronic medication<sup>1</sup> for any therapeutic indication
- C. Headache develops during medication
- D. Headache resolves after discontinuation of medication<sup>2</sup>

#### *Notes:*

- 1. The definition of dose and duration will vary with the medication.
- 2. Time for resolution will vary with the medication but may be months.

#### *Comment:*

Headache can be due to a direct pharmacologic effect of medication, such as vasoconstriction producing malignant hypertension and headache, or to a secondary effect such as drug-induced intracranial hypertension. The latter is a recognised complication of long-term use of anabolic steroids, amiodarone, lithium carbonate, nalidixic acid, thyroid hormone replacement, tetracycline or minocycline.

#### **8.3.1 Exogenous hormone-induced headache**

##### *Diagnostic criteria:*

- A. Headache or migraine fulfilling criteria C and D
- B. Regular use of exogenous hormones
- C. Headache or migraine develops or markedly worsens within 3 months of commencing exogenous hormones
- D. Headache or migraine resolves or reverts to its previous pattern within 3 months after total discontinuation of exogenous hormones

##### *Comments:*

Regular use of exogenous hormones, typically for contraception or hormone replacement therapy, can be associated with increase in frequency or new development of headache or migraine.

When a woman also experiences headache or migraine associated with exogenous oestrogen-withdrawal, both codes 8.3.1 *Exogenous hormone-induced headache* and 8.4.3 *Oestrogen-withdrawal headache* should be used.

## 8.4 Headache attributed to substance withdrawal

### 8.4.1 Caffeine-withdrawal headache

#### *Diagnostic criteria:*

- A. Bilateral and/or pulsating headache fulfilling criteria C and D
- B. Caffeine consumption of  $\geq 200$  mg/day for  $>2$  weeks, which is interrupted or delayed
- C. Headache develops within 24 hours after last caffeine intake and is relieved within 1 hour by 100 mg of caffeine
- D. Headache resolves within 7 days after total caffeine withdrawal

### 8.4.2 Opioid-withdrawal headache

#### *Diagnostic criteria:*

- A. Bilateral and/or pulsating headache fulfilling criteria C and D
- B. Opioid intake daily for  $>3$  months, which is interrupted
- C. Headache develops within 24 hours after last opioid intake
- D. Headache resolves within 7 days after total opioid withdrawal

### 8.4.3 Oestrogen-withdrawal headache

#### *Diagnostic criteria:*

- A. Headache or migraine fulfilling criteria C and D
- B. Daily use of exogenous oestrogen for  $\geq 3$  weeks, which is interrupted
- C. Headache or migraine develops within 5 days after last use of oestrogen
- D. Headache or migraine resolves within 3 days

#### *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.

### 8.4.4 Headache attributed to withdrawal from chronic use of other substances

#### *Diagnostic criteria:*

- A. Bilateral and/or pulsating headache fulfilling criteria C and D
- B. Daily intake of a substance other than those described above for  $>3$  months, which is interrupted
- C. Headache develops in close temporal relation to withdrawal of the substance
- D. Headache resolves within 3 months after withdrawal

#### *Comment:*

It has been suggested, but without sufficient evidence, that withdrawal of the following substances may cause headache: corticosteroids, tricyclic antidepressants, selective serotonin reuptake inhibitors (SSRIs), non-steroidal anti-inflammatory drugs (NSAIDs).

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## 9. HEADACHE ATTRIBUTED TO INFECTION

- 9.1 Headache attributed to intracranial infection
  - 9.1.1 Headache attributed to bacterial meningitis
  - 9.1.2 Headache attributed to lymphocytic meningitis
  - 9.1.3 Headache attributed to encephalitis
  - 9.1.4 Headache attributed to brain abscess
  - 9.1.5 Headache attributed to subdural empyema
- 9.2 Headache attributed to systemic infection
  - 9.2.1 Headache attributed to systemic bacterial infection
  - 9.2.2 Headache attributed to systemic viral infection
  - 9.2.3 Headache attributed to other systemic infection
- 9.3 Headache attributed to HIV/AIDS
- 9.4 Chronic post-infection headache
  - 9.4.1 Chronic post-bacterial meningitis headache

### *Coded elsewhere:*

Headache disorders attributed to extracranial infections of the head (such as ear, eye and sinus infections) are coded as subtypes of 11. *Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures.*

### **General comment**

#### *Primary or secondary headache or both?*

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 the infection. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to an infection, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the infection. Factors that support adding the latter diagnosis are: a very close temporal relation to the infection, a marked worsening of the pre-existing headache, very good evidence that the infection can aggravate the primary headache and, finally, improvement or resolution of the headache after relief from the infection.

#### *Definite, probable or chronic?*

A diagnosis of *Headache attributed to an infection* usually becomes definite only when the headache resolves or greatly improves after effective treatment or spontaneous remission of the infection. If the infection cannot be treated effectively or does not remit spontaneously, or when there has been insufficient time for this to happen, a diagnosis of *Headache probably attributed to infection* is usually applied.

This is not the case for 9.1.1 *Headache attributed to bacterial meningitis*. It is recognised that this headache may become chronic. When the causative infection is effectively treated or remits spontaneously but headache persists after 3 months, the diagnosis changes to 9.4.1 *Chronic post-bacterial meningitis headache*.

In other cases when the infection is eliminated but headache does not resolve or markedly improve after 3 months, a diagnosis of A9.4.2 *Chronic post-non-bacterial infection headache* may be

considered. This is described only in the appendix as such headaches have been poorly documented, and research is needed to establish better criteria for causation.

## **Introduction**

Headache is a common accompaniment of systemic viral infections such as influenza. It is also common with sepsis; more rarely it may accompany other systemic infections.

In intracranial infections headache is usually the first and the most frequently encountered symptom. Occurrence of a new type of headache which is diffuse, pulsating and associated with a general feeling of illness and/or fever should direct attention towards an intracranial infection even in the absence of a stiff neck. Unfortunately, there are no good prospective studies of the headaches associated with intracranial infection and precise diagnostic criteria for these subtypes of headache cannot be developed in all cases.

### **9.1 Headache attributed to intracranial infection**

#### **9.1.1 Headache attributed to bacterial meningitis**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. diffuse pain
  - 2. intensity increasing to severe
  - 3. associated with nausea, photophobia and/or phonophobia
- B. Evidence of bacterial meningitis from examination of CSF
- C. Headache develops during the meningitis
- D. One or other of the following:
  - 1. headache resolves within 3 months after relief from meningitis
  - 2. headache persists but 3 months have not yet passed since relief from meningitis

*Comments:*

Headache is the commonest and may be the first symptom of bacterial meningitis. Headache is a key symptom of meningeal syndrome or meningism consisting usually of headache, neck stiffness and photophobia.

A variety of microorganisms may cause primary or secondary meningitis. Direct stimulation of the sensory terminals located in the meninges by 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 sensitisation and neuropeptide release.

When headache persists after 3 months, code as 9.4.1 *Chronic post-bacterial meningitis headache*.

#### **9.1.2 Headache attributed to lymphocytic meningitis**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:

1. acute onset
  2. severe intensity
  3. associated with nuchal rigidity, fever, nausea, photophobia and/or phonophobia
- B. Examination of CSF shows lymphocytic pleocytosis, mildly elevated protein and normal glucose<sup>1</sup>
- C. Headache develops in close temporal association to meningitis
- D. Headache resolves within 3 months<sup>2</sup> after successful treatment or spontaneous remission of infection

*Notes:*

1. Virus, borrelia, listeria, fungus, tuberculosis or other infective agent(s) may be identified by appropriate methods.
2. Headache usually resolves within 1 week.

*Comments:*

Headache, fever, photophobia and nuchal rigidity are the main symptoms of lymphocytic or non-bacterial meningitis and headache may remain as the main symptom throughout the course of the disease.

Headache can appear with intracranial infection but also in systemic inflammation. Since the signs of systemic inflammation associated with headache do not necessarily mean meningitis or encephalitis, diagnosis of lymphocytic meningitis must be confirmed by CSF examination.

Enteroviruses account for most viral causes. Herpes simplex, adenovirus, mumps and others may also be responsible.

### **9.1.3 Headache attributed to encephalitis**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
1. diffuse pain
  2. intensity increasing to severe
  3. associated with nausea, photophobia or phonophobia
- B. Neurological symptoms and signs of acute encephalitis, and diagnosis confirmed by EEG, CSF examination, neuroimaging and/or other laboratory investigations<sup>1</sup>
- C. Headache develops during encephalitis
- D. Headache resolves within 3 months after successful treatment or spontaneous remission of the infection

*Note:*

1. PCR method gives the specific diagnosis.

*Comments:*

The causes of headache include both meningeal irritation and increased intracranial pressure. Head pain may also be a systemic reaction to the toxic products of the infecting agent(s). Headache may occur early and be the only clinical symptom of encephalitis.

Herpes simplex virus, arbovirus and mumps are known causes of encephalitis. Except for HSV encephalitis (in which 95% of cases are identifiable with PCR), the causative virus is identified in fewer than half of cases of encephalitis.

#### **9.1.4 Headache attributed to brain abscess**

##### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. constant pain
  - 3. intensity gradually increasing to moderate or severe
  - 4. aggravated by straining
  - 5. accompanied by nausea
- B. Neuroimaging and/or laboratory evidence of brain abscess
- C. Headache develops during active infection
- D. Headache resolves within 3 months after successful treatment of the abscess

##### *Comments:*

Direct compression and irritation of the meningeal or arterial structures and increased intracranial pressure are the mechanisms for causing headache.

The most common organisms causing brain abscess include streptococcus, staphylococcus aureus, bacteroides species and enterobacter. Predisposing factors include infections of paranasal sinuses, ears, jaws, teeth or lungs.

#### **9.1.5 Headache attributed to subdural empyema**

##### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. unilateral or much more intense on one side
  - 2. associated with tenderness of the skull
  - 3. accompanied by fever
  - 4. accompanied by stiffness of the neck
- B. Neuroimaging and/or laboratory evidence of subdural empyema
- C. Headache develops during active infection and is localised to or maximal at the site of the empyema
- D. Headache resolves within 3 months after successful treatment of the empyema

##### *Comments:*

Headache is caused by meningeal irritation, increased intracranial pressure and/or fever.

Subdural empyema is often secondary to sinusitis or otitis media. It may also be a complication of meningitis. Early diagnosis is best made by CT or MRI.

## 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*.

### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. diffuse pain
  - 2. intensity increasing to moderate or severe
  - 3. associated with fever, general malaise or other symptoms of systemic infection
- B. Evidence of systemic infection
- C. Headache develops during the systemic infection
- D. Headache resolves within 72 hours after effective treatment 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 systemic symptoms. Nevertheless, some systemic infections, particularly influenza, have headache as a prominent symptom along with fever and other symptoms. In other cases, systemic infection is accompanied by meningitis or encephalitis, and the headache should be coded to these disorders.

The great variability in their propensity for causing headache indicates that systemic infections do not have this effect simply through fever. The mechanisms causing headache include direct effects of the microorganisms themselves. In infectious disease, headache commonly coexists with fever and may be dependent on it, but headache can occur in the absence of fever. The presence or absence of fever may be used in the differential classification of headache. The exact cause of headache by fever is not elucidated. Some infective microorganisms may influence brainstem nuclei which release substances to cause headache, or endotoxins may activate inducible NOS causing production of nitric oxide (NO). The exact nature of these mechanisms remains to be investigated.

### 9.2.1 *Headache attributed to systemic bacterial infection*

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria for 9.2 *Headache attributed to systemic infection*
- B. Laboratory investigation discloses the inflammatory reaction and identifies the organism

#### *Comment:*

Some infective agents have a particular tropism for the central nervous system. They may activate brainstem nuclei where release of toxins induces headache mechanisms.

### 9.2.2 *Headache attributed to systemic viral infection*

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria for 9.2 *Headache attributed to systemic infection*
- B. Clinical and laboratory (serology and/or PCR molecular) diagnosis of viral infection

### 9.2.3 Headache attributed to other systemic infection

#### *Diagnostic criteria:*

- A. Headache fulfilling criteria for 9.2 *Headache attributed to systemic infection*
- B. Clinical and laboratory (serology, microscopy, culture or PCR molecular) diagnosis of infection other than bacterial or viral

### 9.3 Headache attributed to HIV/AIDS

#### *Coded elsewhere:*

Headache attributed to a specific supervening infection is coded according to that infection.

#### *Diagnostic criteria:*

- A. Headache with variable mode of onset, site and intensity<sup>1</sup> fulfilling criteria C and D
- B. Confirmation of HIV infection and/or of the diagnosis of AIDS, and of the presence of HIV/AIDS-related pathophysiology likely to cause headache<sup>2</sup>, by neuroimaging, CSF examination, EEG and laboratory investigations
- C. Headache develops in close temporal relation to the HIV/AIDS-related pathophysiology
- D. Headache resolves within 3 months after the infection subsides

#### *Notes:*

- 1. Headache as a symptom of HIV infection is dull and bilateral. Otherwise, the onset, site and intensity of headache vary according to the HIV/AIDS-related conditions (such as meningitis, encephalitis or systemic infection) that are present.
- 2. See *Comments*.

#### *Comments:*

Dull bilateral headache may be a part of the symptomatology of HIV infection. Headache may also be attributed to aseptic meningitis during HIV infection (but not exclusively in the AIDS stages) and to secondary meningitis or encephalitis associated with opportunistic infections or neoplasms (which mostly occur in the AIDS stages). The most common intracranial infections in HIV/AIDS are toxoplasmosis and cryptococcal meningitis.

Headache occurring in patients with HIV/AIDS but attributed to a specific supervening infection is coded to that infection.

### 9.4 Chronic post-infection headache

#### 9.4.1 *Chronic post-bacterial meningitis headache*

#### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. diffuse continuous pain
  - 2. associated with dizziness
  - 3. associated with difficulty in concentrating and/or loss of memory
- B. Evidence of previous intracranial bacterial infection from CSF examination or neuroimaging

- C. Headache is a direct continuation of 9.1.1 *Headache attributed to bacterial meningitis*
- D. Headache persists for >3 months after resolution of infection

*Comments:*

A reported 32% of survivors of bacterial meningitis suffer from persistent headache (Bohr *et al*, 1983).

There is no evidence for persistent headache following other infections, but criteria for A9.4.2 *Chronic post-non-bacterial infection headache* are in the appendix. More research is needed.

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## 10. HEADACHE ATTRIBUTED TO DISORDER OF HOMOEOSTASIS

- 10.1 Headache attributed to hypoxia and/or hypercapnia
  - 10.1.1 High-altitude headache
  - 10.1.2 Diving headache
  - 10.1.3 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
  - 10.3.5 Headache attributed to eclampsia
  - 10.3.6 Headache attributed to acute pressor response to an exogenous agent
- 10.4 Headache attributed to hypothyroidism
- 10.5 Headache attributed to fasting
- 10.6 Cardiac cephalgia
- 10.7 Headache attributed to other disorder of homoeostasis

*Coded elsewhere:*

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

### General comment

*Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to a disorder of homoeostasis, it is coded as a secondary headache attributed to that disorder. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to a disorder of homoeostasis, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the disorder of homoeostasis. Factors that support adding the latter diagnosis are: a very close temporal relation to the disorder of homoeostasis, a marked worsening of the pre-existing headache, very good evidence that the disorder of homoeostasis can aggravate the primary headache and, finally, improvement or resolution of the headache after relief from the disorder of homoeostasis.

*Definite, probable or chronic?*

A diagnosis of *Headache attributed to disorder of homoeostasis* usually becomes definite only when the headache resolves or greatly improves after effective treatment or spontaneous remission of the disorder. If this disorder cannot be treated effectively or does not remit spontaneously, or when there has been insufficient time for this to happen, a diagnosis of *Headache probably attributed to disorder of homoeostasis* is usually applied.

The alternative, when the disorder of homoeostasis is effectively treated or remits spontaneously but headache does not resolve or markedly improve after 3 months, is a diagnosis of A10.8 *Chronic*

*post-homoeostasis disorder headache*. This is described only in the appendix as such headaches have been poorly documented, and research is needed to establish better criteria for causation.

## **Introduction**

Headache disorders described here were previously referred to as *Headache associated with metabolic or systemic disease*. However, *Headache attributed to disorder of homoeostasis* was felt to capture more accurately the true nature of these headache disorders. Headaches caused by significant disturbances in arterial pressure and by myocardial ischaemia are now included in this section. In addition, disorders of homoeostatic mechanisms affecting a variety of organ systems, including altered arterial blood gases, volume disturbances as in dialysis and disorders of endocrine function, are covered here. Headache attributed to fasting is also included.

### **10.1 Headache attributed to hypoxia and/or hypercapnia**

#### *Comments:*

Headache occurs within 24 hours after acute onset of hypoxia with PaO<sub>2</sub> <70 mm Hg or in chronically hypoxic patients with PaO<sub>2</sub> persistently at or below this level.

It is often difficult to separate the effects of hypoxia and hypercapnia.

#### **10.1.1 High-altitude headache**

##### *Diagnostic criteria:*

- A. Headache with at least two of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. frontal or frontotemporal
  - 3. dull or pressing quality
  - 4. mild or moderate intensity
  - 5. aggravated by exertion, movement, straining, coughing or bending
- B. Ascent to altitude above 2500 m
- C. Headache develops within 24 hours after ascent
- D. Headache resolves within 8 hours after descent

##### *Comments:*

Headache is a frequent complication of ascent to altitude – occurring in more than 80% of cases.

10.1.1 *High-altitude headache* appears to be independent of an individual's previous history of headache, although patients with migraine may describe more severe headache that resembles their typical migraine attacks.

Acute mountain sickness (AMS) consists of at least moderate headache combined with one or more of nausea, anorexia, fatigue, dizziness and sleep disturbances. Acetazolamide (125 mg, two or three times daily) may reduce susceptibility to acute mountain sickness. Preventative strategies include allowing two days of acclimatisation prior to engaging in strenuous exercise at high altitudes, avoiding alcohol, and liberalising fluid intake. Most high-altitude headaches respond to simple analgesics such as paracetamol (acetaminophen) or ibuprofen.

### 10.1.2 Diving headache

*Coded elsewhere:*

1. *Migraine*, 2. *Tension-type headache*, 4.3 *Primary exertional headache*, 11.2.1 *Cervicogenic headache*, 13.6 *Supraorbital neuralgia*, 13.10 *External compression headache* and 13.11 *Cold-stimulus headache* precipitated by diving are coded as those disorders.

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Diving to depth below 10 m
- C. Headache develops during diving and is accompanied by at least one of the following symptoms of CO<sub>2</sub> intoxication in the absence of decompression illness:
  - 1. light-headedness
  - 2. mental confusion
  - 3. dyspnoea
  - 4. flushed feeling in the face
  - 5. motor incoordination
- D. Headache resolves within 1 hour after treatment with 100% O<sub>2</sub>

*Comment:*

Hypercapnia (arterial PCO<sub>2</sub> >50 mm Hg) is known to cause relaxation of cerebrovascular smooth muscle and lead to vasodilatation and increased intracranial pressure. There is some evidence that hypercapnia in the absence of hypoxia is associated with headache. The best clinical example of headache attributed to hypercapnia occurs in divers. Carbon dioxide may accumulate in a diver who intentionally holds his or her breath intermittently (skip breathing) in a mistaken attempt to conserve air, or takes shallow breaths to minimise 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 more than 60 mm Hg. Diving headache usually intensifies during the decompression phase of the dive or upon resurfacing.

Mild non-specific headache is also common in divers with decompression illness, and may be associated with musculoskeletal pain and, in more serious cases, with focal neurological and/or respiratory symptoms, loss of consciousness and/or cognitive deficits.

Headache in divers can also occur as a result of carbon monoxide intoxication which rarely contaminates divers' compressed-air supply if the air intake system is positioned in such a way as to gather improperly directed combustion-engine exhaust. Such headache is coded as 8.1.3 *Carbon monoxide-induced headache*.

Migraine, tension-type headache, primary exertional headache, cervicogenic headache, supraorbital neuralgia, external compression headache and cold-stimulus headache can occur during a dive, but diving in these instances should be considered a precipitating factor rather than the cause.

### 10.1.3 Sleep apnoea headache

#### Diagnostic criteria:

- A. Recurrent headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. occurs on >15 days per month
  - 2. bilateral, pressing quality and not accompanied by nausea, photophobia or phonophobia
  - 3. each headache resolves within 30 minutes
- B. Sleep apnoea (Respiratory Disturbance Index  $\geq 5$ ) demonstrated by overnight polysomnography
- C. Headache is present upon awakening
- D. Headache ceases within 72 hours, and does not recur, after effective treatment of sleep apnoea

#### Comments:

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 which occurs in a variety of primary and secondary headache disorders, in sleep-related respiratory disorders other than sleep apnoea (eg, Pickwickian syndrome, chronic obstructive pulmonary disorder), and in other primary sleep disorders such as periodic leg movements of sleep. A definitive diagnosis of 10.1.3 *Sleep apnoea headache* requires overnight polysomnography.

It is unclear whether the mechanism of 10.1.3 *Sleep apnoea headache* is related to hypoxia, hypercapnia or disturbance in sleep.

## 10.2 Dialysis headache

#### Diagnostic criteria:

- A. At least 3 attacks of acute headache fulfilling criteria C and D
- B. Patient is on haemodialysis
- C. Headache develops during at least half of haemodialysis sessions
- D. Headache resolves within 72 hours after each haemodialysis session and/or ceases altogether after successful transplantation

#### Comment:

Headache commonly occurs in association with hypotension and dialysis disequilibrium syndrome. The disequilibrium syndrome may begin as headache and then progress to obtundation and finally coma, with or without seizures. This syndrome is relatively rare and may be prevented by changing dialysis parameters.

As caffeine is rapidly removed by dialysis, 8.4.1 *Caffeine-withdrawal headache* should be considered in patients who consume large quantities of caffeine.

### 10.3 Headache attributed to arterial hypertension

#### *Comment:*

Mild (140-159/90-99 mm Hg) or moderate (160-179/100-109 mm Hg) chronic arterial hypertension does not appear to *cause* headache. Whether moderate hypertension *predisposes* to headache at all remains controversial, but there is little evidence that it does. Ambulatory blood pressure monitoring in patients with mild and moderate hypertension has shown no convincing relationship between blood pressure fluctuations over a 24-hour period and presence or absence of headache.

#### **10.3.1 Headache attributed to pheochromocytoma**

##### *Diagnostic criteria:*

- A. Intermittent discrete attacks of headache accompanied by at least one of the following and fulfilling criteria C and D:
  - 1. sweating
  - 2. palpitations
  - 3. anxiety
  - 4. pallor
- B. Pheochromocytoma demonstrated by biochemical investigations, imaging and/or surgery
- C. Headache develops concomitantly with abrupt rise in blood pressure
- D. Headache resolves or markedly improves within 1 hour of normalisation of blood pressure

##### *Comments:*

Paroxysmal headache occurs in 51-80% of patients with pheochromocytoma. It is often severe, frontal or occipital and is generally described as either pulsating or steady in quality. An important feature of the headache is its short duration: <15 minutes in 50% and <1 hour in 70% of patients. Other features include apprehension and/or anxiety, often with a sense of impending death, tremor, visual disturbances, abdominal or chest pain, nausea, vomiting and occasionally paraesthesia. The face can blanch or flush during the attack.

The diagnosis is established by the demonstration of increased excretion of catecholamines or catecholamine metabolites, and can usually be secured by analysis of a single 24-hour urine sample collected when the patient is hypertensive or symptomatic.

When hypertensive encephalopathy is present, headache is coded as 10.3.3 *Headache attributed to hypertensive encephalopathy*. When the diagnosis of pheochromocytoma has not yet been made, and hypertensive encephalopathy is not present, patients may meet the diagnostic criteria for 10.3.2 *Headache attributed to hypertensive crisis without hypertensive encephalopathy*.

#### **10.3.2 Headache attributed to hypertensive crisis without hypertensive encephalopathy**

##### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. pulsating quality
  - 3. precipitated by physical activity

- B. Hypertensive crisis defined as a paroxysmal rise in systolic (to >160 mm Hg) and/or diastolic (to >120 mm Hg) blood pressure but no clinical features of hypertensive encephalopathy
- C. Headache develops during hypertensive crisis
- D. Headache resolves within 1 hour after normalisation of blood pressure
- E. Appropriate investigations have ruled out vasopressor toxins or medications as causative factors

*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***

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  1. diffuse pain
  2. pulsating quality
  3. aggravated by physical activity
- B. Persistent blood pressure elevation to >160/100 mm Hg with at least two of the following:
  1. confusion
  2. reduced level of consciousness
  3. visual disturbances (other than those of typical migraine aura) including blindness
  4. seizures
- C. Headache develops in close temporal relation to blood pressure elevation
- D. Headache resolves within 3 months after effective treatment and control of hypertension
- E. Other causes of the neurological symptoms have been excluded

*Comments:*

Hypertensive encephalopathy 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 mm Hg, and by grade 3 or 4 hypertensive retinopathy (Keith-Wagner classification), previously normotensive individuals may develop signs of encephalopathy with blood pressures as low as 160/100. Hypertensive retinopathy may not be present at the time of clinical presentation.

Any cause of hypertension, including phaeochromocytoma and ingestion of vasopressor toxins, can lead to hypertensive encephalopathy.

### ***10.3.4 Headache attributed to pre-eclampsia***

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:

1. bilateral
  2. pulsating quality
  3. aggravated by physical activity
- B. Pregnancy or puerperium (up to 4 weeks post-partum), and pre-eclampsia defined by both of the following:
1. hypertension (>140/90 mm Hg) documented on two blood pressure readings at least 4 hours apart
  2. urinary protein excretion >0.3 g per 24 hours
- C. Headache develops during periods of high blood pressure
- D. Headache resolves within 7 days after effective treatment of hypertension
- E. Appropriate investigations have ruled out vasopressor toxins, medications or phaeochromocytoma as causative factors

*Comment:*

A placenta appears essential for the development of pre-eclampsia. Pre-eclampsia is a multi-system disorder with various forms. In addition to hypertension and proteinuria, tissue oedema, thrombocytopenia and abnormalities in liver function can occur. Pre-eclampsia appears to involve a strong maternal inflammatory response, with broad immunological systemic activity.

### ***10.3.5 Headache attributed to eclampsia***

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
1. bilateral
  2. pulsating quality
  3. aggravated by physical activity
- B. Pregnancy or puerperium (up to 4 weeks post-partum), and eclampsia defined by all of the following:
1. hypertension (>140/90 mm Hg) documented on two blood pressure readings at least 4 hours apart
  2. urinary protein excretion >0.3 g per 24 hours
  3. a seizure has occurred
- C. Headache develops during periods of high blood pressure
- D. Headache resolves within 7 days after effective treatment of hypertension
- E. Appropriate investigations have ruled out vasopressor toxins, medications or phaeochromocytoma as causative factors
- F. Stroke has been excluded

*Comment:*

Case reports indicate that eclampsia can occur in the puerperium as well as during pregnancy.

### ***10.3.6 Headache attributed to acute pressor response to an exogenous agent***

*Coded elsewhere:*

8.1.6 Cocaine-induced headache.

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. An appropriate agent or toxin has been administered or ingested and an acute rise in blood pressure has occurred
- C. Headache develops in close temporal relation to the acute rise in blood pressure
- D. Headache resolves within 24 hours after normalisation of blood pressure
- E. No other mechanism for the headache is apparent

*Comments:*

Apart from cocaine, agents that can produce acute elevations of blood pressure include sympathomimetics and amphetamines, and monoamine oxidase inhibitors when interactions with tyramine-containing foods occur.

There is insufficient evidence to set criteria for how large an elevation in blood pressure is required to produce headache, and this may vary from person to person. Criterion D is arbitrary, but included to increase the specificity of the diagnostic criteria.

#### **10.4 Headache attributed to hypothyroidism**

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. bilateral
  - 2. non-pulsatile
  - 3. continuous
- B. Hypothyroidism is demonstrated by appropriate investigations
- C. Headache develops within 2 months after other symptoms of hypothyroidism become evident
- D. Headache resolves within 2 months after effective treatment of hypothyroidism

*Comment:*

It has been estimated that approximately 30% of patients with hypothyroidism suffer from headache. Its mechanism is unclear. There is a female preponderance and often a history of migraine in childhood. Headache attributed to hypothyroidism is not associated with nausea or vomiting.

#### **10.5 Headache attributed to fasting**

*Coded elsewhere:*

Hypoglycaemia-induced migraine is coded according to subtype under 1. *Migraine*, with hypoglycaemia considered as a precipitating factor.

*Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. frontal location
  - 2. diffuse pain
  - 3. non-pulsating quality
  - 4. mild or moderate intensity

- B. The patient has fasted for >16 hours
- C. Headache develops during fasting
- D. Headache resolves within 72 hours after resumption of food intake

*Comments:*

Headache with fasting is significantly more common in individuals with a prior history of headache. In those individuals with a prior history of migraine, the headache may resemble 1.1 *Migraine without aura*.

The likelihood of headache developing as a result of a fast increases with the duration of the fast.

The headache associated with 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. Fasting headache 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. Well-controlled studies are needed to demonstrate a causal relationship, if one exists.

## 10.6 Cardiac cephalgia

*Diagnostic criteria:*

- A. Headache, which may be severe, aggravated by exertion and accompanied by nausea and fulfilling criteria C and D
- B. Acute myocardial ischaemia has occurred
- C. Headache develops concomitantly with acute myocardial ischaemia
- B. Headache resolves and does not recur after effective medical therapy for myocardial ischaemia or coronary revascularisation

*Comment:*

Diagnosis must include careful documentation of headache and simultaneous cardiac ischaemia during treadmill or nuclear cardiac stress testing. Failure to recognise and correctly diagnose 10.6 *Cardiac cephalgia* can have grave consequences. Therefore, distinguishing this disorder from 1.1 *Migraine without aura* is of crucial importance, particularly since vasoconstrictor medications (eg, 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 disorders 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 homeostasis

*Diagnostic criteria:*

- A. Headache fulfilling criteria C and D
- B. Evidence of a disorder of homeostasis other than those described above
- C. Headache develops within 2 months after onset of the disorder, and other evidence exists that the disorder can cause headache
- D. Headache resolves within 3 months after relief from the disorder of homeostasis

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## 11. HEADACHE OR FACIAL PAIN ATTRIBUTED TO DISORDER OF CRANIUM, NECK, EYES, EARS, NOSE, SINUSES, TEETH, MOUTH OR OTHER FACIAL OR CRANIAL STRUCTURES

- 11.1 Headache attributed to disorder of cranial bone
- 11.2 Headache attributed to disorder of neck
  - 11.2.1 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 eyes
  - 11.3.1 Headache attributed to acute glaucoma
  - 11.3.2 Headache attributed to refractive errors
  - 11.3.3 Headache attributed to heterophoria or heterotropia (latent or manifest squint)
  - 11.3.4 Headache attributed to ocular inflammatory disorder
- 11.4 Headache attributed to disorder of ears
- 11.5 Headache attributed to rhinosinusitis
- 11.6 Headache attributed to disorder of teeth, jaws or related structures
- 11.7 Headache or facial pain attributed to temporomandibular joint (TMJ) disorder
- 11.8 Headache attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structures

### *Coded elsewhere:*

Headaches that are due to head or neck trauma are classified under 5. *Headache attributed to head and/or neck trauma*. Neuralgiform headaches are classified under 13. *Cranial neuralgias and central causes of facial pain*.

### **General comment**

#### *Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to a craniocervical disorder, it is coded as a secondary headache attributed to that disorder. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to a craniocervical disorder, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the craniocervical disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the craniocervical disorder, a marked worsening of the pre-existing headache, very good evidence that the craniocervical disorder can aggravate the primary headache and, finally, improvement or resolution of the headache after relief from the craniocervical disorder.

#### *Definite, probable or chronic?*

A diagnosis of *Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures* usually becomes definite only when the headache resolves or greatly improves after effective treatment or spontaneous remission of the craniocervical disorder. If this disorder cannot be treated effectively or does not remit spontaneously, or when there has been insufficient time for this to happen, a diagnosis of *Headache probably attributed to the [specified] craniocervical disorder* is usually applied.

If the craniocervical disorder is effectively treated or remits spontaneously but headache does not resolve or markedly improve after 1 month, the persisting headache has other mechanisms. Nevertheless, A11.9 *Chronic post-craniocervical disorder headache* is described in the appendix. Headaches meeting these criteria exist but have been poorly studied and the appendix entry is intended to stimulate further research into such headaches and their mechanisms.

## **Introduction**

Disorders of the cervical spine and of other structures of the neck and head have not infrequently been regarded as the commonest causes of headache, since many headaches originate from the cervical, nuchal or occipital regions or are localised there. Moreover, degenerative changes in the cervical spine can be found in virtually all people over 40 years of age. The localisation of pain and the x-ray detection of degenerative changes have been plausible reasons for regarding the cervical spine as the most frequent cause of headaches. However, large-scale controlled studies have shown that such changes are just as widespread among individuals who do not suffer from headaches. Spondylosis or osteochondrosis cannot therefore be seen as the explanation of headaches. A similar situation applies to other widespread disorders: chronic sinusitis, temporomandibular joint disorders and refractive errors of the eyes.

Without specific criteria it would be possible for virtually any type of headache to be classified as *Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures*, and this problem existed in the past. 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 subforms, but rather to establish specific causal relationships between headaches and facial pain and the disorders of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth and other facial or cranial structures where these exist. For this reason it has been necessary to identify strict specific operational criteria for cervicogenic headache and other causes of headache described in this chapter. It is not possible here to take account of diagnostic tests that are unconfirmed or for which quality criteria have not been investigated. Instead the aim of the revised criteria is to motivate as a future task the development of reliable and valid operational tests to establish specific causal relationships between headaches and craniocervical disorders that are currently available only to a very limited extent.

Headache disorders attributed to causes included here for the first time are 11.2.3 *Headache attributed to craniocervical dystonia* and 11.3.4 *Headache attributed to ocular inflammatory disorders*.

### **11.1 Headache attributed to disorder of cranial bone**

*Diagnostic criteria:*

- A. Pain in one or more regions of the head or face fulfilling criteria C and D
- B. Clinical, laboratory and/or imaging evidence of a lesion within the cranial bone known to be, or generally accepted as, a valid cause of headache<sup>1</sup>
- C. Pain develops in close temporal relation to and is maximal over the bone lesion
- D. Pain resolves within 3 months after successful treatment of the bone lesion

*Note:*

1. Most disorders of the skull (*eg*, 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 neck**

*Comment:*

Headache attributed to disorder of neck but not fulfilling the criteria for any of 11.2.1 *Cervicogenic headache*, 11.2.2 *Retropharyngeal tendonitis* or 11.2.3 *Craniocervical dystonia* is not sufficiently validated.

### **11.2.1 Cervicogenic headache**

*Previously used term:*

Cervical headache

*Coded elsewhere:*

Headache causally associated with cervical myofascial tender spots is 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*.

*Diagnostic criteria:*

- A. Pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling criteria C and D
- B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache<sup>1</sup>
- C. Evidence that the pain can be attributed to the neck disorder or lesion based on at least one of the following:
  1. demonstration of clinical signs that implicate a source of pain in the neck<sup>2</sup>
  2. abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls<sup>3</sup>
- D. Pain resolves within 3 months after successful treatment of the causative disorder or lesion

*Notes:*

1. Tumours, fractures, infections and rheumatoid arthritis of the upper cervical spine have not been validated formally as causes of headache, but are nevertheless accepted as valid causes when demonstrated to be so in individual cases. Cervical spondylosis and osteochondritis are NOT accepted as valid causes fulfilling criterion B. When myofascial tender spots are the cause, the headache should be coded under 2. *Tension-type headache*.
2. Clinical signs acceptable for criterion C1 must have demonstrated reliability and validity. The future task is the identification of such reliable and valid operational tests. Clinical features such as neck pain, focal neck tenderness, history of neck trauma, mechanical exacerbation of pain, unilaterality, coexisting shoulder pain, reduced range of motion in the

neck, nuchal onset, nausea, vomiting, photophobia *etc* are not unique to cervicogenic headache. These may be features of cervicogenic headache, but they do not define the relationship between the disorder and the source of the headache.

3. Abolition of headache means complete relief of headache, indicated by a score of zero on a visual analogue scale (VAS). Nevertheless, acceptable as fulfilling criterion C2 is  $\geq 90\%$  reduction in pain to a level of  $< 5$  on a 100-point VAS.

### **11.2.2 Headache attributed to retropharyngeal tendonitis**

#### *Diagnostic criteria:*

- A. Unilateral or bilateral non-pulsating pain in the back of the neck, radiating to the back of the head or to the whole head and fulfilling criteria C and D
- B. Swollen prevertebral soft tissues, in adults measuring  $> 7$  mm at the level between C1 and C4 (special x-ray technique may be required)
- C. Pain is aggravated severely by bending the head backwards
- D. Pain is alleviated within 2 weeks of treatment with non-steroidal anti-inflammatory drugs in their recommended doses

#### *Comments:*

Body temperature and erythrocyte sedimentation rate (ESR) are usually elevated. Although retroflexion of the neck most consistently aggravates pain, this also usually happens with rotation and swallowing. The transverse processes of the upper three vertebrae are usually tender to palpation.

In several cases amorphous calcific material has been aspirated from the swollen prevertebral tissues. Thin calcification in prevertebral tissues is best seen on CT.

Upper carotid dissection should be ruled out.

### **11.2.3 Headache attributed to craniocervical dystonia**

#### *Diagnostic criteria:*

- A. Sensation of cramp, tension or pain in the neck, radiating to the back of the head or to the whole head and fulfilling criteria C and D
- B. Abnormal movements or defective posture of neck or head due to muscular hyperactivity
- C. Evidence that pain is attributed to muscular hyperactivity based on at least one of the following:
  1. demonstration of clinical signs that implicate a source of pain in the hyperactive muscle (*eg*, pain is precipitated or exacerbated by muscle contraction, movements, sustained posture or external pressure)
  2. simultaneous onset of pain and muscular hyperactivity
- D. Pain resolves within 3 months after successful treatment of the causative disorder

#### *Comment:*

Focal dystonias of the head and neck accompanied by pain are pharyngeal dystonia, spasmodic torticollis, mandibular dystonia, lingual dystonia and a combination of the cranial and cervical

dystonias (segmental craniocervical dystonia). Pain is caused by local contractions and secondary changes.

### **11.3 Headache attributed to disorder of eyes**

#### ***11.3.1 Headache attributed to acute glaucoma***

*Diagnostic criteria:*

- A. Pain in the eye and behind or above it, fulfilling criteria C and D
- B. Raised intraocular pressure, with at least one of the following:
  - 1. conjunctival injection
  - 2. clouding of cornea
  - 3. visual disturbances
- C. Pain develops simultaneously with glaucoma
- D. Pain resolves within 72 hours of effective treatment of glaucoma

#### ***11.3.2 Headache attributed to refractive errors***

*Diagnostic criteria:*

- A. Recurrent mild headache, frontal and in the eyes themselves, fulfilling criteria C and D
- B. Uncorrected or miscorrected refractive error (*eg*, hyperopia, astigmatism, presbyopia, wearing of incorrect glasses)
- C. Headache and eye pain first develop in close temporal relation to the refractive error, are absent on awakening and aggravated by prolonged visual tasks at the distance or angle where vision is impaired
- D. Headache and eye pain resolve within 7 days, and do not recur, after full correction of the refractive error

#### ***11.3.3 Headache attributed to heterophoria or heterotropia (latent or manifest squint)***

*Diagnostic criteria:*

- A. Recurrent non-pulsatile mild-to-moderate frontal headache fulfilling criteria C and D
- B. Heterophoria or heterotropia has been demonstrated, with at least one of the following:
  - 1. intermittent blurred vision or diplopia
  - 2. difficulty in adjusting focus from near to distant objects or *vice versa*
- C. At least one of the following:
  - 1. headache develops or worsens during a visual task, especially one that is tiring
  - 2. headache is relieved or improved on closing one eye
- D. Headache resolves within 7 days, and does not recur, after appropriate correction of vision

#### ***11.3.4 Headache attributed to ocular inflammatory disorder***

*Diagnostic criteria:*

- A. Pain in the eye and behind or around it, fulfilling criteria C and D
- B. Ocular inflammation diagnosed by appropriate investigations

- C. Headache develops during inflammation
- D. Headache resolves within 7 days after relief of the inflammatory disorder

*Comment:*

Ocular inflammation takes many forms, and may be categorised variously by anatomical site (*ie*, iritis, cyclitis, choroiditis), by course (acute, subacute, chronic), by presumed cause (infectious agents that are endogenous or exogenous, lens-related, traumatic), or by type of inflammation (granulomatous, non-granulomatous).

#### **11.4 Headache attributed to disorder of ears**

*Coded elsewhere:*

Headache attributed to acoustic neuroma is coded as 7.4.2 *Headache attributed directly to neoplasm*. Headache attributed to a lesion, not of the ear, giving rise to referred otalgia is coded according to the site and/or nature of the lesion.

*Diagnostic criteria:*

- A. Headache accompanied by otalgia and fulfilling criteria C and D
- B. Structural lesion of the ear diagnosed by appropriate investigations
- C. Headache and otalgia develop in close temporal relation to the structural lesion
- D. Headache and otalgia resolve simultaneously with remission or successful treatment of the structural lesion

*Comment:*

There is no evidence that any pathology of the ear can cause *headache* without concomitant otalgia. Structural lesions of the pinna, external auditory canal, tympanic membrane or middle ear may give rise to *primary otalgia* associated with headache.

However, only about 50% of all cases of earache are due to structural lesions of the external or middle ear. Disorders outside this region may lead to referred otalgia as a result of radiation of pain into the ear region. Sensory fibres of the fifth, seventh, ninth and tenth cranial nerves project into the auricle, external auditory canal, tympanic membrane and middle ear. For this reason referred pain from remote structural lesions in any of the anatomical regions to which these nerves project can be felt as *referred otalgia*. Since these are not disorders of the ear they are coded elsewhere according to the site and/or nature of the lesion(s).

#### **11.5 Headache attributed to rhinosinusitis**

*Coded elsewhere:*

“Sinus headaches”

*Diagnostic criteria:*

- A. Frontal headache accompanied by pain in one or more regions of the face, ears or teeth and fulfilling criteria C and D
- B. Clinical, nasal endoscopic, CT and/or MRI imaging and/or laboratory evidence of acute or acute-on-chronic rhinosinusitis<sup>1;2</sup>

- C. Headache and facial pain develop simultaneously with onset or acute exacerbation of rhinosinusitis
- D. Headache and/or facial pain resolve within 7 days after remission or successful treatment of acute or acute-on-chronic rhinosinusitis

*Notes:*

- 1. Clinical evidence may include purulence in the nasal cavity, nasal obstruction, hyposmia/anosmia and/or fever.
- 2. *Chronic sinusitis* is not validated as a cause of headache or facial pain unless relapsing into an acute stage.

*Comments:*

Other conditions that are often considered to induce headache are not sufficiently validated as causes of headache. These include deviation of nasal septum, hypertrophy of turbinates, atrophy of sinus membranes and mucosal contact. The last, however, is defined in the appendix under A11.5.1 *Mucosal contact point headache*.

Migraine and tension-type headache are often confused with 11.5 *Headache attributed to rhinosinusitis* because of similarity in location of the headache. A group of patients can be identified who have all of the features of 1.1 *Migraine without aura* and, additionally, concomitant clinical features such as facial pain, nasal congestion and headache triggered by weather changes. None of these patients have purulent nasal discharge or other features diagnostic of acute rhinosinusitis. Therefore it is necessary to differentiate 11.5 *Headache attributed to rhinosinusitis* from so-called “sinus headaches”, a commonly-made but non-specific diagnosis. Most such cases fulfil the criteria for 1.1 *Migraine without aura*, with headache either accompanied by prominent autonomic symptoms in the nose or triggered by nasal changes.

## **11.6 Headache attributed to disorder of teeth, jaws or related structures**

*Diagnostic criteria:*

- A. Headache accompanied by pain in the teeth and/or jaw(s) and fulfilling criteria C and D
- B. Evidence of disorder of teeth, jaws or related structures
- C. Headache and pain in teeth and/or jaw(s) develop in close temporal relation to the disorder
- D. Headache and pain in teeth and/or jaw(s) resolve within 3 months after successful treatment of the disorder

*Comment:*

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

## **11.7 Headache or facial pain attributed to temporomandibular joint (TMJ) disorder**

*Diagnostic criteria:*

- A. Recurrent pain in one or more regions of the head and/or face fulfilling criteria C and D
- B. X-ray, MRI and/or bone scintigraphy demonstrate TMJ disorder

- C. Evidence that pain can be attributed to the TMJ disorder, based on at least one of the following:
  1. pain is precipitated by jaw movements and/or chewing of hard or tough food
  2. reduced range of or irregular jaw opening
  3. noise from one or both TMJs during jaw movements
  4. tenderness of the joint capsule(s) of one or both TMJs
- D. Headache resolves within 3 months, and does not recur, after successful treatment of the TMJ disorder

*Comment:*

Pain from the temporomandibular joint or related tissues is common. It is due to the so-called temporomandibular joint disorders (*eg*, disk displacements, osteoarthritis, joint hypermobility) or rheumatoid arthritis, and may be associated with myofascial pain and headache.

### **11.8 Headache attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structures**

*Diagnostic criteria:*

- A. Headache, with or without pain in one or more regions of the face, fulfilling criteria C and D
- B. Evidence of disorder, other than those described above, of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
- C. Headache develops in close temporal relation to, or other evidence exists of a causal relationship with, the disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure
- D. Headache resolves within 3 months after successful treatment of the disorder

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## 12. HEADACHE ATTRIBUTED TO PSYCHIATRIC DISORDER

12.1 Headache attributed to somatisation disorder

12.2 Headache attributed to psychotic disorder

### *Coded elsewhere*

Headache attributed to substance-dependence, abuse or withdrawal, headache attributed to acute intoxication and headache attributed to medication overuse are all coded under 8. *Headache attributed to a substance or its withdrawal.*

### **General comment**

#### *Primary or secondary headache or both?*

When a new headache occurs for the first time in close temporal relation to a psychiatric disorder, it is coded as a secondary headache attributed to that disorder. This is also true if the headache has the characteristics of migraine, tension-type headache or cluster headache. When a pre-existing primary headache is made worse in close temporal relation to a psychiatric disorder, there are two possibilities, and judgment is required. The patient can either be given only the diagnosis of the pre-existing primary headache or be given both this diagnosis and the diagnosis of headache attributed to the psychiatric disorder. Factors that support adding the latter diagnosis are: a very close temporal relation to the psychiatric disorder, a marked worsening of the pre-existing headache, very good evidence that the psychiatric disorder can aggravate the primary headache and, finally, improvement or resolution of the headache after relief from the psychiatric disorder.

#### *Definite, probable or chronic?*

A diagnosis of *Headache attributed to psychiatric disorder* usually becomes definite only when the headache resolves or greatly improves after effective treatment or spontaneous remission of the psychiatric disorder. If this disorder cannot be treated effectively or does not remit spontaneously, or when there has been insufficient time for this to happen, a diagnosis of *Headache probably attributed to psychiatric disorder* is usually applied.

Chronic headache attributed to and persisting after resolution of a psychiatric disorder has not yet been described.

### **Introduction**

Overall, there is very limited evidence supporting psychiatric causes of headache. Thus, the only diagnostic categories included in this classification are those rare cases in which a headache occurs in the context of a psychiatric condition that is known to be symptomatically manifested by headache (*eg*, a patient who reports a headache associated with the delusion that a metal plate has been surreptitiously inserted into his or her head, or headache that is a manifestation of somatisation disorder). The vast majority of headaches that occur in association with psychiatric disorders are not causally related to them but instead represent comorbidity (perhaps reflecting a common biological substrate). Headache has been reported to be comorbid with a number of psychiatric disorders, including major depressive disorder, dysthymic disorder, panic disorder, generalised anxiety disorder, somatoform disorders and adjustment disorders. In such cases, both a primary headache diagnosis and the comorbid psychiatric diagnosis should be made.

However, clinical experience suggests that, in some cases, headache occurring exclusively during some common psychiatric disorders such as major depressive disorder, panic disorder, generalised anxiety disorder and undifferentiated somatoform disorder may best be considered as attributed to these disorders. To encourage further research into this area, criteria for headaches attributed to these psychiatric disorders have been included in the appendix.

A headache diagnosis should heighten the clinician's index of suspicion for major depressive disorder, panic disorder and generalised anxiety disorder, and *vice-versa*. Furthermore, evidence suggests that the presence of a comorbid psychiatric disorder tends to worsen the course of migraine and/or tension-type headache by increasing the frequency and severity of headache and making it less responsive to treatment. Thus, identification and treatment of any comorbid psychiatric condition is important for the proper management of the headache. In children and adolescents, primary headache disorders (migraine, episodic tension-type and especially chronic tension-type headache) are often comorbid with psychiatric disorder. Sleep disorder, separation-anxiety disorder, school phobia, adjustment disorder and other disorders usually first diagnosed in infancy, childhood or adolescence (particularly attention-deficit/hyperactivity disorder [ADHD]), conduct disorder, learning disorder, enuresis, encopresis, tic) should be carefully looked for and treated if found, considering their negative burden in disability and prognosis of paediatric headache.

To ascertain whether a headache should be attributed to a psychiatric disorder, it is clearly important first to determine whether or not there is a psychiatric disorder present with the headache. Optimally, this entails conducting a psychiatric evaluation for the presence of a psychiatric disorder. At a minimum, however, it is important to inquire about commonly co-morbid psychiatric symptoms such as generalised anxiety, panic attacks and depression.

## 12.1 Headache attributed to somatisation disorder

### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criterion C
- B. Presence of somatisation disorder fulfilling DSM-IV criteria:
  - 1. history of many physical complaints beginning before age 30 that occur over a period of several years and result in treatment being sought and/or in significant impairment in social, occupational or other important areas of functioning
  - 2. at least four pain symptoms, two non-pain gastrointestinal symptoms, one sexual or reproductive symptom and one pseudoneurological symptom
  - 3. after appropriate investigation, each of these symptoms cannot be fully explained by a known general medical condition or the direct effects of a substance or medication; or, if there is a related medical condition, the complaints or impairment are in excess of what would be expected from the history, examination or laboratory findings
- C. Headache is not attributed to another cause

### *Comments:*

Somatisation disorder, as defined in DSM-IV, is a polysymptomatic disorder characterised by multiple recurring pains and gastrointestinal, sexual and pseudoneurological symptoms occurring for a period of years with onset before age 30. These symptoms are, by definition, considered to be *somatoform*: that is, they are complaints of physical symptoms suggestive of, but not fully explained by, a medical condition or the direct effect(s) of a substance. In the United States, it is found predominantly in women, in whom the lifetime risk is estimated to be 2.0%, with a female/male ratio of approximately 10:1. This ratio is not as large in some other cultures (*eg*, in Greeks and Puerto Ricans).

It should be noted that the symptom requirement laid out in DSM-IV is quite extensive: a minimum of eight somatoform symptoms must have occurred over the patient's lifetime, each one severe enough to result in the seeking of medical help or the taking of a medication (prescribed or over-the-counter), or to affect the person's functioning (eg, causing missed days at work). DSM-IV has set such a high threshold in order to reduce false positives, most particularly the possibility that the "unexplained" symptoms are in fact part of a complex and as yet undiagnosed medical condition with variable symptom presentation such as multiple sclerosis or systemic lupus erythematosus. Somatoform disorders with fewer than eight symptoms are diagnosed in DSM-IV as *Undifferentiated somatoform disorder*. Because of the difficulty and uncertainty associated with this diagnosis, A12.6 *Headache attributed to undifferentiated somatoform disorder* is included only in the appendix.

To ascertain whether headache is part of the presentation of somatisation disorder, it is important to ask whether the patient has a history of multiple somatic complaints, since at any one time the patient may be focused on one particular complaint. Consider the following case scenario (from Yutzy, 2003):

A 35-year-old woman presented with a complaint of extreme headaches, "like a knife being stuck through the back of my head into my eye," as well as other headaches virtually every day. After medical and neurological examinations failed to suggest any specific aetiology for either headache, it was important to take a careful history of past symptoms. In this case, the woman also reported a history of other pains, including abdominal pain associated at times with nausea and vomiting, periods of constipation followed by diarrhoea which had resulted in investigation for gallbladder and peptic ulcer disease with no significant findings, and pain "in all of my joints" but particularly in her knees and her back that she said had been diagnosed as degenerative arthritis at age 27 years yet no deformities had developed since. She had had menstrual problems since menarche, with pain that put her to bed and excessive flow with "big blue clots", which had resolved only after hysterectomy two years earlier at age 33 years. The mother of four, she reported a long history of sexual problems including pain with intercourse. She had been told that she had a "tipped uterus". Throughout her life, she was seldom orgasmic and had not enjoyed sex "for years". She reported episodes of blurred vision with "spots" in front of her eyes, which caused her to stop work, and other episodes when she just could not hear anything, "like someone put their hands over my ears." She also reported periods of uncontrollable shaking and a feeling that she was losing control of her body, for which she had been investigated for seizures. She reported that, at times, she had feared having some serious medical disease but "with all the work-ups I have had, I am sure they would have found something by now."

As was evident after a complete medical history, the headaches were part of a much more involved syndrome. This woman had had multiple physical complaints with onset before age 30 that had no adequate medical explanation, were severe enough to cause her to seek medical attention and affected a variety of organ systems meeting the DSM-IV criteria for *Somatization disorder* (ie, at least four pain symptoms [headaches, abdominal pain, back pain and knee pain], at least two non-pain gastrointestinal symptoms [nausea, vomiting, diarrhoea and constipation], at least one sexual or reproductive symptom [pain on intercourse, excessive menstrual flow, loss of sexual enjoyment] and at least one pseudoneurological symptom [muffled hearing, uncontrollable shaking, blurred vision, spots in visual field]). Thus, her headaches would be correctly diagnosed as 12.1 *Headache attributed to somatisation disorder*.

## 12.2 Headache attributed to psychotic disorder

*Previously used terms:*

Delusional headache

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Delusional belief about the presence and/or aetiology of headache<sup>1</sup> occurring in the context of delusional disorder, schizophrenia, major depressive episode with psychotic features, manic episode with psychotic features or other psychotic disorder fulfilling DSM-IV criteria
- C. Headache occurs only when delusional
- D. Headache resolves when delusions remit
- E. Headache is not attributed to another cause

*Note:*

1. For example, a patient's false conviction that he or she has a brain tumour or intracranial mass giving rise to headache, which would fulfil DSM-IV criteria for *Delusional disorder, somatic type*.

*Comments:*

Delusions, as defined in DSM-IV, are false fixed beliefs based on incorrect inference about reality that are firmly held despite obvious proof to the contrary. Delusions, like any firmly-held belief, can be about virtually anything. In 12.2 *Headache attributed to psychotic disorder*, the delusion specifically involves the presence of headache. In some instances, the delusion may involve a false belief that a serious medical condition (*eg*, brain tumour) is present and is the cause of the headache, despite repeated and appropriate authoritative reassurance that no such medical condition is present. In other cases, the content of the delusion may be more bizarre: for example, a delusion that a transmitter has been surgically implanted into one's head and that the transmitter is the cause of the headache.

Delusional headache is apparently very rare and no empirical data are available about this condition.

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# **PART THREE**

## **CRANIAL NEURALGIAS, CENTRAL AND PRIMARY FACIAL PAIN AND OTHER HEADACHES**

## 13. CRANIAL NEURALGIAS AND CENTRAL CAUSES OF FACIAL PAIN

- 13.1 Trigeminal neuralgia
  - 13.1.1 Classical trigeminal neuralgia
  - 13.1.2 Symptomatic trigeminal neuralgia
- 13.2 Glossopharyngeal neuralgia
  - 13.2.1 Classical glossopharyngeal neuralgia
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### Introduction

Pain in the head and neck is mediated by afferent fibres in the trigeminal nerve, nervus intermedius, glossopharyngeal and vagus nerves and the upper cervical roots *via* the occipital nerves. Stimulation of these nerves by compression, distortion, exposure to cold or other forms of irritation or by a lesion in central pathways may give rise to stabbing or constant pain felt in the area innervated.

The cause may be clear, such as infection by herpes zoster or a structural abnormality demonstrated by imaging, but in some cases there may be no cause apparent for neuralgic pain.

Trigeminal and glossopharyngeal neuralgias present a problem of terminology. When pain is found to result from compression of the nerve by a vascular loop at operation, the neuralgia should strictly be regarded as secondary. Since many patients do not come to operation, it remains uncertain as to whether they have primary or secondary neuralgias. For this reason the term *classical* rather than

*primary* has been applied to those patients with a typical history even though a vascular source of compression may be discovered during its course. The term *secondary* can then be reserved for those patients in whom a neuroma or similar lesion is demonstrated.

Another difficulty arises with the condition that used to be known as *atypical facial pain* (an inappropriate term since many cases conform to a pattern). The fact that some cases follow surgery or injury to the face, teeth or gums suggests the possibility of an infectious or traumatic cause. Until more is known of the condition, *persistent idiopathic facial pain* seems a preferable non-committal title.

### 13.1 Trigeminal neuralgia

*Previously used term:*

Tic douloureux

#### 13.1.1 Classical trigeminal neuralgia

*Description:*

Trigeminal neuralgia is a unilateral disorder characterised by brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve. Pain is commonly evoked by trivial stimuli including washing, shaving, smoking, talking and/or brushing the teeth (trigger factors) and frequently occurs spontaneously. Small areas in the nasolabial fold and/or chin may be particularly susceptible to the precipitation of pain (trigger areas). The pains usually remit for variable periods.

*Diagnostic criteria:*

- A. Paroxysmal attacks of pain lasting from a fraction of a second to 2 minutes, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C
- B. Pain has at least one of the following characteristics:
  - 1. intense, sharp, superficial or stabbing
  - 2. precipitated from trigger areas or by trigger factors
- C. Attacks are stereotyped in the individual patient
- D. There is no clinically evident neurological deficit
- E. Not attributed to another disorder

*Comments:*

Classical trigeminal neuralgia usually starts in the second or third divisions, affecting the cheek or the chin. In <5% of patients the first division is affected. The pain never crosses to the opposite side but it may rarely occur bilaterally, in which case a central cause such as multiple sclerosis must be considered. Between paroxysms the patient is usually asymptomatic but a dull background pain may persist in some long-standing cases. Following a painful paroxysm there is usually a refractory period during which pain cannot be triggered. In some cases a paroxysm may be triggered from somatosensory stimuli outside the trigeminal area, such as a limb, or by other sensory stimulation such as bright lights, loud noises or tastes.

The pain often evokes spasm of the muscle of the face on the affected side (*tic douloureux*).

The increasing frequency of posterior fossa exploration and magnetic resonance imaging has demonstrated that many, possibly most, patients with this condition have compression of the trigeminal root by tortuous or aberrant vessels.

Classical trigeminal neuralgia is usually responsive, at least initially, to pharmacotherapy.

### **13.1.2 Symptomatic trigeminal neuralgia**

#### *Description:*

Pain indistinguishable from 13.1.1 *Classical trigeminal neuralgia* but caused by a demonstrable structural lesion other than vascular compression.

#### *Diagnostic criteria:*

- A. Paroxysmal attacks of pain lasting from a fraction of a second to 2 minutes, with or without persistence of aching between paroxysms, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C
- B. Pain has at least one of the following characteristics:
  - 1. intense, sharp, superficial or stabbing
  - 2. precipitated from trigger areas or by trigger factors
- C. Attacks are stereotyped in the individual patient
- D. A causative lesion, other than vascular compression, has been demonstrated by special investigations and/or posterior fossa exploration

#### *Comment:*

There may be sensory impairment in the distribution of the appropriate trigeminal division. 13.1.2 *Symptomatic trigeminal neuralgia* demonstrates no refractory period after a paroxysm, unlike 13.1.1 *Classical trigeminal neuralgia*.

## **13.2 Glossopharyngeal neuralgia**

### **13.2.1. Classical glossopharyngeal neuralgia**

#### *Description:*

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

#### *Diagnostic criteria:*

- A. Paroxysmal attacks of facial pain lasting from a fraction of a second to 2 minutes and fulfilling criteria B and C
- B. Pain has all of the following characteristics:
  - 1. unilateral location
  - 2. distribution within the posterior part of the tongue, tonsillar fossa, pharynx or beneath the angle of the lower jaw and/or in the ear
  - 3. sharp, stabbing and severe

- 4. precipitated by swallowing, chewing, talking, coughing and/or yawning
- C. Attacks are stereotyped in the individual patient
- D. There is no clinically evident neurological deficit
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

- 1. Other causes have been ruled out by history, physical examination and/or special investigations.

### **13.2.2 Symptomatic glossopharyngeal neuralgia**

*Description:*

Pain as in 13.2.1 *Classical glossopharyngeal neuralgia* with the proviso that aching pain may persist between paroxysms and sensory impairment may be found in the distribution of the glossopharyngeal nerve.

*Diagnostic criteria:*

- A. Paroxysmal attacks of facial pain lasting from a fraction of a second to 2 minutes, with or without persistence of aching between paroxysms, and fulfilling criteria B and C
- B. Pain has all of the following characteristics:
  - 1. unilateral location
  - 2. distribution within the posterior part of the tongue, tonsillar fossa, pharynx or beneath the angle of the lower jaw and/or in the ear
  - 3. sharp, stabbing and severe
  - 4. precipitated by swallowing, chewing, talking, coughing and/or yawning
- C. Attacks are stereotyped in the individual patient
- D. A causative lesion has been demonstrated by special investigations and/or surgery

### **13.3 Nervus intermedius neuralgia**

*Description:*

A rare disorder characterised by brief paroxysms of pain felt deeply in the auditory canal.

*Diagnostic criteria:*

- A. Pain paroxysms of intermittent occurrence, lasting for seconds or minutes, in the depth of the ear
- B. Presence of a trigger area in the posterior wall of the auditory canal
- C. Not attributed to another disorder<sup>1</sup>

*Note:*

- 1. Other causes, in particular a structural lesion, have been ruled out by history, physical examination and special investigations.

*Comment:*

Disorders of lacrimation, salivation and/or taste sometimes accompany the pain. There is a common association with herpes zoster. In view of the sparse innervation of the affected area by the nervus intermedius some patients may have an otalgic variant of glossopharyngeal neuralgia.

### **13.4 Superior laryngeal neuralgia**

*Description:*

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

*Diagnostic criteria:*

- A. Pain paroxysms lasting for seconds or minutes in the throat, submandibular region and/or under the ear and fulfilling criteria B-D
- B. Paroxysms are triggered by swallowing, straining the voice or head turning
- C. A trigger point is present on the lateral aspect of the throat overlying the hypothyroid membrane
- D. The condition is relieved by local anaesthetic block and cured by section of the superior laryngeal nerve
- E. Not attributed to another disorder<sup>1</sup>

*Note:*

- 1. Other causes, in particular a structural lesion, have been ruled out by history, physical examination and special investigations.

### **13.5 Nasociliary neuralgia**

*Previously used term:*

Charlin's neuralgia

*Description:*

A rare condition in which touching the outer aspect of one nostril causes a lancinating pain radiating to the medial frontal region.

*Diagnostic criteria:*

- A. Stabbing pain lasting seconds to hours in one side of the nose, radiating upwards to the medial frontal region and fulfilling criteria B and C
- B. Pain is precipitated by touching the lateral aspect of the ipsilateral nostril
- C. Pain is abolished by block or section of the nasociliary nerve, or by the application of cocaine to the nostril on the affected side

### **13.6 Supraorbital neuralgia**

*Description:*

An uncommon disorder characterised by pain in the region of the supraorbital notch and medial aspect of the forehead in the area supplied by the supraorbital nerve.

*Diagnostic criteria:*

- A. Paroxysmal or constant pain in the region of the supraorbital notch and medial aspect of the forehead in the area supplied by the supraorbital nerve
- B. Tenderness over the nerve in the supraorbital notch
- C. Pain is abolished by local anaesthetic blockade or ablation of the supraorbital nerve

### 13.7 Other terminal branch neuralgias

*Description:*

Injury or entrapment of peripheral branches of the trigeminal nerve other than the nasociliary and supraorbital nerves may give rise to pain referred to the area innervated by the branch affected. Examples are neuralgias of the infraorbital, lingual, alveolar and mental nerves.

*Diagnostic criteria:*

- A. Pain in the distribution of a peripheral branch of the trigeminal nerve other than the nasociliary or supraorbital nerves
- B. Tenderness over the affected nerve
- C. Pain is abolished by local anaesthetic blockade or ablation of the nerve

*Comment:*

A13.7.1 *Nummular headache*, described in the appendix, is probably a localised terminal branch neuralgia of the trigeminal nerve.

### 13.8 Occipital neuralgia

*Description:*

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

*Diagnostic criteria:*

- A. Paroxysmal stabbing pain, with or without persistent aching between paroxysms, in the distribution(s) of the greater, lesser and/or third occipital nerves
- B. Tenderness over the affected nerve
- C. Pain is eased temporarily by local anaesthetic block of the nerve

*Comment:*

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

### 13.9 Neck-tongue syndrome

*Description:*

The sudden onset of pain in the occiput or upper neck associated with abnormal sensation in the same side of the tongue.

*Diagnostic criteria:*

- A. Pain lasting seconds or minutes, with or without simultaneous dysaesthesia, in the area of distribution of the lingual nerve and second cervical root and fulfilling criteria B and C
- B. Pain has acute onset

- C. Pain is commonly precipitated by sudden turning of the head

*Comment:*

Proprioceptive fibres from the tongue enter the central nervous system through the second cervical dorsal root *via* connections between lingual and hypoglossal nerves and between the latter and the second cervical root. There is clinical and surgical evidence that the C2 root is compromised by sudden rotation of the neck, which is particularly likely when subluxation of the atlantoaxial joint occurs. The abnormal sensation in the ipsilateral side of the tongue may be numbness, paraesthesia or the sensation of involuntary movement.

### **13.10 External compression headache**

*Description:*

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

*Diagnostic criteria:*

- A. Headache with all of the following characteristics and fulfilling criteria C and D:
  - 1. non-pulsating
  - 2. increasing over minutes
  - 3. no accompanying symptoms
- B. Continuing application of external pressure to the forehead or scalp
- C. Headache develops during and is maximal at the site of pressure
- D. Headache resolves after pressure is relieved

*Comment:*

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

### **13.11 Cold-stimulus headache**

#### ***13.11.1 Headache attributed to external application of a cold stimulus***

*Description:*

Generalised headache following exposure of the unprotected head to a low environmental temperature as in very cold weather or in diving into cold water.

*Diagnostic criteria:*

- A. Diffuse and/or non-pulsating headache fulfilling criteria C and D:
- B. Presence of external cold stimulus to the head
- C. Headache develops during cold stimulus
- D. Headache resolves after removal of cold stimulus

### **13.11.2 Headache attributed to ingestion or inhalation of a cold stimulus**

*Previously used term:*

Ice-cream headache

*Description:*

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

*Diagnostic criteria:*

- A. Acute frontal<sup>1</sup> non-pulsatile headache fulfilling criteria C and D
- B. Cold stimulus to palate and/or posterior pharyngeal wall due to ingestion of cold food or drink or to inhalation of cold air
- C. Headache develops immediately, and only, after cold stimulus
- D. Headache resolves within 5 minutes after removal of cold stimulus

*Note:*

1. In migrainous patients, the headache may be referred to the usual site of migraine headache.

### **13.12 Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions**

*Description:*

Constant headache or facial pain caused by a lesion directly compromising afferent fibres in nerves mediating pain sensation from the head and/or neck. Sensory deficit may be detected within the appropriate distribution.

*Diagnostic criteria:*

- A. Constant and/or jabbing pain in the territory supplied by a cranial sensory nerve, fulfilling criteria C and D
- B. Evidence of compression, irritation or distortion of the appropriate cranial nerve
- C. Pain and compression, irritation or distortion occur simultaneously and correspond in location
- D. Pain is relieved by removal of the cause of compression, irritation or distortion

*Comments:*

Structural lesions may be space-occupying (*eg*, tumour or aneurysm) or contained within anatomical boundaries (*eg*, osteomyelitis of the cranial bones). If there is no sensory deficit or supporting imaging evidence, the diagnosis is doubtful.

Facial pain around the ear or temple may result from invasion of the vagus nerve by lung carcinoma.

### **13.13 Optic neuritis**

*Description:*

Pain behind one or both eyes accompanied by impairment of central vision caused by demyelination of the optic nerve.

*Diagnostic criteria:*

- A. Dull pain behind one or both eyes, worsened by eye movement and fulfilling criteria C and D
- B. Visual impairment due to a central or paracentral scotoma
- C. Onset of pain and onset of visual impairment separated by <4 weeks<sup>1</sup>
- D. Pain resolves within 4 weeks
- E. A compressive lesion has been ruled out

*Note:*

1. Pain precedes impairment of vision by <4 weeks. During this time, criterion B is not fulfilled and the diagnosis is *Probable optic neuritis*.

*Comments:*

Vision usually improves within 4 weeks.

Optic neuritis is often a manifestation of multiple sclerosis.

### **13.14 Ocular diabetic neuropathy**

*Description:*

Pain around the eye and forehead associated with paresis of one or more ocular cranial nerves (usually the third cranial nerve) in a patient with diabetes mellitus.

*Diagnostic criteria:*

- A. Pain, in a patient with diabetes mellitus, developing over a few hours around one eye
- B. Third cranial nerve palsy, often with sparing of pupillary function, and/or paresis of the fourth and/or sixth cranial nerves
- C. Neuropathy develops within 7 days of onset of pain<sup>1</sup>
- D. Not attributed to another disorder

*Note:*

1. Pain precedes signs of neuropathy by <7 days. During this time, criterion B is not fulfilled and the diagnosis is *Probable ocular diabetic neuropathy*.

### **13.15 Head or facial pain attributed to herpes zoster**

#### ***13.15.1 Head or facial pain attributed to acute herpes zoster***

*Description:*

Head or facial pain caused by herpes zoster.

*Diagnostic criteria:*

- A. Head or facial pain in the distribution of a nerve or nerve division and fulfilling criteria C and D
- B. Herpetic eruption in the territory of the same nerve
- C. Pain precedes herpetic eruption by <7 days<sup>1</sup>
- D. Pain resolves within 3 months

*Note:*

1. Pain precedes herpetic eruption by <7 days. During this time, criterion B is not fulfilled and the diagnosis is *Head or facial pain probably attributed to acute herpes zoster*.

*Comments:*

Herpes zoster affects the trigeminal ganglion in 10-15% of patients with the disease, and the ophthalmic division is singled out in some 80% of those patients. Herpes zoster may also involve the geniculate ganglion, causing an eruption in the external auditory meatus. The soft palate or areas of distribution of upper cervical roots may be involved in some patients.

Ophthalmic herpes may be associated with third, fourth and/or sixth cranial nerve palsies and geniculate herpes with facial palsy and/or acoustic symptoms. Zoster occurs in about 10% of patients with lymphoma and 25% of patients with Hodgkin's disease.

### **13.15.2 Post-herpetic neuralgia**

*Description:*

Facial pain persisting or recurring  $\geq 3$  months after the onset of herpes zoster.

*Diagnostic criteria:*

- A. Head or facial pain in the distribution of a nerve or nerve division
- B. Herpetic eruption in the territory of the same nerve
- C. Pain preceded herpetic eruption by <7 days
- D. Pain persists after 3 months

*Comment:*

Post-herpetic neuralgia is more often a sequel of herpes zoster as age advances, afflicting 50% of patients contracting zoster over the age of 60 years. Hypaesthesia or hyperalgesia and/or allodynia are usually present in the territory involved.

### **13.16 Tolosa-Hunt syndrome**

*Description:*

Episodic orbital pain associated with paralysis of one or more of the third, fourth and/or sixth cranial nerves which usually resolves spontaneously but tends to relapse and remit.

*Diagnostic criteria:*

- A. One or more episodes of unilateral orbital pain persisting for weeks if untreated
- B. Paresis of one or more of the third, fourth and/or sixth cranial nerves and/or demonstration of granuloma by MRI or biopsy
- C. Paresis coincides with the onset of pain or follows it within 2 weeks
- D. Pain and paresis resolve within 72 hours when treated adequately with corticosteroids
- E. Other causes have been excluded by appropriate investigations<sup>1</sup>

*Note:*

1. Other causes of painful ophthalmoplegia include tumours, vasculitis, basal meningitis, sarcoid, diabetes mellitus and ophthalmoplegic “migraine”.

*Comments:*

Some reported cases of Tolosa-Hunt syndrome had additional involvement of the trigeminal nerve (commonly the first division) or optic, facial or acoustic nerves. Sympathetic innervation of the pupil is occasionally affected.

The syndrome has been caused by granulomatous material in the cavernous sinus, superior orbital fissure or orbit in some biopsied cases.

Careful follow-up is required to exclude other possible causes of painful ophthalmoplegia.

### **13.17 Ophthalmoplegic “migraine”**

*Description:*

Recurrent attacks of headache with migrainous characteristics associated with paresis of one or more ocular cranial nerves (commonly the third nerve) in the absence of any demonstrable intracranial lesion other than MRI changes within the affected nerve.

*Diagnostic criteria:*

- A. At least 2 attacks fulfilling criterion B
- B. Migraine-like headache accompanied or followed within 4 days of its onset by paresis of one or more of the third, fourth and/or sixth cranial nerves
- C. Parasellar, orbital fissure and posterior fossa lesions ruled out by appropriate investigations

*Comment:*

This condition is very rare. It is unlikely that 13.17 *Ophthalmoplegic “migraine”* is a variant of migraine since the headache often lasts for a week or more and there is a latent period of up to 4 days from the onset of headache to the onset of ophthalmoplegia. Furthermore, in some cases MRI shows gadolinium uptake in the cisternal part of the affected cranial nerve which suggests that the condition may be a recurrent demyelinating neuropathy.

### **13.18 Central causes of facial pain**

#### **13.18.1 Anaesthesia dolorosa**

*Description:*

Persistent and painful anaesthesia or hypaesthesia in the distribution of the trigeminal nerve or one of its divisions or of the occipital nerves.

*Diagnostic criteria:*

- A. Persistent pain and dysaesthesia within the area of distribution of one or more divisions of the trigeminal nerve or of the occipital nerves
- B. Diminished sensation to pin-prick and sometimes other sensory loss over the affected area

- C. There is a lesion of the relevant nerve or its central connections

*Comment:*

Anaesthesia dolorosa is often related to surgical trauma of the occipital nerves or trigeminal ganglion, evoked most frequently after rhizotomy or thermocoagulation has been performed for treatment of 13.1.1 *Classical trigeminal neuralgia*.

### **13.18.2 Central post-stroke pain**

*Description:*

Unilateral pain and dysaesthesia associated with impaired sensation involving part or the whole of the face, not explicable by a lesion of the trigeminal nerve. It is attributed to a lesion of the quintothalamic (trigeminothalamic) pathway, thalamus or thalamocortical projection. Symptoms may also involve the trunk and/or limbs of the affected *or contralateral* side.

*Diagnostic criteria:*

- A. Pain and dysaesthesia in one half of the face, associated with loss of sensation to pin-prick, temperature and/or touch, fulfilling criteria C and D
- B. One or both of the following:
  - 1. history of sudden onset suggesting a vascular lesion (stroke)
  - 2. demonstration by CT or MRI of a vascular lesion in an appropriate site
- C. Pain and dysaesthesia develop within 6 months after stroke
- D. Not explicable by a lesion of the trigeminal nerve

*Comment:*

Facial pain following a thalamic lesion is part of a hemisyndrome. With lateral medullary lesions hemifacial pain may occur in isolation, but it is more often accompanied by crossed hemidysaesthesia.

The pain and dysaesthesia are usually persistent.

### **13.18.3 Facial pain attributed to multiple sclerosis**

*Coded elsewhere:*

Pain attributed to optic neuritis occurring as a manifestation of multiple sclerosis is coded as 13.13 *Optic neuritis*.

*Description:*

Unilateral or bilateral facial pain, with or without dysaesthesia, attributed to a demyelinating lesion of the central connections of the trigeminal nerve, which commonly remits and relapses.

*Diagnostic criteria:*

- A. Pain, with or without dysaesthesia, in one or both sides of the face
- B. Evidence that the patient has multiple sclerosis
- C. Pain and dysaesthesia develop in close temporal relation to, and with MRI demonstration of, a demyelinating lesion in the pons or quintothalamic (trigeminothalamic) pathway
- D. Other causes have been ruled out

*Comment:*

Pain may be tic-like, as in 13.1 *Trigeminal neuralgia*, or continuous. Trigeminal neuralgia occurring in young people or affecting one and then the other side should arouse the suspicion of multiple sclerosis.

#### **13.18.4 Persistent idiopathic facial pain**

*Previously used term:*

Atypical facial pain

*Description:*

Persistent facial pain that does not have the characteristics of the cranial neuralgias described above and is not attributed to another disorder.

*Diagnostic criteria:*

- A. Pain in the face, present daily and persisting for all or most of the day, fulfilling criteria B and C
- B. Pain is confined at onset to a limited area on one side of the face<sup>1</sup>, and is deep and poorly localised
- C. Pain is not associated with sensory loss or other physical signs
- D. Investigations including x-ray of face and jaws do not demonstrate any relevant abnormality

*Note:*

- 1. Pain at onset is commonly in the nasolabial fold or side of the chin, and may spread to the upper or lower jaw or a wider area of the face and neck.

*Comments:*

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

Facial pain around the ear or temple may precede the detection of an ipsilateral lung carcinoma causing referred pain by invasion of the vagus nerve.

The term *atypical odontalgia* has been applied to a continuous pain in the teeth or in a tooth socket after extraction in the absence of any identifiable dental cause.

#### **13.18.5 Burning mouth syndrome**

*Description:*

An intraoral burning sensation for which no medical or dental cause can be found.

*Diagnostic criteria:*

- A. Pain in the mouth present daily and persisting for most of the day
- B. Oral mucosa is of normal appearance
- C. Local and systemic diseases have been excluded

*Comment:*

Pain may be confined to the tongue (*glossodynia*). Subjective dryness of the mouth, paraesthesia and altered taste may be associated symptoms.

### **13.19 Other cranial neuralgia or other centrally-mediated facial pain**

Vail's Vidian neuralgia and Sluder's sphenopalatine neuralgia are not sufficiently validated. The recognition of Eagle's syndrome (Montalbetti *et al*, 1995) as a distinct entity awaits clarification.

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#### *13.6 Supraorbital neuralgia*

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## **14. OTHER HEADACHE, CRANIAL NEURALGIA, CENTRAL OR PRIMARY FACIAL PAIN**

14.1 Headache not elsewhere classified

14.2 Headache unspecified

### **Introduction**

In order to make this classification exhaustive there is, after the entries for many disorders, a subcategory for conditions that fulfil all but one criterion for that disorder. Still there may be headaches that cannot fit into any of the existing chapters because they are being described for the first time, or because there simply is not enough information available. This chapter is intended for these types or subtypes of headaches.

### **14.1 Headache not elsewhere classified**

*Previously used term:*

Headache not classifiable

*Diagnostic criteria:*

- A. Headache with characteristic features suggesting that it is a unique diagnostic entity
- B. Does not fulfil criteria for any of the headache disorders described above

*Comment:*

Several new headache entities have been described in the time between the first edition of *The International Classification of Headache Disorders* and this second edition. It is anticipated that there are more entities still to be described. Such headaches, until classified, can be coded as 14.1 *Headache not elsewhere classified*.

### **14.2 Headache unspecified**

*Previously used term:*

Headache not classifiable

*Diagnostic criteria:*

- A. Headache is or has been present
- B. Not enough information is available to classify the headache at any level of this classification

*Comment:*

It is also apparent that a diagnosis must be made in a large number of patients where very little information is available, allowing only to state that they have headache but not which type of headache. Such patients are coded as 14.2 *Headache unspecified*. This code, however, must never be used as an excuse for not gathering detailed information about a headache when such information is available. It should be used only in situations where information cannot be obtained because the patient is dead, unable to communicate or unavailable.

# APPENDIX

## Introduction

In the first edition of *The International Classification of Headache Disorders* there was no appendix. This time an appendix is added which, we hope, will be used in several ways.

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 studies. However, the experience of the experts in the Headache Classification Subcommittee and publications of variable quality suggest that there are a number of diagnostic entities that are believed to be real but for which further scientific evidence must be presented before they can be formally accepted. Therefore it is anticipated that a number of the disorders now in the appendix will move into the main body of the classification next time the classification is revised.

In a few places we present an alternative set 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 this may be a good idea, but the subcommittee still does not feel that the evidence is sufficient to change the main classification. This is, for example, the case for the accompanying symptoms of migraine without aura. The alternative diagnostic criterion D in the appendix is easier both to understand and to apply, but not yet sufficiently validated.

Finally, the appendix is used as a first step in eliminating disorders included as diagnostic entities in the first edition because of tradition but for which sufficient evidence has still not been published.

## **A1. Migraine**

### **A1.1 Migraine without aura**

#### *Alternative diagnostic criteria:*

- A. At least 5 attacks fulfilling criteria B-D
- B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated) and occurring on <15 days/month
- C. Headache has at least two of the following characteristics:
  - 1. unilateral location
  - 2. pulsating quality
  - 3. moderate or severe pain intensity
  - 4. aggravation by or causing avoidance of routine physical activity (*eg*, walking or climbing stairs)
- D. During headache at least two of the following:
  - 1. nausea
  - 2. vomiting
  - 3. photophobia
  - 4. phonophobia
  - 5. osmophobia
- E. Not attributed to another disorder

#### *Comment:*

Only criterion D is different from those in the main body of the classification. Whilst this alternative appears easier both to understand and to apply, it is not yet sufficiently validated.

#### ***A1.1.1 Pure menstrual migraine without aura***

##### *Diagnostic criteria:*

- A. Attacks, in a menstruating woman, fulfilling criteria for 1.1 *Migraine without aura*
- B. Attacks occur exclusively on day  $1 \pm 2$  (*ie*, days  $-2$  to  $+3$ )<sup>1</sup> of menstruation<sup>2</sup> in at least two out of three menstrual cycles and at no other times of the cycle

##### *Notes:*

- 1. The first day of menstruation is day 1 and the preceding day is day  $-1$ ; there is no day 0.
- 2. For the purposes of this classification, menstruation is considered to be endometrial bleeding resulting from either the normal menstrual cycle or from the withdrawal of exogenous progestogens, as in the case of combined oral contraceptives and cyclical hormone replacement therapy.

#### ***A1.1.2 Menstrually-related migraine without aura***

##### *Diagnostic criteria:*

- A. Attacks, in a menstruating woman, fulfilling criteria for 1.1 *Migraine without aura*

- B. Attacks occur on day  $1 \pm 2$  (ie, days  $-2$  to  $+3$ )<sup>1</sup> of menstruation<sup>2</sup> in at least two out of three menstrual cycles and additionally at other times of the cycle

*Notes:*

1. The first day of menstruation is day 1 and the preceding day is day  $-1$ ; there is no day 0.
2. For the purposes of this classification, menstruation is considered to be endometrial bleeding resulting from either the normal menstrual cycle or from the withdrawal of exogenous progestogens, as in the case of combined oral contraceptives and cyclical hormone replacement therapy.

### ***A1.1.3 Non-menstrual migraine without aura***

*Diagnostic criteria:*

- A. Attacks, in a menstruating woman, fulfilling criteria for 1.1 *Migraine without aura*  
B. Attacks have no menstrual relationship<sup>1</sup>

*Note:*

1. That is, they do not fulfil criterion B for A1.1.1 *Pure menstrual migraine without aura* or A1.1.2 *Menstrually-related migraine without aura*.

*Comments:*

This subclassification of 1.1 *Migraine without aura* is applicable only to menstruating women.

The importance of distinguishing between A1.1.1 *Pure menstrual migraine* and A1.1.2 *Menstrually-related migraine* is that hormone prophylaxis is more likely to be effective for pure menstrual migraine. Documented prospectively-recorded evidence, kept for a minimum of three cycles, is necessary to confirm the diagnosis as many women over-report an association between attacks and menstruation.

Menstrual attacks are mostly migraine without aura. In a woman who has migraine both with and without aura, migraine with aura does not appear to be associated with menstruation.

The mechanism(s) of migraine may be different with endometrial bleeding resulting from the normal menstrual cycle and bleeding 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 subpopulations. Management strategies may also differ for these distinct subpopulations.

There is some evidence that menstrual 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. If 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 8.4.3 *Oestrogen-withdrawal headache* should be used.

### ***A1.2.7 Migraine aura status***

#### *Diagnostic criteria:*

- A. Migraine aura fulfilling aura criteria for 1.2 *Migraine with aura* or one of its subtypes
- B. At least 2 auras per day for  $\geq 5$  consecutive days

### ***A1.3.4 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
- B. Onset before the age of 18 months
- C. At least one other paroxysmal phenomenon is associated with the bouts of hemiplegia or occurs independently, such as tonic spells, dystonic posturing, choreoathetoid movements, nystagmus or other ocular motor abnormalities, autonomic disturbances
- D. Evidence of mental and/or neurological deficit(s)
- E. Not attributed to another disorder

#### *Comment:*

This is a heterogeneous condition that includes neurodegenerative disorders. A relationship with migraine is suggested on clinical grounds. The possibility that it is an unusual form of epilepsy cannot be ruled out.

### ***A1.3.5 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. It may evolve into 1.3.3 *Benign paroxysmal vertigo of childhood* or 1.2 *Migraine with aura*, or cease without further symptoms.

#### *Diagnostic criteria:*

- A. Episodic attacks, in a young child, with all of the following characteristics and fulfilling criterion B:
  - 1. tilt of the head to one side (not always the same side), with or without slight rotation
  - 2. lasting minutes to days
  - 3. remitting spontaneously and tending to recur monthly
- B. During attacks, symptoms and/or signs of one or more of the following:
  - 1. pallor
  - 2. irritability
  - 3. malaise
  - 4. vomiting
  - 5. ataxia<sup>1</sup>

- C. Normal neurological examination between attacks
- D. Not attributed to another disorder

*Note:*

1. Ataxia is more likely in older children within the affected age group.

*Comments:*

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

A1.3.5 *Benign paroxysmal torticollis* may evolve to 1.3.3 *Benign paroxysmal vertigo of childhood* or 1.2 *Migraine with aura* (particularly 1.2.6 *Basilar-type migraine*).

These observations need further validation by patient diaries, structured interviews and longitudinal data collection. 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.

## **A2. Tension-type headache**

*Comment:*

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. Headache lasting from 30 minutes to 7 days
- C. At least three of the following pain 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 (anorexia may occur), vomiting, photophobia or phonophobia
- E. Not attributed to other disorder<sup>1;2</sup>

*Notes:*

1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorder but it is ruled out by appropriate investigations, or such disorder is present but headache does not occur for the first time in close temporal relation to the disorder.
2. In the case of A2.3 *Chronic tension-type headache* and when medication overuse is present and fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache*, it is

uncertain whether this criterion E is fulfilled until 2 months after medication has been withdrawn without improvement (see *Comment*).

*Comment:*

In many uncertain cases of chronic tension-type headache there is overuse of medication. When this fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache*, the default rule is to code for 2.4.3 *Probable chronic tension-type headache* plus 8.2.8 *Probable medication-overuse headache*. When these criteria A-E are still fulfilled 2 months after medication overuse has ceased, A2.3 *Chronic tension-type headache* should be diagnosed and 8.2.8 *Probable medication-overuse headache* must be discarded. Alternatively, if at any time sooner they are no longer fulfilled because improvement has followed withdrawal, 8.2 *Medication-overuse headache* should be diagnosed and 2.4.3 *Probable chronic tension-type headache* must be discarded.

### **A3. Cluster headache and other trigeminal autonomic cephalalgias**

#### **A3.3 Short-lasting Unilateral Neuralgiform headache attacks with cranial Autonomic symptoms (SUNA)**

*Comments:*

The current classification for 3.3 *SUNCT* has some notable problems. First, the name implies that all patients must have both conjunctival injection and tearing. This is not the Subcommittee's invariable clinical experience. It is possible that 3.3 *SUNCT* is a subform of a broader problem of A3.3 *SUNA*. This proposal requires validation. Second, the pain of the attacks can be difficult to differentiate from that of 13.1 *Trigeminal neuralgia* affecting the ophthalmic division. One suggested distinction is the absence of a refractory period to cutaneous stimulation in A3.3 *SUNA*. Third, the criterion for attack frequency in 3.3 *SUNCT* is rather unhelpful given the breadth of variation it allows. Since attacks are usually at least daily, simplifying the frequency requirement may be more useful.

The following proposed criteria for A3.3 *SUNA* (as an alternative to 3.3 *SUNCT*) are for research purposes and need to be tested. Cranial autonomic features should be prominent to distinguish this disorder from ophthalmic division trigeminal neuralgia.

*Diagnostic criteria:*

- A. At least 20 attacks fulfilling criteria B-E
- B. Attacks of unilateral orbital, supraorbital or temporal stabbing or pulsating pain lasting from 2 seconds to 10 minutes
- C. Pain is accompanied by one of:
  - 1. conjunctival injection and/or lacrimation
  - 2. nasal congestion and/or rhinorrhoea
  - 3. eyelid oedema
- D. Attacks occur with a frequency of  $\geq 1$  per day for more than half of the time
- E. No refractory period follows attacks triggered from trigger areas
- F. Not attributed to another disorder

### **A3.3.1 Episodic SUNA**

#### *Description:*

SUNA attacks occurring in periods lasting 7 days to 1 year separated by pain-free intervals lasting one month or longer.

#### *Diagnostic criteria:*

- A. Attacks fulfilling criteria A-F for A3.3 SUNA
- B. At least 2 attack periods lasting (if untreated) from 7 days to 1 year and separated by pain-free remission periods of  $\geq 1$  month

### **A3.3.2 Chronic SUNA**

#### *Description:*

SUNA attacks occurring for more than 1 year without remission, or with remissions lasting less than 1 month.

#### *Diagnostic criteria:*

- A. Attacks fulfilling criteria A-F for A3.3 SUNA
- B. Attacks recur over  $>1$  year without remission periods or with remission periods lasting  $<1$  month

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## **A6. Headache attributed to cranial or cervical vascular disorder**

### **A6.5.6 Carotidynia**

Carotidynia has been removed from the main classification to the appendix because an extensive literature survey suggests that it is not an entity but a syndrome encompassing many varieties of pain in the carotid region. In particular, carotidynia as described in the first edition of *The International Classification of Headache Disorders* – neck pain lasting  $<2$  weeks with tenderness to palpation over the carotid bifurcation – can be due to carotid dissection (which should be coded as 6.5.1 *Arterial dissection*). Some cases have recently been published of carotidynia with MRI abnormalities (described as an intermediate signal on T1W1 and a masked ring enhancement after gadolinium injection) in the tissue surrounding the symptomatic artery. Until the specificity of this finding is established, carotidynia is better considered as a syndrome rather than as a distinct entity.

## **A6.8 Chronic post-vascular disorder headache**

### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. A vascular disorder has been present but has been effectively treated or has remitted spontaneously
- C. Headache has been attributed to the vascular disorder
- D. Headache persists for >3 months after effective treatment or spontaneous remission of the vascular disorder

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## **A7. Headache attributed to non-vascular intracranial disorder**

### **A7.9.1 Post-radiosurgery headache**

#### *Diagnostic criteria:*

- A. Diffuse and/or holocranial headache fulfilling criteria C and D
- B. Radiosurgery of the brain has been performed
- C. Headache develops within 7 days after radiosurgery
- D. Headache resolves within 3 months after radiosurgery

#### *Comment:*

Although *de novo* headache has been described after radiosurgery, most studies do not provide a detailed description of the clinical characteristics of the headache, nor 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, the relationships between these headaches and the radiosurgical procedures preceding them are highly doubtful. Carefully controlled prospective studies are necessary to determine whether a unique headache disorder can occur after radiosurgery and, if so, how it is related to the type and location of lesion being irradiated and/or the dosage and radiation field employed.

### **A7.9.2 Post-electroconvulsive therapy (ECT) headache**

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Electroconvulsive therapy (ECT) has been given
- C. Headache develops within 4 hours after ECT and after at least 50% of treatments
- D. Headache resolves within 72 hours after ECT

#### *Comments:*

Clear descriptions of headache associated with ECT are sparse. Published data may not be adequate to define post-ECT headache operationally.

The characteristics of headache after ECT are noted in several case reports. Hawken *et al* (2001) reported on a patient who suffered from “mild migraine” every two to three days and “more severe” migraine every 7-10 days after ECT (the symptoms listed correspond with diagnostic criteria for 1.1 *Migraine without aura*). Headache developed immediately after the patient regained consciousness following sessions of ECT. On one of six occasions the headache was associated with nausea but other symptoms of migraine were not described in this report. The headache did not respond to sumatriptan but was alleviated by a combination of propranolol and naproxen, and appeared to be prevented by administration of propranolol prior to ECT. De Battista and Mueller (1995) described a patient who developed severe post-ECT unilateral headaches associated with nausea/vomiting and photophobia. The patient had a history of similar although less intense headaches. Prophylactic administration of sumatriptan appeared to prevent the headache whereas prophylactic administration of beta-blockers did not. Ghoname *et al* (1999) reported on five patients who experienced headaches immediately after sessions of ECT. The headaches were severe and bilateral in each case (pulsating in two), but no other symptoms of migraine were described. Several other letters and case reports have documented attacks of severe headache (associated with symptoms of migraine or described as being similar to migraine) triggered by ECT in patients with a history of migraine (eg, Folkerts, 1995; Oms *et al*, 1998). Markowitz *et al* (2001) reported that, of 13 moderate or severe attacks of headache after ECT, six were associated with sensitivity to light, four with sensitivity to noise, three with nausea and one with vomiting. All but one of the attacks decreased within 1.5 hours after intranasal administration of sumatriptan 20 mg.

### **A7.10 Chronic post-intracranial disorder headache**

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. An intracranial disorder has been present but has been effectively treated or has remitted spontaneously
- C. Headache has been attributed to the intracranial disorder
- D. Headache persists for >3 months after effective treatment or spontaneous remission of the intracranial disorder

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## A8. Headache attributed to a substance or its withdrawal

### 8.1.10 Headache as an acute adverse event attributed to medication used for other indications

Table 1: Drugs that may induce headache or worsen pre-existing headache

Acetazolamide	Codeine	Interferons	Ondansetron
Ajmaline	Didanosine	Isoniazid	Paroxetine
Amantadine	Dihydralazine	Meprobamate	Pentoxifylline
Antihistaminics	Dihydroergotamine	Methaqualone	Perhexiline
Barbiturates	Dipyridamole	Metronidazole	Primidone
Beta-interferon	Disopyramide	Morphine and derivatives	Prostacyclines
Bromocriptine	Disulfiram	Nalidixic acid	Ranitidine
Caffeine	Ergotamine	Nifedipine	Rifampicin
Calcium antagonists	Etofibrate	Nitrofurantoin	Sildenafil
Carbimazol	Gestagens	Nitrates	Theophylline and derivatives
Chinidine	Glycosides	Non-steroidal anti-inflammatory drugs	Thiamazole

Chloroquine	Griseofulvin	Octreotide	Trimethoprim + sulfamethoxazole
Cimetidine	Guanethidine	Oestrogens	Triptans
Clofibrate	Immunoglobulins	Omeprazole	Vitamin A

### A8.5 Chronic post-substance exposure headache

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. Exposure to a substance has been present but has ceased
- C. Headache has been attributed to exposure to the substance
- D. Headache persists for >3 months after exposure to the substance ceased

### A9. Headache attributed to infection

#### *A9.1.6 Headache attributed to space-occupying intracranial infectious lesion or infestation*

#### *Comment:*

There are space-occupying intracranial infectious lesions causing headache other than brain abscess or subdural empyema. Since the pathophysiology is miscellaneous and the systematic studies to classify these headaches are inadequate, tentative diagnostic criteria are given here in the appendix.

#### *Diagnostic criteria:*

- A. Headache with at least one of the following characteristics and fulfilling criteria C and D:
  - 1. diffuse continuous pain
  - 2. aggravated by straining
  - 3. accompanied by nausea and/or focal neurological symptoms and/or signs
- B. Evidence of a space-occupying intracranial infectious lesion or infestation from neuroimaging and/or laboratory investigations
- C. Headache develops during the space-occupying intracranial infection or infestation
- D. Headache resolves within 3 months<sup>1</sup> after successful treatment of the lesion

#### *Note:*

- 1. Headache usually resolves within 1 month.

#### *Comments:*

A direct space-occupying effect leading to raised intracranial pressure and/or irritation of the meningeal or arterial structures are the mechanisms for causing headache of this subtype.

The most common organisms causing space occupying granulomatous or cystic central nervous system diseases are mycobacteria, fungi (*eg*, *Cryptococcus neoformans* and others), *Toxoplasma gondii*, free living amoebae, cestodes (*eg*, *Cysticercus cellulosae*, *Coenurus cerebralis*, *Sparganum* species), nematodes (*eg*, *Toxocara canis*, lymphatic filariae, *Onchocerca volvulus*, *Anisakis* species) and trematodes (*eg*, *Schistosoma* species, in particular *Schistosoma japonicum*, and *Paragonimus* species).

### ***A9.1.7 Headache attributed to intracranial parasitic infestation***

#### *Coded elsewhere:*

Headache attributed to space occupation by rather than to a direct effect of an intracranial parasitic infestation is coded as A9.1.6 *Headache attributed to space-occupying intracranial infectious lesion or infestation*.

#### *Comment:*

Parasitic infestations are characterised by an acute stage and a chronic stage. Headache in the acute stage is usually due to meningitis while headache in the chronic stage is believed to be due to encephalitic changes or secondary to neuropsychological deterioration. Systematic studies of the headaches caused by these disorders are lacking and therefore diagnostic criteria can only be proposed with great uncertainty.

#### *Diagnostic criteria:*

- A. Headache with one or other of the following characteristics, with or without focal neurological symptoms and/or signs and fulfilling criteria C and D:
  - 1. headache, with acute onset, resembling 9.1.1 *Headache attributed to bacterial meningitis*
  - 2. headache with more insidious onset and characteristic of chronic meningoencephalitis
- B. Evidence of an intracranial parasitic infestation from CSF examination, blood serology and/or neuroimaging
- C. Headache develops during the parasitic infestation
- D. Headache resolves within 3 months after successful treatment of the infestation

#### *Comments:*

Headache is a common and frequently the first symptom of intracranial parasitic infestation. A wide variety of parasitic organisms may infest the central nervous system, directly or indirectly. Whereas *Trypanosoma cruzi* (American trypanosomiasis, Chagas' disease) may cause acute meningitis, *T. brucei gambiense* (West African trypanosomiasis, Gambian sleeping sickness) and *T. brucei rhodesiense* (East African trypanosomiasis, East African sleeping sickness) cause a chronic meningoencephalitis.

Predisposing factors include exposure to parasites in tropical and/or subtropical areas of prevalence and, in a few instances, immunocompromised status.

### ***A9.4.2 Chronic post-non-bacterial infection headache***

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. A non-bacterial infection has been present but has been effectively treated or has remitted spontaneously
- C. Headache has been attributed to the infection
- D. Headache persists for >3 months after effective treatment or spontaneous remission of the infection

*Comment:*

There is little evidence for the existence of chronic headache attributed to non-bacterial infections. More research is needed.

## **Bibliography**

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## **A10. Headache attributed to disorder of homoeostasis**

### *A10.7.1 Headache attributed to other metabolic or systemic disorders*

Headaches attributed to the following disorders are not sufficiently validated: anaemia, hypercapnia, adrenocortical insufficiency, mineralocorticoid deficiency, hyperaldosteronism, polycythaemia, hyperviscosity syndrome, thrombotic thrombocytopenic purpura, plasmapheresis-induced headache, 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.

### **A10.8 Chronic post-homoeostasis disorder headache**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. A disorder of homeostasis has been present but has been effectively treated or has remitted spontaneously
- C. Headache has been attributed to the disorder of homoeostasis
- D. Headache persists for >3 months after treatment or remission of the disorder of homoeostasis

*Comment:*

Some patients may suffer from persistent headache after resolution of a disorder of homoeostasis. Such headache has never been the subject of systematic study.

## **A11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures**

### ***A11.5.1 Mucosal contact point headache***

#### *Diagnostic criteria:*

- A. Intermittent pain localised to the periorbital and medial canthal or temporozygomatic regions and fulfilling criteria C and D
- B. Clinical, nasal endoscopic and/or CT imaging evidence of mucosal contact points without acute rhinosinusitis
- C. Evidence that the pain can be attributed to mucosal contact based on at least one of the following:
  - 1. pain corresponds to gravitational variations in mucosal congestion as the patient moves between upright and recumbent postures
  - 2. abolition of pain within 5 minutes after diagnostic topical application of local anaesthesia to the middle turbinate using placebo- or other controls<sup>1</sup>
- D. Pain resolves within 7 days, and does not recur, after surgical removal of mucosal contact points

#### *Note:*

- 1. Abolition of pain means complete relief of pain, indicated by a score of zero on a visual analogue scale (VAS).

#### *Comment:*

A11.5.1 *Mucosal contact point headache* is a new entry to the classification for which evidence is limited. Controlled trials are recommended to validate it, using the listed criteria for patient selection.

## **A11.9 Chronic post-cranio-cervical disorder headache**

#### *Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C and D
- B. A cranio-cervical disorder has been present but has been effectively treated or has remitted spontaneously
- C. Headache has been attributed to the cranio-cervical disorder
- D. Headache persists for >3 months after effective treatment or spontaneous remission of the cranio-cervical disorder

## **A12. Headache attributed to psychiatric disorder**

While headaches are commonly associated with various psychiatric disorders, whether this relationship is causal and, if so, the direction of the causation remain objects of study. The following are offered as candidate criteria sets to facilitate research into the possible causal relationships between certain psychiatric disorders and headache. It is not recommended that they be used routinely in clinical practice to describe the association between comorbid headache and

psychiatric disorders. In the vast majority of cases, headache associated with these disorders most probably reflects common underlying risk factors or aetiologies.

Note that when making any of the diagnoses listed below, it is crucial to establish that the headache in question occurs exclusively during the course of the psychiatric disorder. This should be interpreted to mean that the headache is manifest only during times when the symptoms of the psychiatric disorder are also manifest. Thus, for example, in a child with separation anxiety disorder, headache should be attributed to separation anxiety disorder only in those cases where it occurs solely in the context of actual or threatened separation. Similarly, in an adult with panic disorder, headache should be attributed to panic disorder only in those cases where it occurs solely as one of the symptoms of a panic attack.

### **A12.3 Headache attributed to major depressive disorder**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Presence of major depressive disorder fulfilling DSM-IV criteria:
  - 1. one or more episodes in which, during the same 2-week period, at least five of the following symptoms are present:
    - a) depressed mood
    - b) markedly diminished interest or pleasure
    - c) weight or appetite change
    - d) insomnia or hypersomnia
    - e) psychomotor agitation or retardation
    - f) fatigue or loss of energy
    - g) feelings of worthlessness or excessive or inappropriate guilt
    - h) diminished ability to concentrate or indecisiveness
    - i) recurrent thoughts of death, suicidal idea, plan or attempt
  - 2. occurring in the absence of any manic or hypomanic episodes
  - 3. not better accounted for by bereavement and not due to the direct physiological effects of a medical condition or substance
- C. Headache occurs exclusively during major depressive episodes
- D. Headache resolves or greatly improves within 3 months after the major depressive disorder is in full remission
- E. Headache is not attributed to another cause

*Comment:*

Since tricyclic antidepressants are effective against certain types of headache, remission of headache is more suggestive of a psychiatric cause of the headache when major depressive disorder improves under treatment with other antidepressants than tricyclic antidepressants.

### **A12.4 Headache attributed to panic disorder**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Presence of panic disorder fulfilling DSM-IV criteria:

1. recurrent unexpected panic attacks followed by a month or more of worry about having another attack, worry about the implications of an attack or a change in behaviour because of the attack
  2. panic attack is defined as a discrete period of intense fear or discomfort in which at least four of the following symptoms develop abruptly and reach a peak within 10 minutes:
    - a) palpitations
    - b) pounding heart or accelerated heart rate
    - c) sweating, trembling or shaking
    - d) sensations of shortness of breath or smothering
    - e) feelings of choking, chest pain or discomfort
    - f) nausea or abdominal distress
    - g) feeling dizzy, unsteady, light-headed or faint
    - h) derealisation or depersonalisation
    - i) fear of losing control or going crazy
    - j) fear of dying
    - k) paraesthesias
    - l) chills or hot flushes
  3. panic attacks are not due to the physiological effects of a medical condition or substance
- C. Headache occurs exclusively during panic attacks
- D. Headache resolves, and does not recur, after panic disorder remits
- E. Headache is not attributed to another cause

### **A12.5 Headache attributed to generalised anxiety disorder**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Presence of generalised anxiety disorder fulfilling DSM-IV criteria:
  1. excessive anxiety and worry, occurring on more days than not for at least 6 months, about a number of events or activities and which the patient finds difficult to control
  2. associated with at least three of the following:
    - a) restlessness or feeling keyed-up or on edge
    - b) being easily fatigued
    - c) difficulty concentrating or mind going blank
    - d) irritability
    - e) muscle tension
    - f) sleep disturbance
  3. not occurring exclusively during a mood disorder
  4. not due to the direct physiological effects of a medical condition or substance
- C. Headache occurs exclusively during the course of the generalised anxiety disorder
- D. Headache resolves, and does not recur, after the generalised anxiety disorder remits
- E. Headache is not attributed to another cause

### **A12.6 Headache attributed to undifferentiated somatoform disorder**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C-E

- B. Presence of undifferentiated somatoform disorder defined by a somatoform symptom, in addition to headache, fulfilling DSM-IV criteria for *Undifferentiated somatoform disorder*:
  1. a physical complaint, plus headache, that, after appropriate investigation, cannot be fully explained by a known general medical condition or by the direct effects of a substance or medication or, when there is a related medical condition, the complaint or impairment is in excess of what would be expected from the history, examination and/or laboratory findings
  2. the physical complaint and headache cause distress or impairment and last at least 6 months
- C. Headache occurs exclusively during the course of the other physical complaint
- D. Headache resolves after the undifferentiated somatoform disorder remits
- E. Headache is not attributed to another cause

### **A12.7 Headache attributed to social phobia**

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Presence of social phobia fulfilling DSM-IV criteria:
  1. a marked and persistent fear of one or more social or performance situations, including school, in which the patient is exposed to unfamiliar people or to possible scrutiny by others, and in which the patient fears that he or she will act in a way that will be humiliating or embarrassing
  2. the patient either avoids social situations or endures them with marked distress
  3. the phobia is a source of distress or causes impairment in social or occupational functioning
- C. Headache occurs exclusively during the course of social phobia
- D. Headache resolves after social phobia remits
- E. Headache is not attributed to another cause

### **A12.8 Headache attributed to separation anxiety disorder**

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Presence of separation anxiety disorder fulfilling at least three of the following DSM-IV criteria, lasting  $\geq 6$  months and with onset before age 18:
  1. recurrent excessive distress when separation from home or major attachment figures occurs or is anticipated
  2. persistent and excessive worry about losing, or about possible harm befalling, major attachment figures
  3. persistent and excessive worry that an untoward event will lead to separation from a major attachment figure (*eg*, getting lost or being kidnapped)
  4. persistent reluctance or refusal to go to school or elsewhere because of fear of separation
  5. persistently and excessively fearful or reluctant to be alone or without major attachment figures at home or without significant adults in other settings
  6. persistent reluctance or refusal to go to sleep without being near a major attachment figure or to sleep away from home
  7. repeated nightmares involving themes of separation
  8. repeated complaints of physical symptoms (such as headaches, stomach-aches, nausea or vomiting) when separation from major attachment figures occurs or is anticipated

- C. Headache occurs exclusively during the course of the separation anxiety disorder
- D. Headache resolves after separation anxiety disorder remits
- E. Headache is not attributed to another cause

### **A12.9 Headache attributed to post-traumatic stress disorder**

*Diagnostic criteria:*

- A. Headache, no typical characteristics known, fulfilling criteria C-E
- B. Presence of post-traumatic stress disorder fulfilling DSM-IV criteria:
  1. the patient has been exposed to a traumatic event in which both of the following were present:
    - a) the patient experienced, witnessed or was confronted with an event or events that involved actual or threatened death or serious injury, or a threat to the physical integrity of self or others
    - b) the patient's response involved intense fear, helplessness or horror
  2. the traumatic event is persistently re-experienced in at least one of the following ways:
    - a) recurrent and intrusive distressing recollections of the event, including images, thoughts or perceptions
    - b) recurrent distressing dreams of the event
    - c) acting or feeling as if the traumatic event were recurring (including a sense of reliving the experience, illusions, hallucinations and dissociative flashback episodes, including any of these that occur on awakening or when intoxicated)
    - d) intense psychological distress at exposure to internal or external cues that symbolise or resemble an aspect of the traumatic event
    - e) physiological reactivity on exposure to internal or external cues that symbolise or resemble an aspect of the traumatic event
  3. persistent avoidance of stimuli associated with the trauma and a numbing of general responsiveness (not present before the trauma), as indicated by at least three of the following:
    - a) efforts to avoid thoughts, feelings or conversations associated with the trauma
    - b) efforts to avoid activities, places or people that arouse recollections of the trauma
    - c) inability to recall an important aspect of the trauma
    - d) markedly diminished interest or participation in significant activities
    - e) feeling of detachment or estrangement from others
    - f) restricted range of affect (*eg*, unable to have loving feelings)
    - g) sense of a foreshortened future (*eg*, not expecting to have a career, marriage, children or a normal life span)
  4. persistent symptoms of increased arousal (not present before the trauma), as indicated by at least two of the following:
    - a) difficulty falling asleep or staying asleep
    - b) irritability or outbursts of anger
    - c) difficulty concentrating
    - d) hypervigilance
    - e) exaggerated startle response
  5. symptoms in B2, B3 and B4 have lasted >1 month
- C. Headache occurs exclusively during the course of the post-traumatic stress disorder
- D. Headache resolves after the post-traumatic stress disorder remits

- E. Headache is not attributed to another cause

## **A13. Cranial neuralgias and central causes of facial pain**

### ***A13.7.1 Nummular headache***

#### *Previously used terms:*

Coin-shaped cephalgia

#### *Description:*

Pain in a small circumscribed area of the head in the absence of any lesion of the underlying structures.

#### *Diagnostic criteria:*

- A. Mild to moderate head pain fulfilling criteria B and C:
- B. Pain is felt exclusively in a rounded or elliptical area typically 2-6 cm in diameter
- C. Pain is chronic and either continuous or interrupted by spontaneous remissions lasting weeks to months
- D. Not attributed to another disorder

#### *Comments:*

There is a slight female preponderance.

Nummular headache is probably a localised terminal branch neuralgia of the trigeminal nerve.

The painful area may be localised in any part of the head but is usually in the parietal region. The pain remains confined to the same symptomatic area which does not change in shape or size over time. Lancinating exacerbations lasting for several seconds or gradually increasing over 10 minutes to 2 hours may be superimposed on the base-line pain. During and between symptomatic periods, the affected area may show variable combinations of hypaesthesia, dysaesthesia, paraesthesia, tenderness and/or discomfort.

Spontaneous periods of remission have been observed in 38% of patients, with return to continuous pain after weeks or months.

## **Bibliography**

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## DEFINITION OF TERMS

*Accompanying symptoms:* Symptoms that typically accompany rather than precede or follow headache. In migraine, for example, the most frequent are nausea, vomiting, photophobia and phonophobia; osmophobia, diarrhoea and other symptoms occur more rarely.

*Anorexia:* Lack of appetite and dislike for food to a mild degree.

*Attack of headache (or pain):* Headache (or pain) that builds up, remains at a certain level for minutes to 72 hours, then wanes until it is gone completely.

*Aura:* Early symptoms of an attack of migraine with aura, being the manifestations of focal cerebral dysfunction. The aura typically lasts 20-30 minutes and precedes the headache. See also: *focal symptoms, prodromes, premonitory symptoms, warning symptoms* and *neurological symptoms*.

*Chronic:* In pain terminology, *chronic* denotes persistence over a period longer than 3 months. In headache terminology, it retains this meaning for secondary headache disorders. For primary headache disorders that are more usually episodic (*qv*), *chronic* is used whenever attacks of headache (*qv*) occur on more days than not over a period longer than 3 months. The trigeminal autonomic cephalalgias are the exception: in these disorders, *chronic* is not used until the disorder has been unremitting for more than 1 year.

*Close temporal relation:* This term is used to describe the relation between an organic disorder and headache. Specific temporal relations may be known for disorders of acute onset where causation is likely, but have often not been studied sufficiently. For chronic disorders the temporal relation as well as causation are often very difficult to ascertain.

*Cluster headache attack:* One episode of continuous pain lasting 15-180 minutes.

*Cluster period:* The time during which cluster headache attacks occur regularly and at least once every other day.

*Cluster remission period:* The time during which attacks cease to occur spontaneously and cannot be induced with alcohol or nitroglycerine. To be considered a remission, the attack-free period must exceed 1 month.

*Duration of attack:* Time from onset until termination of an attack of headache (or pain) (*qv*) meeting criteria for a particular headache type or subtype. After migraine or cluster headache, a low-grade non-pulsating headache without accompanying symptoms may persist, but this is not part of the attack and is not included in duration. If the patient falls asleep during an attack and wakes up relieved, duration is until time of awakening. If an attack of migraine is successfully relieved by medication but symptoms recur within 48 hours, these may represent a relapse of the same attack or a new attack. Judgement is required to make the distinction (see Frequency of attacks).

*Episodic:* Recurring and remitting in a regular or irregular pattern of attacks of headache (or pain) (*qv*) of constant or variable duration. Through long usage the term has acquired special meaning in the context of *episodic cluster headache*, referring to the occurrence of cluster periods separated by cluster remission periods (*qv*) rather than to attacks. Similar usage has been adopted in paroxysmal hemicrania.

*Facial pain:* Pain below the orbitomeatal line, above the neck and anterior to the pinnae.

*Focal symptoms:* Symptoms of focal brain (usually cerebral) disturbance such as occur in migraine aura.

*Fortification spectrum:* Angulated, arcuate and gradually enlarging visual hallucination typical of migrainous visual aura.

*Frequency of attacks:* The rate of occurrence of attacks of headache (or pain) (*qv*) per time period (commonly one month). Successful relief of a migraine attack with medication may be followed by relapse within 48 hours. The IHS *Guidelines for Controlled Trials of Drugs in Migraine*, 2<sup>nd</sup> 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 an entire day headache-free.

*Headache:* Pain located above the orbitomeatal line.

*Headache days:* Number of days during an observed period of time (commonly 1 month) affected by headache for any part or the whole of the day.

*Heterophoria:* Latent strabismus.

*Heterotropia:* Manifest strabismus.

*Intensity of pain:* Degree of pain usually expressed in terms of its functional consequence and scored on a verbal 4-point scale: 0, no pain; 1, mild pain, does not interfere with usual activities; 2, moderate pain, inhibits but does not wholly prevent usual activities; 3, severe pain, prevents all activities. It may also be expressed on a visual analogue scale.

*Lancinating:* Brief, electric, shock-like along a root or nerve.

*Neuroimaging:* CT, MRI, PET, SPECT or scintigraphy of the brain.

*New headache:* Any type of headache from which the patient was not previously suffering.

*Not sufficiently validated:* Of doubtful validity as a diagnostic entity judged from the experience of the subcommittee and/or controversy in the literature.

*Nuchal region:* Dorsal (posterior) aspect of upper neck including the region of insertion of neck muscles on the cranium.

*Pericranial muscles:* Neck muscles, muscles of mastication, facial muscles of expression and speech and muscles of the inner ear (tensor tympani, stapedius).

*Phonophobia:* Hypersensitivity to sound, usually causing avoidance.

*Photophobia:* Hypersensitivity to light, usually causing avoidance.

*Premonitory symptoms:* Symptoms preceding and forewarning of a migraine attack by 2-48 hours, occurring before the aura in migraine with aura and before the onset of pain in migraine without aura.. Among the common premonitory symptoms are: fatigue, elation, depression, unusual hunger, craving for certain foods.

*Pressing/tightening:* Pain of a constant quality often compared to an iron band around the head.

*Pressure algometer:* Device to measure the detection threshold or tolerance threshold of pressure-induced pain.

*Previously used term:* A diagnostic term that has been used previously with a similar or identical meaning to the classified term or is subsumed within it. Previously used terms are often ambiguous and/or have been used differently in different countries.

*Prodrome:* This term has been used with different meanings, most often synonymously with premonitory symptoms. It should be avoided in the future.

*Pulsating:* Varying with the heart beat; throbbing.

*Referred pain:* Pain perceived in another area than the one where nociception arises.

*Refraction error:* Myopia, hypermetropia or astigmatism.

*Scintillation:* Visual hallucinations that are bright and fluctuate in intensity, often at approximately 8-10 cycles/second. They are typical of migraine aura.

*Scotoma:* Loss of part(s) of the visual field of one or both eyes. Scotoma may be absolute (no vision) or relative (obscured or reduced vision).

*Stab of pain:* Sudden pain lasting a minute or less (usually a second or less).

*Substance:* Drug, chemical, wine, vapour, etc.

*Teichopsia:* Synonym for fortification spectrum (*qv*).

*Tenderness:* A feeling of discomfort or pain caused by pressure that would not normally be sufficient to cause such sensations.

*Throbbing:* Synonym for pulsating (*qv*).

*Unilateral:* On either the right or the left side, not crossing the mid line. Unilateral headache does not necessarily involve all of the right or left side of the head, but may be frontal, temporal or occipital only. When used for sensory or motor disturbances of migraine aura it includes complete or partial hemidistribution.

*Vasospasm:* Constriction of artery or arterioles to such a degree that tissue perfusion is reduced.

*Warning symptoms:* Previously used term for either aura or premonitory symptoms and therefore ambiguous. It should not be used.

*Zig zag line:* Synonym for fortification spectrum (*qv*).

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