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2nd Edition

Headache Classification Subcommittee of the International Headache Society

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First Headache Classification Subcommittee Members

Jes Olesen, Denmark (*Chairman*)
André Bes, France
Robert Kunkel, USA
James W Lance, Australia
Giuseppe Nappi, Italy
Volker Pfaffenrath, Germany
Frank Clifford Rose, UK
Bruce S Schoenberg, USA (deceased)
Dieter Soyka, Germany
Peer Tfelt-Hansen, Denmark (*Secretary*)
K Michael A Welch, USA
Marcia Wilkinson, UK

Second Headache Classification Subcommittee Members

Jes Olesen, Denmark (*Chairman*)
Marie-Germaine Bousser, France
Hans-Christoph Diener, Germany
David Dodick, USA
Michael First, USA
Peter J Goadsby, UK
Hartmut Göbel, Germany
Miguel JA Lainez, Spain
James W Lance, Australia
Richard B Lipton, USA
Giuseppe Nappi, Italy
Fumihiko Sakai, Japan
Jean Schoenen, Belgium
Stephen D Silberstein, USA
Timothy J Steiner, UK (*Secretary*)

Members of headache classification working groups and advisors

Please address comments and suggestions to the relevant working-group chairmen with a copy to:

Jes Olesen
Chairman of the Classification Subcommittee of the International Headache Society
Professor of Neurology, University of Copenhagen
Department of Neurology, Glostrup Hospital, 2600 Glostrup, Copenhagen, Denmark

1. *Working group on* **Migraine:**
   RB Lipton, USA (*Chairman*); R Daroff, USA; J Haan, The Netherlands; H Massiou, France; J Olesen, Denmark; J Pascual, Spain; BK Rasmussen, Denmark; SD Silberstein, USA; S Solomon, USA; TJ Steiner, UK; D Symon, UK; P Winner, USA.
   Advisors: MK Eriksen, Denmark; P Goadsby, UK; S Graff-Radford, USA; JW Lance, Australia; LL Thomsen, Denmark.
2. *Working group on* **Tension-Type Headache:**
   J Schoenen, Belgium (*Chairman*); EA MacGregor, UK; K Holroyd, USA; R Jensen, Denmark; N Mathew, USA; J Olesen, Denmark; T Paiva, Portugal; R Pothmann, Germany; P Sandor, Switzerland; G Sandrini, Italy.
3. *Working group on* **Cluster Headache and Other Trigeminal Autonomic Cephalalgias:**
   P Goadsby, UK (*Chairman*); F Antonaci, Italy; A Bahra, UK; D Dodick, USA; MS Matharu, UK; A May, Germany; J Olesen, Denmark; L Newman, USA; J Pareja, Spain; D Rothner, USA; M-B Russell, Denmark; M Vincent, Brazil; E Waldenlind, Sweden.
   Advisor: CJ Boes, USA.
4. *Working group on* **Other Primary Headaches:**
   G Nappi, Italy (*Chairman*); P Goadsby, UK; JW Lance, Australia; PO Lundberg, Sweden; IP Martins, Portugal; J Olesen, Denmark; JA Pareja, Spain; NH Raskin, USA; G Sandrini, Italy; TJ Steiner, UK; A Straube, Germany.
   Advisors: L Bonamico, Argentina; S Evers, Germany.
5. *Working group on* **Headache Attributed to Head and/or Neck Trauma:**
   M Lainez, Spain (*Chairman*); R Agosti, Switzerland; F Antonaci, Italy; D Dodick, USA; R Evans, USA; A Mosek, Israel; R Nelson, Canada; D Obelieniene, Lithuania; N Ramadan, USA; J Pascual, Spain; P Sandor, Switzerland.
6. *Working group on* **Headache Attributed to Cranial or Cervical Vascular Disorder:**
   M-G Bousser, France (*Chairman*); J-P Castel, France; A Ducros, France; J Ferro, Portugal; S Kittner, USA; H Mattle, Switzerland; J Olesen, Denmark; S Solomon, USA.
7. *Working group on* **Headache Attributed to Non-Vascular Intracranial Disorder:**
   H-C Diener, Germany (*Chairman*); M-G Bousser, France; D Dodick, USA; A Dowson, UK; P Drummond, Australia; J Gladstein, USA; A Mosek, Israel; R Nelson, Canada; J Olesen, Denmark; N Ramadan, USA; K Ravishankar, India; P Sandor, Switzerland; SD Silberstein, USA; J Swanson, USA; F Taylor, USA; L Watkins, UK.
8. *Working group on* **Headache Attributed to a Substance or its Withdrawal:**
   S Silberstein, USA (*Chairman*); H-C Diener, Germany; M Ferrari, The Netherlands; J Olesen, Denmark; JM Pereira Monteiro, Portugal; J Saper, USA; F Sheftell, USA; P Tfelt-Hansen, Denmark; WB Young, USA.
9. *Working group on* **Headache Attributed to Infection:**
   F Sakai, Japan (*Chairman*); M De Marinis, Italy; A Pradalier, France; D Russell, Norway; N Suzuki, Japan; JR Weber, Germany; KMA Welch, USA.
   Advisor: E Schmutzhard, Austria.

10. *Working group on* **Headache Attributed to Disorder of Homoeostasis:**
D Dodick, USA (*Chairman*); W Becker, Canada; G Bussone, Italy; D Capobianco, USA; FM Cutrer, USA; J Edmeads, Canada; A Kuritzky, Israel; J Olesen, Denmark; A Purdy, Canada; P Spira, Australia.

11. *Working group on* **Headache or Facial Pain Attributed to Disorder of Cranium, Neck, Eyes, Ears, Nose, Sinuses, Teeth, Mouth or Other Facial or Cranial Structures:**
H Göbel, Germany (*Chairman*); M Bakke, Denmark; RW Baloh, USA; N Bogduk, Australia; RB Daroff, USA; S Graff-Radford, USA; J Olesen, Denmark; D Soyka, Germany.
Advisors: H Blumenthal, USA; G Deuschl, Germany; HL Levine, USA; NT Mathew, USA.

12. *Working group on* **Headache Attributed to Psychiatric Disorder:**
MB First, USA (*Chairman*); RM Agosti, Switzerland; S Baskin, USA; N Breslau, USA; V Guidetti, Italy; J Olesen, Denmark; F Sheftell, USA.

13. *Working group on* **Cranial Neuralgias and Central Causes of Facial Pain:**
J Lance, Australia (*Chairman*); D Bowsher, UK; KL Casey, USA; J Olesen, Denmark; T Staehelin-Jensen, Denmark; A Zagami, Australia.

14. *Working group on* **Other Headache, Cranial Neuralgia and Central or Primary Facial Pain:**
D Dodick, USA (*Chairman*); J Olesen, Denmark.

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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
President
International Headache Society
Jes Olesen
Chairman
Headache Classification Committee

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, new criteria such as aggravation by physical activity are 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*, *2nd 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

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, 2nd 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, 2nd 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 on 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 homoeostasis* 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 13. *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*, *2nd 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 patients 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*, *2nd 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, 2nd 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 systemic 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 and WHO ICD-10NA Codes

IHS ICHD-II code	WHO ICD-10NA code	Diagnosis [and aetiological ICD-10 code for secondary headache disorders]
1.	**[G43]**	**Migraine**
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][1]	Migraine-triggered seizure
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
2.	**[G44.2]**	**Tension-type headache (TTH)**
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
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

[1] The additional code specifies the type of seizure.

2.4.2	[G44.28]	Probable frequent episodic tension-type headache
2.4.3	[G44.28]	Probable chronic tension-type headache
3.	**[G44.0]**	**Cluster headache and other trigeminal autonomic cephalalgias**
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 cephalalgia
3.4.1	[G44.08]	Probable cluster headache
3.4.2	[G44.08]	Probable paroxysmal hemicrania
3.4.3	[G44.08]	Probable SUNCT
4.	**[G44.80]**	**Other primary headaches**
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)
5.	**[G44.88]**	**Headache attributed to head and/or neck trauma**
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
6.	**[G44.81]**	**Headache attributed to cranial or cervical vascular disorder**
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]
7.	**[G44.82]**	**Headache attributed to non-vascular intracranial disorder**
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
8.	**[G44.4 or G44.83]**	**Headache attributed to a substance[2] or its withdrawal**
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 induced by 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 medication-overuse headache [F55.2]
8.2.6	[G44.410]	Headache attributed to other medication overuse [code to specify substance]
8.2.7	[G44.41 or G44.83]	Probable medication-overuse headache [code to specify substance]

[2] 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.

8.3	[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]
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 homoeostasis**
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 phaeochromocytoma [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]
11.	**[G44.84]**	**Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures**
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]
12.	**[R51]**	**Headache attributed to psychiatric disorder**
12.1	[R51]	Headache attributed to somatisation disorder [F45.0]
12.2	[R51]	Headache attributed to psychotic disorder [code to specify aetiology]
13.	**[G44.847, G44.848 or G44.85]**	**Cranial neuralgias and central causes of facial pain**
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]
14.	**[R51]**	**Other headache, cranial neuralgia, central or primary facial pain**
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
  1.3.1 Cyclical vomiting
  1.3.2 Abdominal migraine
  1.3.3 Benign paroxysmal vertigo of childhood
1.4 Retinal migraine
1.5 Complications of migraine
  1.5.1 Chronic migraine
  1.5.2 Status migrainosus
  1.5.3 Persistent aura without infarction
  1.5.4 Migrainous infarction
  1.5.5 Migraine-triggered seizure
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[1] fulfilling criteria B–D
B. Headache attacks lasting 4–72 hours (untreated or unsuccessfully treated)[2;3;4]
C. Headache has at least two of the following characteristics:
  1. unilateral location[5;6]
  2. pulsating quality[7]
  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[8]

E. Not attributed to another disorder[9]

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 ≥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 oligaemia 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, $5HT_{1B/D}$ 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[1]

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 1.1 *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[1] and/or unilateral sensory symptoms

2. at least one aura symptom develops gradually over ≥5 minutes and/or different aura symptoms occur in succession over ≥5 minutes
3. each symptom lasts ≥5 and ≤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[2]

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[1] and/or unilateral sensory symptoms
   2. at least one aura symptom develops gradually over ≥5 minutes and/or different aura symptoms occur in succession over ≥5 minutes
   3. each symptom lasts ≥5 and ≤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[2]

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[1] and/or unilateral sensory symptoms
   2. at least one aura symptom develops gradually over ≥5 minutes and/or different aura symptoms occur in succession over ≥5 minutes
   3. each symptom lasts ≥5 and ≤60 minutes
D. Headache does not occur during aura nor follow aura within 60 minutes
E. Not attributed to another disorder[2]

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 when aura begins after age 40, when negative features (*eg*, hemianopia) are predominant, or when 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 ≥5 minutes and/or different aura symptoms occur in succession over ≥5 minutes
   2. each aura symptom lasts ≥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[1]

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 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 ≥5 minutes and/or different aura symptoms occur in succession over ≥5 minutes
   2. each aura symptom lasts ≥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[1]

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 ≥5 minutes and/or different aura symptoms occur in succession over ≥5 minutes
2. each aura symptom lasts ≥5 and ≤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[1]

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 B and 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[1]

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[1]

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

Comments:
Pain 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[1], occurring without warning and resolving spontaneously after minutes to hours
C. Normal neurological examination and 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[1]

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 ≥15 days/month for >3 months
B. Not attributed to another disorder[1,2]

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. When medication overuse is present and fulfils criterion B for any of the subforms of 8.2 *Medication-overuse headache*, it is uncertain whether criterion B for 1.5.1 *Chronic migraine* 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 8.2.7 *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 8.2.7 *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 persisting 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 ≥15 days/month for >3 months
B. Not attributed to another disorder[1] 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.

Bibliography

1. Migraine in general

Bille B. Migraine in childhood and its prognosis. Cephalalgia 1991; 1:71–5.

Blau JN. Migraine prodromes separated from the aura: complete migraine. BMJ 1980; 281:658–60.

Diener HC, Tfelt-Hansen P. Headaches associated with chronic use of substances. In Olesen J, Tfelt-Hansen P, Welch KMA (eds). The Headaches. New York, Haven Press; 1993: pp. 721–7.

Goadsby PJ, Lipton RB, Ferrari MD. Migraine – current understanding and treatment. New Engl J Med 2002; 346:257–70.

Lance JW, Anthony M. Some clinical aspects of migraine. A prospective survey of 500 patients. Arch Neurol 1966; 15:356–61.

Olesen J, Lipton RB. Migraine classification and diagnosis. International Headache Society criteria. Neurology 1994; 44(Suppl 4):6–10.

Rasmussen BK. Epidemiology of headache. Cephalalgia 1995; 15:45–68.

1.1 Migraine without aura

Bille B. A 40-year follow-up of school children with migraine. Cephalalgia 1997; 17:488–91.

Friedman AP, Storch TJC, Merritt HH. Migraine and tension headaches. A clinical study of 2000 cases. Neurology 1954; 4:773–8.

Guidetti V, Galli F. Evolution of headache in childhood and adolescence: an 8-year follow-up. Cephalalgia. 1998; 18:449–54.

Lance JW, Anthony M. Some clinical aspects of migraine. Arch Neurol 1966; 15:356–61.

Lewis DW, Ashwal S, Dahl G et al. Practice parameter: evaluation of children and adolescents with recurrent headaches: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology 2002; 59:490–8.

Maytal J, Young M, Shechter A et al. Pediatric migraine and the International Headache Society (IHS) criteria. Neurology 1997; 48:602–7.

Olesen J. Some clinical features of the acute migraine attack. An analysis of 750 patients. Headache 1978; 18:268–71.

Olesen J, Lipton RB. Migraine classification and diagnosis. International Headache Society criteria. Neurology 1994; 44(Suppl 4):6–10.

Rasmussen BK, Jensen R, Olesen J. A population-based analysis of the diagnostic criteria of the International Headache Society. Cephalalgia 1991; 11:129–34.

Rasmussen BK, Jensen R, Schroll M et al. Interrelations between migraine and tension-type headache in the general population. Arch Neurol 1992, 49:914–8.

Rothner AD, Winner P. Headaches in children and adolescents. In Silberstein SD, Lipton RB, Dalessio DJ. Wolff's Headache and other Head Pain. New York, Oxford University Press 2001: pp. 539–61.

Silberstein SD, Lipton RB. Overview of diagnosis and treatment of migraine. Neurology 1994; 44(Suppl 7):6–16.

Silberstein SD. Migraine symptoms: results of a survey of self-reported migraineurs. Headache 1995; 35:387–96.

Solomon S. Migraine diagnosis and clinical symptomatology. Headache 1994; 34:S8–12.

1.2 Migraine with aura

Blau JN. Migraine prodromes separated from the aura: complete migraine. BMJ 1980; 281:658–60.

Blau JN. Resolution of migraine attacks: sleep and the recovery phase. J Neurol Neurosurg Psychiatr 1982; 45:223–6.

Diamond S, Freitag FG, Prager J et al. Olfactory aura in migraine. N Engl J Med 1985; 312:1390–1.

Goadsby PJ, Lipton RB, Ferrari MD. Migraine–current understanding and treatment. N Engl J Med 2002; 346:257–70.

Goadsby PJ. Migraine, aura, and cortical spreading depression: why are we still talking about it? Ann Neurol 2001; 49:4–6.

Jensen K, Tfelt-Hansen P, Lauritzen M, Olesen J. Classic migraine, a prospective recording of symptoms. Acta Neurol Scand 1986; 73:359–62.

Lauritzen M. Pathophysiology of the migraine aura. The spreading depression theory. Brain 1994; 117:199–210.

Leão AA. Spreading depression of activity in cerebral cortex. J Neurophysiol 1944; 7:359–90.

Manzoni G, Farina S, Lanfranchi M et al. Classic migraine: clinical findings in 164 patients. Eur Neurol 1985; 24:163–9.

Olesen J, Friberg L, Olsen TS et al. Timing and topography of cerebral blood flow, aura, and headache during migraine attacks. Ann Neurol 1990; 28:791–8.

Podoll K, Robinson D. Illusory splitting as visual aura symptom in migraine. Cephalalgia 2000; 20:228–32.

Queiroz LP, Rapoport AM, Weeks RE et al. Characteristics of migraine visual aura. Headache 1997; 37:137–41.

Rasmussen BK, Olesen J. Migraine with aura and migraine without aura: an epidemiological study. Cephalalgia 1992; 12:221–8.

Russell MB, Iversen HK, Olesen J. Improved description of the migraine aura by a diagnostic aura diary. Cephalalgia 1994; 14:107–17.

Russel MB, Olesen J. A nosographic analysis of the migraine aura in a general population. Brain 1996; 119:355–61.

Selby G, Lance JW. Observations on 500 cases of migraine and allied vascular headaches. J Neurol Neurosur Psychiatr 1960; 23:23–32.

Silberstein SD, Young WB. Migraine aura and prodrome. Semin Neurol 1995; 15:175–82.

1.2.2 Typical aura with non-migraine headache

Matharu MJ, Goadsby PJ. Post-traumatic chronic paroxysmal hemicrania (CPH) with aura. Neurology 2001; 56:273–5.

Peres MF, Siow HC, Rozen TD. Hemicrania continua with aura. Cephalalgia 2002; 22:246–8.

Silberstein SD, Niknam R, Rozen TD et al. Cluster headache with aura. Neurology 2000; 54:219–21.

1.2.3 Typical aura without headache

Evans RW, Tietjen GE. Migrainous aura versus transient ischemic attack in an elderly migraineur. Headache 2001; 41:201–3.

Fisher CM. Late-life migraine accompaniments as a cause of unexplained transient ischemic attacks. Can J Neurol Sci 1980; 7:9–17.

Lipton RB, Pfeffer D, Newman LC et al. Headaches in the elderly. J Pain Symptom Manage 1993; 8:87–97.

Russell MB, Olesen J. A nosographic analysis of the migraine aura in a general population. Brain 1996; 119:355–61.
Whitty CVM. Migraine without headache. Lancet 1967; ii:283–85.
Willey RG. The scintillating scotoma without headache. Ann Ophthalmol 1979; 11:581–5.
Ziegler DK, Hanassein RS. Specific headache phenomena: their frequency and coincidence. Headache 1990; 30:152–60.

1.2.4 Familial hemiplegic migraine, and 1.2.5 Sporadic hemiplegic migraine

Carrera P, Stenirri S, Ferrari M et al. Familial hemiplegic migraine: a ion channel disorder. Brain Res Bull 2001, 56:239–41.
De Fusco M, Marconi R, Silvestri L et al. Haploinsufficiency of ATP1A2 encoding the Na/K pump a2 subunit associated with familial hemiplegic migraine type 2. Nat Genet 2003; advance online publication.
Ducros A, Denier C, Joutel A et al. The clinical spectrum of familial hemiplegic migraine associated with mutations in a neuronal calcium channel. N Engl J Med 2001, 345:17–24.
Gomez-Aranda F, Canadillas F, Marti-Masso JF et al. Pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis. A report of 50 cases. Brain 1997; 120:1105–13.
Haan J, Terwindt GM, Ferrari MD. Genetics of migraine. Neurol Clin 1997; 15:43–60.
Kors EE, Terwindt GM, Vermeulen FL, Fitzsimons RB, Jardine PE, Heywood P, Love S, van den Maagdenberg AM, Haan J, Frants RR, Ferrari MD. Delayed cerebral edema and fatal coma after minor head trauma: role of the CACNA1A calcium channel subunit gene and relationship with familial hemiplegic migraine. Ann Neurol 2001; 49:753–60.
Ophoff RA, Terwindt GM, Vergouwe MN et al. Familial hemiplegic migraine and episodic ataxia type-2 are caused by mutations in the Ca2+ channel gene CACNL1A4. Cell 1996; 87:543–52.
Ophoff RA, Terwindt GM, Vergouwe MN et al. Wolff Award 1997. Involvement of a Ca2+ channel gene in familial hemiplegic migraine and migraine with and without aura. Dutch Migraine Genetics Research Group. Headache 1997; 37:479–85.
Staehelin-Jensen T, Olivarius B, Kraft M, Hansen H. Familial hemiplegic migraine. A reappraisal and long-term follow-up study. Cephalalgia 1981; 1:33–9.
Thomsen LL, Ostergaard E, Olesen J, Russell MB. Evidence for a separate type of migraine with aura: Sporadic hemiplegic migraine. Neurology 2003; 60:595–601.
Thomsen LL, Eriksen MK, Roemer SF et al. A population-based study of familial hemiplegic migraine suggests revised diagnostic criteria. Brain 2002; 125:1379–91.

1.2.6 Basilar-type migraine

Bickerstaff ER. Basilar artery migraine. Lancet 1967; i:1517.
Diamond S. Basilar artery migraine. A commonly misdiagnosed disorder. Postgrad Med 1987; 81:45–6.
Erdemoglu AK. Psychogenic basilar migraine. Neurology 1996; 47:302–3.
Kuhn WF, Kuhn SC, Daylida L. Basilar migraine. Eur J Emerg Med 1997; 4:33–8.
Muellbacher W, Mamoli B. Prolonged impaired consciousness in basilar artery migraine. Headache 1994; 34:282–5.
Panayiotopoulos CP. Basilar migraine. Neurology 1991; 41:1707.
Sturzenegger MH, Meienberg O. Basilar artery migraine: a follow-up study of 82 cases. Headache 1985; 25:408–15.
Sudo K, Tashiro K. Psychogenic basilar migraine. Neurology 1996; 46:1786–7.
Swanson JW, Vick NA. Basilar artery migraine. Neurology 1978; 28:782–6.
Thomsen LL, Eriksen MK, Roemer SF, Andersen I, Olesen J, Russell MB. A population-based study of familial hemiplegic migraine suggests revised diagnostic criteria. Brain 2002; 125:1379–91.

1.3.1 Cyclical vomiting

Fleisher DR. Cyclic vomiting syndrome and migraine. J Pediatr 1999; 134:533–5.
Haan J, Kors EE, Ferrari MD. Familial cyclic vomiting syndrome [In Process Citation]. Cephalalgia 2002; 22:552–4.
Li BU. Cyclic vomiting syndrome: age-old syndrome and new insights. Semin Pediatr Neurol 2001; 8:13–21.
Rashed H, Abell TL, Familoni BO et al. Autonomic function in cyclic vomiting syndrome and classic migraine. Dig Dis Sci 1999; 44(8 Suppl):74S–78S.
Welch KM. Scientific basis of migraine: speculation on the relationship to cyclic vomiting. Dig Dis Sci 1999; 44(8 Suppl):26–30.

1.3.2 Abdominal migraine

Abu-Arafeh I, Russel G. Prevalence and clinical features of abdominal migraine compared with those of migraine headache. Arch Dis Child 1995; 72:413–7.
Al-Twaijri WA, Shevell MI. Pediatric migraine equivalents: occurrence and clinical features in practice. Pediatr Neurol 2002; 26:365–8.
Dignan F, Abu-Arafeh I, Russell G. The prognosis of childhood abdominal migraine. Arch Dis Child 2001; 84:415–8.
Farquar HA. Abdominal migraine in children. BMJ 1956; i:1082–5.
Russell G, Abu-Arafeh I, Symon DN. Abdominal migraine: evidence for existence and treatment options. Paediatr Drugs 2002; 4:1–8.

1.3.3 Benign paroxysmal vertigo of childhood

Drigo P, Carli G, Laverda AM. Benign paroxysmal vertigo of childhood. Brain Dev (Netherlands) 2001; 23:38–41.
Dunn DW, Snyder CH. Benign paroxysmal vertigo of childhood. Am J Dis Child 1976; 130:1099–100.
Fenichel GM. Migraine as a cause of benign paroxysmal vertigo of childhood. J Pediatr 1967; 71:114–5.

1.4 Retinal migraine

Carroll D. Retinal migraine. Headache 1970; 10:9–13.
Chronicle EP, Mulleners WM. Visual system dysfunction in migraine: a review of clinical and psychophysical findings. Cephalalgia 1996; 16:525–35.
Hedges TR. Isolated ophthalmic migraine in the differential diagnosis of cerebro-ocular ischemia. Stroke 1976; 7:379–81.
Martin TJ, Corbett JJ. Disorders of the eye. In Silberstein SD, Lipton RB, Dalessio DJ. Wolff's Headache and other Head Pain. New York, Oxford University Press; 2001:459–74.

Troost T, Zagami AS. Ophthalmoplegic migraine and retinal migraine. In Olesen J, Tfelt-Hansen P, Welch KMA. The Headaches. Philadelphia, Lippincott Willians & Wilkins, 2000: pp 511–6.

1.5.1 Chronic migraine

Bigal ME, Sheftell FD, Rapoprt AM, Lipton RB, Tepper SJ. Chronic daily headache in a tertiary care population: correlation between the International Headache Society diagnostic criteria and proposed revisions of criteria for chronic daily headache. Cephalalgia 2002; 22:432–8.

Diamond S. A view of chronic daily headache. Headache Quarterly 2000; 11:177.

Mathew NT, Stubits E, Nigam MR. Transformation of migraine into daily headache: analysis of factors. Headache. 1982; 22:66–8.

Mathew NT, Stubits E, Nigam MP. Transformed or evolutive migraine. Headache 1987; 27:102–6.

Scher AI, Stewart WF, Liberman J, Lipton RB. Prevalence of frequent headache in a population sample. Headache 1998; 38:497–506.

Silberstein SD, Lipton RB, Sliwinski M. Classification of daily and near-daily headaches: field trial of revised IHS criteria. Neurology 1996; 47:871–5.

Silberstein SD, Lipton RB, Solomon S, Mathew N. Classification of daily and near-daily headaches in the headache clinic. Proposed revision to the International Headache Society Criteria. In: Olesen J. Frontiers in Headache Research, vol 4: Headache Classificaton and Epidemiology. New York: Raven Press, 1994: pp 117–26.

Silberstein SD, Lipton RB, Solomon S, Mathew NT. Classification of daily and near-daily headaches: proposed revisions to the IHS criteria. Headache 1994; 34:1–7.

Stewart WF, Scher AI, Lipton RB. Stressful life events and risk of chronic daily headache: results from the frequent headache epidemiology study. Cephalalgia 2001; 21:279.

1.5.2 Status migrainosus

Couch JR, Diamond S. Status migrainosus. Causative and therapeutic aspects. Headache 1983; 23:94–101.

Raskin NH. Treatment of status migrainosus: the American experience. Headache 1990; 30(Suppl 2):550–3.

1.5.3 Persistent aura without infarction

Ambrosini A, de Noordhout AM, Schoenen J. Neuromuscular transmission in migraine patients with prolonged aura. Acta Neurol Belg 2001; 101:166–70.

Bento MS, Esperanca P. Migraine with prolonged aura. Headache 2000; 40:52–3.

Evans RW, Lay CL. A persistent migraine aura. Headache 2000; 40:696–8.

Haan J, Sluis P, Sluis IH, Ferrari MD. Acetazolamide treatment for migraine aura status. Neurology 2000; 55:1588–9.

Haas DC. Prolonged migraine aura status. Ann Neurol 1982; 11:197–9.

Liu GT, Schatz NJ, Galetta SK et al. Persistent positive visual phenomena in migraine. Neurology 1995; 45:664–8.

Luda E, Bo E, Sicuro L et al. Sustained visual aura: a totally new variation of migraine. Headache 1991; 31:582–3.

Rothrock JF. Successful treatment of persistent migraine aura with divalproex sodium. Neurology 1997; 48:261–2.

Smith M, Cros D, Sheen V. Hyperperfusion with vasogenic leakage by fMRI in migraine with prolonged aura. Neurology 2002; 58:1308–10.

1.5.4 Migrainous infarction

Bousser MG, Conard J, Kittner S et al. Recommendations on the risk of ischaemic stroke associated with use of combined oral contraceptives and hormone replacement therapy in women with migraine. The International Headache Society Task Force on Combined Oral Contraceptives & Hormone Replacement Therapy. Cephalalgia 2000; 20:155–6.

Chang CL, Donaghy M, Poulter N. Migraine and stroke in young women: case-control study. The World Health Organisation Collaborative Study of Cardiovascular Disease and Steroid Hormone Contraception. BMJ 1999; 318:13–8.

Connor CCR. Complicated migraine. A study of permanent neurological and visual defects. Lancet 1962; ii:1072–5.

MacGregor EA, Guillebaud J. Combined oral contraceptives, migraine and ischaemic stroke. Clinical and Scientific Committee of the Faculty of Family Planning and Reproductive Health Care and the Family Planning Association. Br J Fam Plann 1998; 24:55–60.

Olesen J, Friberg L, Olsen TS, Andersen AR, Lassen NA, Hansen PE et al. Ischaemia-induced (symptomatic) migraine attacks may be more frequent than migraine-induced ischaemic insults. Brain 1993; 116:187–202.

Rothrock JF, Walicke P, Swenson MR et al. Migrainous stroke. Arch Neurol 1988; 45:63–7.

Tietjen GE. The relationship of migraine and stroke. Neuroepidemiology 2000; 19:13–9.

Tzourio C, Kittner SJ, Bousser MG et al. Migraine and stroke in young women. Cephalalgia 2000; 20:190–9.

1.5.5 Migraine-triggered seizure

Friedenberg S, Dodick DW. Migraine-associated seizure: a case of reversible MRI abnormalities and persistent nondominant hemisphere syndrome. Headache 2000; 40:487–90.

Marks DA, Ehrenberg BL. Migraine-related seizures in adults with epilepsy, with EEG correlation. Neurology. 1993; 43:2476–83.

Ter Berg HW. Migraine-associated seizure: a case of reversible MRI abnormalities and persistent nondominant hemisphere syndrome. Headache 2001; 41:326–8.

Velioglu SK, Ozmenoglu M. Migraine-related seizures in an epileptic population. Cephalalgia 1999; 19:797–801.

1.6 Probable migraine

Granella F, Alessandro RD, Manzoni GC et al. International Headache Society classification: interobserver reliability in the diagnosis of primary headaches. Cephalalgia 1994; 14:16–20.

Rains JC, Penzien DB, Lipchik GL et al. Diagnosis of migraine: empirical analysis of a large clinical sample of atypical migraine (IHS 1.7) patients and proposed revision of the IHS criteria. Cephalalgia 2001, 21:584–95.

Rasmussen BK, Jensen R, Olesen J. A population-based analysis of the diagnostic criteria of the International Headache Society. Cephalalgia 1991; 11:129–34.

Russell MB, Olesen J. Migrainous disorder and its relation to migraine without aura and migraine with aura. A genetic epidemiological study. Cephalalgia 1996; 16:431–5.

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 encour-

ages 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[1]

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 ≥1 but <15 days per month for at least 3 months (≥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[1]

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 ≥15 days per month on average for >3 months (≥180 days per year)[1] 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[2;3]

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, unam-

biguously, 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*, *2nd 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 8.2.7 *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 8.2.7 *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 ≥15 days per month on average for >3 months (≥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*

Bibliography and references

Ad hoc committee on classification of headache. Classification of headache. JAMA 1962; 179:127–8.

Andersson HI, Ejlertsson G, Leden I, Rosenberg C. Chronic pain in a geographically defined general population: studies. Clin J Pain 1993; 9:174–82.

Ashina M, Bendtsen L, Jensen R, Ekman R, Olesen J. Plasma levels of substance P, neuropeptide Y and vasoactive intestinal polypeptide in patients with chronic tension-type headache. Pain 1999; 83:541–7.

Ashina M, Bendtsen L, Jensen R, Olesen J. Nitric oxide-induced headache in patients with chronic tension-type headache. Brain 2000; 23:1830–7.

Ashina M, Bendtsen L, Jensen R, Sakai F, Olesen J. Muscle hardness in patients with chronic tension-type headache: relation to actual headache state. Pain 1999; 79:201–5.

Ashina M, Bendtsen L, Jensen R, Sakai F, Olesen J. Possible mechanisms of glyceryl trinitrate induced headache in patients with chronic tension-type headache. Cephalalgia 2000; 20;919–24.

Ashina M, Bendtsen L, Jensen R, Schifter S, Janssen-Olesen I, Olesen J. Plasma levels of calcitonin gene-related peptide in chronic tension-type headache. Neurology 2000; 55:1335–9.

Ashina M, Bendtsen L, Jensen R, Schifter S, Janssen-Olesen I, Olesen J. Calcitonin gene-related peptide (CGRP) in chronic tension-type headache. Adv Stud Med 2001; 1:54–5.

Ashina M, Bendtsen L, Jensen R, Schifter S, Olesen J. Evidence for increased plasma levels of calcitonin gene-related peptide in migraine outside of attacks. Pain 2000; 86:133–8.

Ashina M, Lassen LH, Bendtsen L, Jensen R, Olesen J. Effect of inhibition of nitric oxide synthase on chronic tension-type headache: a randomised crossover trial. Lancet 1999; 353:287–9.

Ashina M, Lassen LH, Bendtsen L, Jensen R, Sakai F, Olesen J. Possible mechanisms of action of nitric oxide synthase inhibitors in chronic tension-type headache. Brain 1999; 122:1629–35.

Ashina M, Stallknecht B, Bendtsen L et al. In vivo evidence of altered skeletal muscle blood flow in chronic tension-type headache. Brain 2002; 125:320–6.

Bach F, Langemark M, Secher NH, Olesen J. Plasma and cerebrospinal fluid β-endorphin in chronic tension-type headache. Pain 1992; 51:163–8.

Bendtsen L. Central sensitization in tension-type headache – possible pathophysiological mechanisms. Cephalalgia 2000; 20:486–508.

Bendtsen L, Jensen R. Amitriptyline reduces myofascial tenderness in patients with chronic tension-type headache. Cephalalgia 2000; 20;603–10.

Bendtsen L, Jensen R, Brennum J, Arendt-Nielsen L, Olesen J. Exteroceptive suppresion of temporal muscle activity is normal in chronic tension-type headache and not related to actual headache state. Cephalalgia 1996; 16:251–6.

Bendtsen L, Jensen R, Jensen NK, Olesen J. Pressure-controlled palpation: a new technique which increases the reliability of manual palpation. Cephalalgia 1995; 15:205–10.

Bendtsen L, Jensen R, Olesen J. Decreased pain detection and tolerance thresholds in chronic tension-type headache. Arch Neurol 1996; 53:373–6.

Bendtsen L, Jensen R, Olesen J. Qualitatively altered nociception in chronic myofascial pain. Pain 1996; 65:259–64.

Bovim G. Cervicogenic headache, migraine and tension headache. Pressure pain thresholds measurements. Pain 1992; 51:169–73.

Clark GT, Sakai S, Merrill R, Flack VF, McCreary C. Cross-correlation between stress, pain, physical activity, and temporalis muscle EMG in tension-type headache. Cephalalgia 1995; 15:511–8.

Diehr P, Wood RW, Barr V, Wolcott B, Slay L, Tompkins RK. Acute headaches: presenting symptoms and diagnostic rules to identify patients with tension and migraine headache. J Chron Dis 1981; 34:147–58.

Drummond PD. Scalp tenderness and sensitivity to pain in migraine and tension headache. Headache 1987; 27:45–50.

Friedman AP. Characteristics of tension-type headache. A profile of 1420 cases. Psychosomatics 1979; 20:451–61.

Friedman AP, von Storch TJC, Merritt HH. Migraine and tension headaches: a clinical study of two thousand cases. Neurology 1954; 4:773–8.

Göbel H, Petersen-Braun M, Soyka D. The epidemiology of headache in Germany: a nationwide survey of a representative sample on the basis of the headache classification of the International Headache Society. Cephalalgia 1994; 14:97–106.

Göbel H, Weigle L, Kropp P, Soyka D. Pain sensitivity and pain reactivity of pericranialmuscles in migraine and tension-type headache. Cephalalgia 1992; 12:142–51.

Hatch JP, Moore PJ, Cyr-Provost M, Boutros NN, Seleshi E, Borcherding S. The use of electromyography and muscle palpation in the diagnosis of tension-type headache with and without pericranial muscle involvement. Pain 1992; 49:175–8.

Headache Classification Committee of the International Headache Society. Classification and Diagnostic criteria for headache disorders, cranial neuralgias and facial pain. Cephalalgia 1988; 8(suppl 7):1–96.

Jensen K. Quantification of tenderness by palpation and use of pressure algometer. Adv Pain Res Ther 1990; 17:165–81.

Jensen R. Mechanisms of spontaneous tension-type headaches: an analysis of tenderness, pain thresholds and EMG. Pain 1996; 64:251–6.

Jensen R. Pathophysiological mechanisms of tension-type headache: a review of epidemiological and experimental studies. Cephalalgia 1999; 19:602–21.

Jensen R. The tension-type headache alternative. Peripheral pathophysiological mechanisms. Cephalalgia 1999; 19(suppl 25):9–10.

Jensen R. Tension-type headache. Cur Treat Opt Neurol 2001; 3:169–80.

Jensen R. Mechanisms of tension-type headache. Cephalalgia 2001; 21:786–9.

Jensen R. Chronic tension-type headache. Adv Stud Med 2001; 1(11):449–50.

Jensen R, Bendtsen L, Olesen J. Muscular factors are of importance in tension-type headache. Headache 1998; 38:10–17.

Jensen R, Fuglsang-Frederiksen A. Quantitative surface EMG of pericranial muscles. Relation to age and sex in a general population. Electroenceph Clin Neurophysiol 1994; 93:175–83.

Jensen R, Fuglsang-Frederiksen A, Olesen J. Quantitative surface EMG of pericranial muscles in headache. A population study. Electroenceph Clin Neurophysiol 1994; 93:335–44.

Jensen R, Olesen J. Initiating mechanisms of experimentally induced tension-type headache. Cephalalgia 1996; 16:175–82.

Jensen R, Olesen J. Tension-type headache: an update on mechanisms and treatment. Cur Opin Neurol 2000; 13:285–9.

Jensen R, Rasmussen BK. Muscular disorders in tension-type headache. Cephalalgia 1996; 16:97–103.

Jensen R, Rasmussen BK, Pedersen B, Lous I, Olesen J. Cephalic muscle tenderness and pressure pain threshold in a general population. Pain 1992; 48:197–203.

Jensen R, Rasmussen BK, Pedersen B, Olesen J. Muscle tenderness and pressure pain thresholds in headache. A population study. Pain 1993; 52:193–9.

Kunkel RS. Diagnosis and treatment of muscle contraction (tension-type) headaches. Med Clin North Am 1991; 75:595–603.

Langemark M, Bach FW, Ekman R, Olesen J. Increased cerebrospinal fluid met-enkephalin immunoreactivity in patients with chronic tension-type headache. Pain 1995; 63:103–7.

Langemark M, Bach FW, Jensen TS, Olesen J. Decreased nociceptive flexion reflex threshold in chronic tension-type headache. Arch Neurol 1993; 50:1061–4.

Langemark M, Jensen K. Myofascial mechanisms of pain. In: Olesen J, Edvinson EL (Eds). Basic Mechanisms of Headache. Elsevier, Amsterdam 1988, 331–341.

Langemark M, Jensen K, Jensen TS, Olesen J. Pressure pain thresholds and thermal nociceptive thresholds in chronic tension-type headache. Pain 1989; 38:203–10.

Langemark M, Jensen K, Olesen J. Temporal muscle blood flow in chronic tension-type headache. Arch Neurol 1990; 47:654–8.

Langemark M, Olesen J. Pericranial tenderness in tension headache. Cephalalgia 1987; 7:249–55.

Langemark M, Olesen J, Poulsen DL, Bech P. Clinical characteristics of patients with tension-type headache. Headache 1988; 28:590–6.

Lipchik GL, Holroyd KA, France CR, Kvaal SA, Segal D, Cordingley GE, Rokicki LA, McCool HR. Central and peripheral mechanisms in chronic tension-type headache. Pain 1996; 64;467–75.

Lipton RB, Schwartz BS, Stewart WF. Epidemiology of tension-type headache. Cephalalgia 1997; 17:233.

Lous I, Olesen J. Evaluation of pericranial tenderness and oral function in patients with common migraine, muscle contraction headache and combination headache. Pain 1982; 12:385–93.

Marcus DA. Serotonin and its role in headache pathogenesis and treatment. Clin J Pain 1993; 9:159–67.

Martin PR, Mathews AM. Tension headaches: psychosociological investigation and treatment. J Psychosom Res 1978; 22:389–99.

Mennel J. Myofascial trigger points as a cause of headaches. J Manipulative Physiol Therap 1989; 12:308–13.

Mense S. Nociception from skeletal muscle in relation to clinical muscle pain. Pain 1993; 54:241–89.

Olesen J. Clinical and pathophysiological observations in migraine and tension-type headache explained by integration of vascular, supraspinal and myofascial inputs. Pain 1991; 46:125–32.

Olesen J, Jensen R. Getting away from simple muscle contraction as a mechanism of tension-type headache. Pain 1991; 46:123–4.

Östergaard S, Russell MB, Bendtsen L, Olesen J. Increased familial risk of chronic tension-type headache. BMJ 1997; 314:1092–3.

Passchier J, Van der Helm-Hylkema H, Orlebeke JF. Psychophysiological characteristics of migraine and tension headache patients. Differential effects of sex and pain state. Headache 1984; 24:131–9.

Peterson AL, Talcott GW, Kelleher WJ, Haddock CK. Site specificity of pain and tension in tension-type headaches. Headache 1995; 35:89–92.

Pikoff H. Is the muscular model of headache still viable? A review of conflicting data. Headache 1984; 24:186–98.

Quinter JL, Cohen ML. Referred pain of peripheral nerve origin: An alternative to the myofascial pain construct. Clin J Pain 1994; 10:243–51.

Rasmussen BK. Migraine and tension-type headache in a general population: precipitating factors, female hormones, sleep pattern and relation to lifestyle. Pain 1993; 53:65–72.

Rasmussen BK, Jensen R, Olesen J. A population-based analysis of the criteria of the International Headache Society. Cephalalgia 1991; 11:129–34.

Rasmussen BK, Jensen R, Olesen J. Questionnaire versus clinical interview in the diagnosis of headache. Headache 1991; 31:290–5.

Rasmussen BK, Jensen R, Olesen J. Impact of headache on sickness absence and utilisation of medical services: a Danish population study. J Epidemiol Community Health 1992; 46:443–6.

Rasmussen BK, Jensen R, Schroll M, Olesen J. Epidemiology

of headache in a general population. A prevalence study. J Clin Epidemiol 1991; 44:1147–57.

Sakai F, Ebihara S, Akiyama M, Horikawa M. Pericranial muscle hardness in tension-type headache. A non-invasive measurement method and its clinical application. Brain 1995; 118:523–31.

Sandrini G, Antonaci F, Pucci E, Bono G, Nappi G. Comparative study with EMG, pressure algometry and manual palpation in tension-type headache and migraine. Cephalalgia 1994; 14:451–7.

Sandrini G, Ruiz L, Alfonsi E, Cravera, Nappi G. Antinociceptive system in primary headache disorders: A neurophysiological approach. In: Nappi G et al.(Eds.) Headache and Depression: Serotonin pathways as a common clue. Raven Press, New York 1991; pp 67–78.

Simons DG. Clinical and etiological update of myofascial pain from trigger points. J Musculoskeletal Pain 1996; 4:93–121.

Schoenen J, Bottin D, Hardy F, Gerard P. Cephalic and extracephalic pressure pain thresholds in chronic tension-type headache. Pain 1991; 47:145–9.

Schoenen J, Gerard P, De Pasqua V, Juprelle M. EMG activity in pericranial muscles during postural variation and mental activity in healthy volunteers and patients with chronic tension-type headache. Headache 1991; 31:321–4.

Schoenen J, Gerard P, De Pasqua V, Sianard-Gainko J. Multiple clinical and paraclinical analyses of chronic tension-type headache associated or unassociated with disorder of pericranial muscles. Cephalalgia 1991; 11:135–9.

Schoenen J, Jamart B, Gerard P, Lenarduzzi P, Delwaide PJ. Exteroceptive suppression of temporalis muscle activity in chronic headache. Neurology 1987; 37:1834–6.

Solomon S, Lipton RB, Newman LC. Clinical features of chronic daily headache. Headache 1992; 32:325–9.

Travell JG, Simons DG. Myofascial pain and dysfunction. The trigger point manual. Williams & Wilkins, Baltimore 1983.

Ulrich V, Russel MB, Jensen R, Olesen J. A comparision of tension-type headache in migraineurs and in non-migraineurs: a population-based study. Pain 1996; 67:501–6.

Wang W, Schoenen J. Reduction of temporalis exteroceptive suppression by peripheral electrical stimulation in migraine and tension-type headaches. Pain 1994; 59:327–34.

Zwart JA, Sand T. Exteroceptive suppression of temporalis muscle activity: a blind study of tension-type headache, migraine and cervicogenic headache. Headache 1995; 5:338–43.

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
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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).

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[1]
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[2]
E. Not attributed to another disorder[3]

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[1] and separated by pain-free remission periods of ≥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 s*econdary 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[1]
F. Not attributed to another disorder[2]

Notes:
1. In order to rule out incomplete response, indomethacin should be used in a dose of ≥150 mg daily orally or rectally, or ≥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 1 month or longer.

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 ≥1 month

3.2.2 Chronic paroxysmal hemicrania (CPH)

Description:
Attacks of paroxysmal hemicrania 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 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[1]

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

References

General

Goadsby PJ. Pathophysiology of cluster headache: a trigeminal autonomic cephalgia. Lancet Neurology 2002; 1:37–43.

Goadsby PJ, Lipton RB. A review of paroxysmal hemicranias, SUNCT syndrome and other short-lasting headaches with autonomic features, including new cases. Brain 1997; 120:193–209.

May A, Goadsby PJ. The trigeminovascular system in humans: pathophysiological implications for primary headache syndromes of the neural influences on the cerebral circulation. Journal of Cerebral Blood Flow and Metabolism 1999; 19:115–27.

3.1 Cluster headache

Alberca R, Ochoa JJ. Cluster tic syndrome. Neurology 1994; 44:996–9.

Bahra A, May A, Goadsby PJ. Diagnostic patterns in cluster headache. In: Olesen J, Goadsby PJ, editors. Cluster Headache and Related Conditions. Oxford: Oxford University Press 1999: pp. 61–65.

Bahra A, May A, Goadsby PJ. Cluster headache: a prospective clinical study in 230 patients with diagnostic implications. Neurology 2002; 58:354–61.

Bing R. Uber traumatische Erythromelalgie und Erthroprosopalgie. Nervenarzt 1930; 3:506–12.

de Fine Olivarius B. Hemicrania neuralgiformis chronica (Chronic migrainous neuralgia). Quoted by Sjaastad O, ed. Proceedings of the Scandinavian Migraine Society Annual Meeting 1971: p. 8.

Ekbom K. Ergotamine tartrate orally in Horton's 'histaminic cephalalgia' (also called Harris's ciliary neuralgia). Acta Psychiatrica Scandinavia 1947; 46:106.

Ekbom K. Nitroglycerin as a provocative agent in cluster headache. Archives of Neurology 1968; 19:487–93.

Eulenberg A. Lehrbuch der Nervenkrankheiten. 2nd edn. Berlin: Hirschwald 1878.

Harris W. Ciliary (migrainous) neuralgia and its treatment. British Medical Journal 1936; 1:457–60.

Horton BT. Histaminic cephalgia. Lancet 1952; ii:92–8.

Kudrow L. Cluster headache: Mechanisms and Management. Oxford: Oxford University Press 1980.

Manzoni GC. Gender ratio of cluster headache over the years: a possible role of changes in lifestyle. Cephalalgia 1998; 18:138–42.

Manzoni GC, Micieli G, Granella F, Tassorelli C, Zanferrari C,

Cavallini A. Cluster headache-course over ten years in 189 patients. Cephalalgia 1991; 11:169–74.
Manzoni GC, G.Terzano M, Bono G, Micieli G, Martucci N, Nappi G. Cluster headache – clinical findings in 180 patients. Cephalalgia 1983; 3:21–30.
May A, Bahra A, Buchel C, Frackowiak RSJ, Goadsby PJ. Hypothalamic activation in cluster headache attacks. Lancet 1998; 351:275–8.
Mulleners WM, Verhagen WIM. Cluster-tic syndrome. Neurology 1996; 47:302.
Pascual J, Berciano J. Relief of cluster-tic syndrome by the combination of lithium and carbamazepine. Cephalalgia 1993; 13:205–6.
Romberg MH. Lehrbuch der Nervenkrankheiten des Menschen. Berlin: Dunker 1840.
Russell MB, Andersson PG, Thomsen LL, Iselius L. Cluster headache is an autosomal dominantly inherited disorder in some families: a complex segregation analysis. Journal of Medical Genetics 1995; 32:954–6.
Sjostrand C, Waldenlind E, Ekbom K. A follow up study of 60 patients after an assumed first period of cluster headache. Cephalalgia 2000; 20:653–7.
Sluder G. The syndrome of sphenopalatine ganglion neurosis. American Journal of Medicine 1910; 140:868–78.
Solomon S, Apfelbaum RI, Guglielmo KM. The cluster-tic syndrome and its surgical therapy. Cephalalgia 1985; 5:83–9.
Torelli P, Cologno D, Cademartiri C, Manzoni GC. Application of the International Headache Society classification criteria in 652 cluster headache patients. Cephalalgia 2001; 21:145–50.
Vail HH. Vidian neuralgia. Ann Otol Rhinol Laryngol 1932; 41:837–56.
Watson P, Evans R. Cluster-tic syndrome. Headache 1985; 25:123–6.

3.2 Paroxysmal hemicrania

Antonaci F, Pareja JA, Caminero AB, Sjaastad O. Chronic paroxysmal hemicrania and hemicrania continua. Parenteral indomethacin: the 'Indotest'. Headache 1998; 38:122–8.
Antonaci F, Sjaastad O. Chronic paroxysmal hemicrania (CPH): a review of the clinical manifestations. Headache 1989; 29:648–56.
Broeske D, Lenn NJ, Cantos E. Chronic paroxysmal hemicrania in a young child: possible relation to ipsilateral occipital infarction. Journal of Child Neurology 1993; 8:235–6.
Caminero AB, Pareja JA, Dobato JL. Chronic paroxysmal hemicrania-tic syndrome. Cephalalgia 1998; 18:159–61.
Hannerz J. The second case of chronic paroxysmal hemicrania-tic syndrome [Editorial Comment]. Cephalalgia 1998; 18:124.
Kudrow DB, Kudrow L. Successful aspirin prophylaxis in a child with chronic paroxysmal hemicrania. Headache 1989; 29:280–1.
Sjaastad O, Dale I. Evidence for a new (?) treatable headache entity. Headache 1974;14:105–108.
Zukerman E, Peres MFP, Kaup AO, Monzillo PH, Costa AR. Chronic paroxysmal hemicrania-tic syndrome. Neurology 2000; 54:1524–6.

3.3 SUNCT

Benoliel R, Sharav Y. Trigeminal neuralgia with lacrimation or SUNCT syndrome? Cephalalgia 1998; 18:85–90.
Bouhassira D, Attal N, Esteve M, Chauvin M. SUNCT syndrome. A case of transformation from trigeminal neuralgia. Cephalalgia 1994; 14:168–70.
Bussone G, Leone M, Volta GD, Strada L, Gasparotti R. Short-lasting unilateral neuralgiform headache attacks with tearing and conjunctival injection: the first symptomatic case. Cephalalgia 1991; 11:123–7.
De Benedittis G. SUNCT syndrome associated with cavernous angioma of the brain stem. Cephalalgia 1996; 16:503–6.
Ferrari MD, Haan J, van Seters AP. Bromocriptine-induced trigeminal neuralgia attacks in a patient with pituitary tumor. Neurology 1988; 38:1482–4.
Goadsby PJ, Lipton RB. A review of paroxysmal hemicranias, SUNCT syndrome and other short-lasting headaches with autonomic features, including new cases. Brain 1997; 120:193–209.
Goadsby PJ, Matharu MS, Boes CJ. SUNCT syndrome or trigeminal neuralgia with lacrimation. Cephalalgia 2001; 21:82–3.
Levy MJ, Matharu MS, Goadsby PJ. Prolactinomas, dopamine agonist and headache: two case reports. European Journal of Neurology 2003; 10:169–74.
Massiou H, Launay JM, Levy C, El Amran M, Emperauger B, Bousser M-G. SUNCT syndrome in two patients with prolactinomas and bromocriptine-induced attacks. Neurology 2002; 58:1698–9.
Matharu MS, Levy MJ, Merry RT, Goadsby PJ. SUNCT syndrome secondary to prolactinoma. J. Neurol. Neurosurg. Psychiatry 2003; in press.
Morales F, Mostacero E, Marta J, Sanchez S. Vascular malformation of the cerebellopontine angle associated with SUNCT syndrome. Cephalalgia 1994; 14:301–2.
Moris G, Ribacoba R, Solar DN, Vidal JA. SUNCT syndrome and seborrheic dermatitis associated with craneosynostosis. Cephalalgia 2001; 21:157–9.
Pareja JA, Sjaastad O. SUNCT syndrome. A clinical review. Headache 1997; 37:195–202.
Penart A, Firth M, Bowen JRC. Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) following presumed dorsolateral brainstem infarction. Cephalalgia 2001; 21:236–9.
Sjaastad O, Saunte C, Salvesen R, Fredriksen TA, Seim A, Roe OD et al. Shortlasting unilateral neuralgiform headache attacks with conjunctival injection, tearing, sweating, and rhinorrhea. Cephalalgia 1989; 9:147–56.
ter Berg HWM, Goadsby PJ. Significance of atypical presentation of symptomatic SUNCT: a case report. J Neurol Neurosurg Psychiat 2001; 70:244–46.

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[1]

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[1]

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[1]

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 fulfilling 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 fulfilling criterion B
B. Occurs at orgasm
C. Not attributed to another disorder[1]

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[1]

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[1]
D. Not attributed to another disorder[2]

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[1]

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 for >3 months fulfilling criteria B–D
B. Headache is daily and unremitting from onset or from <3 days from onset[1]
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 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[2]

Notes:
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 8.2.7 *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 entity, especially studies comparing 4.8 *New daily-persistent headache* with 2.3 *Chronic tension-type headache*.

Bibliography

4.1 Primary stabbing headache

Ammache Z, Graber M, Davis P. Idiopathic stabbing headache associated with mononuclear visual loss. Arch Neurol 2000; 57:745–6.

Dangond F, Spierings EL. Idiopathic stabbing headaches lasting a few seconds. Headache 1993; 33:257–8.

Martins IP, Parreira B, Costa I. Extratrigeminal ice-pick status. Headache 1995; 35:107–10.

Pareja JA, Kruszewski P, Caminero AB. SUNCT syndrome versus idiopathic stabbing headache (jabs and jolts syndrome). Cephalalgia 1999; 19(Suppl 25):46–8.

Pareja JA, Ruiz J, de Isla C, al-Sabbab H, Espejo J. Idiopathic stabbing headache (jabs and jolts syndrome). Cephalalgia 1996; 16:93–6.

Sjaastad O, Batnes J, Haugen S. The Vågå Study: an outline of the design. Cephalalgia 1999; 19(Suppl 25):24–30.

Soriani G, Battistella PA, Arnaldi C, De Carlo L, Cemetti R, Corra S, Tosato G. Juvenile idiopathic stabbing headache. Headache 1996; 36:565–7.

4.2 Primary cough headache

Calandre L, Hernandez-Lain A, Lopez-Valdes E. Benign Valsalva's maneuver-related headache: an MRI study of six cases. Headache 1996; 36:251–3.

Ertsey C, Jelencsik I. Cough headache associated with Chiari type-I malformation: responsiveness to indomethacin. Cephalalgia 2000; 20:518–20.

Pascual J, Berciano J. Experience in the diagnosis of headaches that start in elderly people. J Neurol Neurosurg Psychiatry 1994; 57:1255–7.

Pascual J, Iglesias F, Oterino A, Vazquez-Barquero A, Berciano J. Cough, exertional, and sexual headaches: an analysis of 72 benign and symptomatic cases. Neurology 1996; 46: 1520–4.

Perini F, Toso V. Benign cough 'cluster' headache. Cephalalgia 1998; 18:493–4.

Raskin NH. The cough headache syndrome: treatment. Neurology 1995; 45:1784.

Smith WS, Messing RO. Cerebral aneurysm presenting as cough headache. Headache 1993; 33:203–4.

4.3 Primary exertional headache

Edis RH, Silbert PL. Sequential benign sexual headache and exertional headache (letter). Lancet 1988; 30: 993.

Green MW. A spectrum of exertional headaches. Headache 2001; 4:1085–92.

Heckmann JG, Hilz MJ, Muck-Weymann M, Neundorfer B. Benign exertional headache/benign sexual headache: a disorder of myogenic cerebrovascular autoregulation? Headache 1997; 37:597–8.

Lance JW, Lambros J. Unilateral exertional headache as a symptom of cardiac ischemia. Headache 1998; 38:315–6.

Lipton RB, Lowenkopf T, Bajwa ZH, Leckie RS, Ribeiro S, Newman LC, Greenberg MA. Cardiac cephalgia: a treatable form of exertional headache. Neurology 1997; 49:813–6.

Pascual J, Iglesias F, Oterino A, Vazquez-Barquero A, Berciano J. Cough, exertional, and sexual headaches: an analysis of 72 benign and symptomatic cases. Neurology 1996; 46: 1520–4.

Silbert PL, Edis RH, Stewart-Wynne EG, Gubbay SS. Benign vascular sexual headache and exertional headache: interrelationships and long term prognosis. J Neurol Neurosurg Psychiatry 1991; 54:417–21.

4.4 Primary headache associated with sexual activity

D'Andrea G, Granella F, Verdelli F. Migraine with aura triggered by orgasm. Cephalalgia 2002; 22:485–6.

Jacome DE. Masturbatory-orgasmic extracephalic pain. Headache 1998; 38:138–41.

Kumar KL, Reuler JB. Uncommon headaches: diagnosis and treatment. J Gen Int Med 1993; 8:333–41.

Lance JW. Headaches related to sexual activity. J Neurol Neurosurg Psychiat 1976; 39:1226—30.

Lundberg PO, Osterman PO. Intercourse and headache. In: Genazzani AR, Nappi G, Facchinetti F, Martignoni E (eds). Pain and reproduction. Parthenon Publishing 1988: pp 149–53.

Maliszewski M, Diamond S, Freitag FG. Sexual headache occuring in cluster headache patients. Clinical J Pain 1989; 5:45–7.

Pascual J, Iglesias F, Oterino A, Vazquez-Barquero A, Berciano J. Cough, exertional, and sexual headaches: an analysis of 72 benign and symptomatic cases. Neurology 1996; 46: 1520–4.

Robbins L. Masturbatory-orgasmic extracephalic pain. Headache 1994; 34:214–16.

4.5 Hypnic headache

Arjona JA, Jimenez-Jimenez FJ, Vela-Bueno A, Tallon-Barranco A. Hypnic Headache associated with stage 3 slow wave sleep. Headache 2000; 40:753–4.

Bruni O, Galli F, Guidetti V. Sleep hygiene and migraine in children and adolescents. Cephalalgia 1999; 19(Suppl 25):57–9.

Centonze V, D'Amico D, Usai S, Causarano V, Bassi A, Bussone G. First Italian case of hypnic headache, with literature review and discussion of nosology. Cephalalgia 2001; 21:71–4.

Dodick DW. Polysomnography in hypnic headache syndrome. Headache 2000; 40:748–52.

Dodick DW, Jones JM, Capobianco DJ. Hypnic headache: another indomethacin-responsive headache syndrome? Headache 2000; 40:830–5.

Dodick DW, Mosek AC, Campbell IK. The hypnic ('alarm clock') headache syndrome. Cephalalgia 1998; 18:152–6.

Ghiotto N, Sances G, Di Lorenzo G, Trucco M, Loi M, Sandrini G, Nappi G. Report of eight new cases of hypnic headache and a mini-review of the literature. Funct Neurol 2002; 17:211–9.

Gould JD, Silberstein SD. Unilateral hypnic headache: a case study. Neurology 1997; 49:1749–51.

lvanez V, Soler R, Barreiro P. Hypnic headache syndrome: a case with good response to indomethacin. Cephalalgia 1998; 18:225–6.

Martins IP, Gouveia RG. Hypnic headache and travel across time zones: a case report. Cephalalgia 2001; 21:928–31.

Morales-Asin F, Mauri JA, Iniguez C, Espada F, Mostacero E. The hypnic headache syndrome: report of three new cases. Cephalalgia 1998; 18:157–8.

Newman LC, Lipton RB, Solomon S. The hypnic headache syndrome: a benign headache disorder of the elderly. Neurology 1990; 40:1904–5.

Raskin NH. The hypnic headache syndrome. Headache 1988; 28:534–6.

Ravishankar K. Hypnic headache syndrome. Cephalalgia 1998; 18:358–9.

4.6 Primary thunderclap headache

Bassi P, Bandera R, Loiero M, Tognoni G, Mangoni A. Warning signs in subarachnoid hemorrhage: a cooperative study. Acta Neurol Scand 1991; 84:277–81.

Dodick DW, Brown RD, Britton JW, Huston J. Nonaneurysmal thunderclap headache with diffuse, multifocal, segmental and reversible vasospasm. Cephalalgia 1999; 19:118–23.

Garg RK. Recurrent thunderclap headache associated with reversible vasospasm causing stroke. Cephalalgia 2001; 21:78–9.

Landtblom AM, Fridriksson S, Boivie J, Hillman J, Johansson G, Johansson I. Sudden onset headache: a prospective study of features incidence and causes. Cephalalgia 2002; 22:354–60.

Linn FHH, Rinkel GJE, Algra A, van Gijn J. Headache characteristics in subarachnoid haemorrhage and benign thunderclap headache. J Neurol Neurosurg Psychiat 1998; 65:791–3.

Linn FHH, Rinkel GJE, Algra A, van Gijn J. Follow-up of idiopathic thunderclap headache in general practice. J Neurol 1999; 246:946–8.

Linn FHH, Wijdicks EFM. Causes and management of thunderclap headache: a comprehensive review. The Neurologist 2002; 8:279–89.

Markus HS. A prospective follow-up of thunderclap headache mimicking subarachnoid haemorrhage. J Neurol Neurosurg Psychiat 1991; 54:1117–25.

Mauriño J, Saposnik G, Lepera S, Rey RC, Sica RE. Multiple simultaneous intracerebral haemorrhages. Arch Neurol 2001; 58:629–32.

Nowak DA, Rodiek SO, Henneken S, Zinner J, Schreiner R, Fuchs H-H, Topka H. Reversible segmental cerebral vasoconstriction (Call-Fleming syndrome): are calcium channel inhibitors a potential treatment option? Cephalalgia 2003; 23:218–22.

Sturm JW, Macdonell RAL. Recurrent thunderclap headache associated with reversible intracerebral vasospasm causing stroke. Cephalalgia 2000; 20:132–5.

Slivka A, Philbrook B. Clinical and angiographic features of thunderclap headache. Headache 1995; 35:1–6.

Wijdicks EFM, Kerkhoff H, van Gjin J. Cerebral vasospasm and unruptured aneurysm in thunderclap headache. Lancet 1988; ii:1020.

Witham TF, Kaufmann AM. Unruptured cerebral aneurysm producing a thunderclap headache. Am J Emergency Med 2000; 1:88–90.

4.7 Hemicrania continua

Antonaci F, Pareja JA, Caminero AB, Sjaastad O. Chronic paroxysmal hemicrania and hemicrania continua: anaesthetic blockades of pericranial nerves. Funct Neurol 1997; 1:11–5.

Antonaci F, Pareja JA, Caminero AB, Sjaastad O. Chronic paroxysmal hemicrania and hemicrania continua. Parenteral indomethacin: the 'Indotest'. Headache 1998; 8:235–6.

Bordini C, Antonaci F, Stovner LJ, Schrader H, Sjaastad O. 'Hemicrania continua': a clinical review. Headache 1991; 31: 20–6.

Newman LC, Lipton RB, Solomon S. Hemicrania continua: ten new cases and a review of the literature. Neurology 1994; 44:2111–4.

Pareja J, Antonaci F, Vincent M. The hemicrania continua diagnosis. Cepahalalgia 2002; 7:563–4.

Pareja J, Vincent M, Antonaci F, Sjaastad O. Hemicrania continua: diagnostic criteria and nosologic status. Cepahalalgia 2001; 9:874–7.

Sjaastad O, Antonaci F. Chronic paroxysmal hemicrania (CPH) and hemicrania continua: transition from one stage to another. Headache 1993; 33:551–4.

Sjaastad O, Antonaci F. A piroxicam derivative partly effective in chronic paroxysmal hemicrania and hemicrania continua. Headache 1995; 35:549–50.

Sjaastad O, Spierings EL. Hemicrania continua: another headache absolutely responsive to indomethacin. Cephalalgia 1984; 4:65–70.

4.8 New daily-persistent headache

Evans RW, Rozen TD. Etiology and treatment of new daily persistent headache. Headache 2001; 4:830–2.

Goadsby PJ, Boes C. New daily persistent headache. J Neurol Neurosurg Psychiat 2002; 72(Suppl 2):ii6–ii9.

Li D, Rozen TD. The clinical characterisation of new daily persistent headache. Cephalalgia 2002; 22:66–9.

Silberstein SD, Lipton RB, Solomon S, Mathew NT. Classification of daily and near daily headaches: proposed revisions to the IHS-criteria. Headache 1994; 34:1–7.

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

Headache attributed to a substance or its withdrawal

Headache attributed to infection

Headache attributed to disturbance of homoeostasis

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[1,2] 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[3]

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 8.2.7 *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) ≥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 ≥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) ≥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 ≥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[1]
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[1]
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.

Bibliography

5.1, 5.2 Acute and chronic post-traumatic headache

Branca B, Giordani B, Lutz T, Saper JR. Self-report of cognition and objective test performance in posttraumatic headache. Headache 1996; 36:300–6.

Duckro PN, Chibnall JT, Tomazic TJ. Anger, depression, and disability: a path analysis of relationships in a sample of chronic posttraumatic headache patients. Headache 1995; 35:7–9.

Editorial. Definition of mild traumatic brain injury. J Head Trauma Rehabil 1993; 6:86–7.

Elkind AH. Headache and head trauma. Clin J Pain 1989; 5:77–87.

Evans RW. Some observations on whiplash injuries. Neurol Clin 1992; 10:975–98.

Gfeller JD, Chibnall JT, Duckro PN. Postconcussion symptoms and cognitive functioning in posttraumatic headache patients. Headache 1994; 34:503–7.

Gilkey SJ, Ramadan NM, Aurora TK, Welch KM. Cerebral blood flow in chronic posttraumatic headache. Headache 1997; 37:583–7.

Haas DC. Chronic postraumatic headaches classified and compared with natural headaches. Cephalalgia 1996; 16:486–93.

Hachinski VV. Posttraumatic headache. Arch Neurol 2000; 57:1780.

Jiménez MD. Cefalea postraumática. In: Cefaleas secundarias. F. Titus. C. Targa, JM Láinez, eds. Ediciones Ergon. Madrid. 1995: pp 17–36.

Keidel M, Ramadan N. Acute posttraumatic headache. In: The Headaches. Olesen J, Tfelt-Hansen P, Welch KMA (eds). Lippincott & Wilkins. Philadelphia 2000: pp 765–70.

Kelly JP, Rosenberg JH. Diagnosis and management of concussion in sports. Neurology 1997; 48:575–80.

Landy PJB. Neurogical sequelae of minor head and neck injuries. Injury 1998; 29:129–206.

Lanzi G, Balottin U, Borgatti R, De Agostini G, Pezzota S, Spanu G. Late post-traumatic headache in pediatric age. Cephalalgia 1985; 5:211–5.

Martelli MF, Grayson RL, Zasler ND. Posttraumatic headache: neuropsychological and psychological effects and treatment implications. J Head Trauma Rehabil 1999; 14:49–69.

Packard RC. Epidemiology and pathogenesis of posttraumatic headache. J Head Trauma Rehabil 1999; 14:9–21.

Packard RC. Treatment of chronic daily posttraumatic headache with divalproex sodium. Headache 2000; 40:736–9.

Packard RC, Ham LP. Posttraumatic headache: determining chronicity. Headache 1993; 33:133–4.

Packard RC, Ham LP. Pathogenesis of posttraumatic headache and migraine: a common headache pathway? Headache 1997; 37:142–52.

Packard RC, Weaver R, Ham LP. Cognitive symptoms in patients with posttraumatic headache. Headache 1993; 33:365–8.

Ramadan N, Keidel M. Chronic posttraumatic headache. In: The Headaches. Olesen J, Tfelt-Hansen P, Welch KMA (eds). Lippincott & Wilkins. Philadelphia 2000: pp 771–80.

Weiss HD, Stern BJ, Goldberg J. Postraumatic migraine: chronic migraine precipitated by minor head and neck trauma. Headache 1991; 31:451–6.

Yagamuchi M. Incidence of headache and severity of head injury. Headache 1992; 32:427–31.

Zasler ND. Posttraumatic headache: caveats and controversies. J Head Trauma Rehabil 1999; 14:1–8.

5.3, 5.4 Acute and chronic headache attributed to whiplash injury

Bono G, Antonaci F, Ghirmai S, D'Angelo F, Berger M, Nappi G. Whiplash injuries: clinical picture and diagnosis work-up. Clin Exp Rheumatol 2000; 18(S19):S23–S28.

Borchgrevink GE, Kaasa A, McDonagh D, Stiles TC, Haraldseth O, Lereim I. Acute treatment of whiplash neck sprain injuries. A randomized trial of treatment during the first 14 days after a car accident. Spine 1998; 23:25–31.

Cassidy J (ed). Scientific Monograph of the Quebec Task Force on Whiplash Associated Disorders: redefining 'whiplash' and its management. Spine 1995; 20:S8.

Cassidy JD, Carrol LJ, Cote P, Lemstra M, Berglund A, Nygren A. Effect of eliminating compensation for pain and suffering on the outcome of insurance claims for whiplash injury. N Engl J Med 2000; 342:1179–86.

Cote P, Cassidy JD, Carroll L. Is a lifetime history of neck injury in a traffic collision associated with prevalent neck pain, headache and depressive symptomatology? Accid Anal Prev 2000; 32:151–9.

Evans RW. Some observations on whiplash injuries. Neurol Clin 1992; 10:975–98.

Evans RW, Evans RI, Sharp MJ. The physician survey on the postconcussion and whiplash syndromes. Headache 1994; 34:268–74.

Hagstrom Y, Carlsson J. Prolonged functional impairments after whiplash injury. Scand J Rehabil Med 1996; 28:139–46.

Karlsborg M, Smed A, Jespersen H, Stephensen S, Cortsen M, Jennum P, Herning M, Korfitsen E, Werdelin L. A prospective study of 39 patients with whiplash injury. Acta Neurol Scand 1997; 95:65–72.

Keidel M, Rieschke P, Stude P, Eisentraut R, van Schayck R, Diener H. Antinociceptive reflex alteration in acute posttraumatic headache following whiplash injury. Pain 2001; 92:319–26.

Kolbinson DA, Epstein JB, Burgess JA. Temporomandibular disorders, headaches, and neck pain following motor vehicle accidents and the effect of litigation: review of the literature. J Orofac Pain 1996; 10:101–25.

Magnusson T. Extracervical symptoms after whiplash trauma. Cephalalgia 1994; 14:223–7.

Obelieniene D, Bovim G, Schrader H, Surkiene D, Mickeviciene D, Miseviciene I, Sand T. Headache after whiplash: a historical cohort study outside the medico-legal context. Cephalalgia 1998; 18:559–64.

Obelieniene D, Schrader H, Bovim G, Miseviciene I, Sand T. Pain after whiplash: a prospective controlled inception cohort study. J Neurol Neurosurg Psychiat 1999; 66:279–83.

Pettersson K, Hildingsson C, Toolanen G, Fagerlund M, Bjornebrink J. MRI and neurology in acute whiplash trauma. No correlation in prospective examination of 39 cases. Acta Orthop Scand 1994; 65:525–8.

Radanov BP, Sturzenegger M, Di Stefano G, Schnidrig A, Aljinovic M. Factors influencing recovery from headache after common whiplash. BMJ 1993; 307:652–5.

Radanov BP, Begre S, Sturzenegger M, Augustiny KF. Course of psychological variables in whiplash injury – a 2-year follow-up with age, gender and education pair-matched patients. Pain 1996; 64:429–34.

Schrader H, Obelieniene D, Bovim G, Surkiene D, Mickeviciene D, Miseviciene I, Sand T. Natural evolution of late whiplash syndrome outside the medicolegal context. Lancet 1996; 347:1207–11.

Sturzenegger M, Radanov BP, Di Stefano G. The effect of accident mechanisms and initial findings on the long-term course of whiplash injury. J Neurol 1995; 242:443–9.

Wallis BJ, Lord SM, Barnsley L, Bogduk N. The psychological profiles of patients with whiplash-associated headache. Cephalalgia 1998; 18:101–5.

5.7 Post-craniotomy headache

Feghali JG, Elowitz EH. Split calvarial graft cranioplasty for the prevention of headache after retrosigmoid resection of acoustic neuromas. Laryngoscope. 1998; 108:1450–2.

Ferber J, Juniewicz H, Glogowska E, Wronski J, Abraszko R, Mierzwa J. Tramadol for postoperative analgesia in intracranial surgery. Its effect on ICP and CPP. Neurologia i Neurochirurgia Polska. 2000; 34(6 Suppl):70–9.

Fetterman BL, Lanman TH, House JW. Relief of headache by cranioplasty after skull base surgery. Skull Base Surg 1997; 7:1–4.

Hanson MB, Glasscock ME, Brandes JL, Jackson CG. Medical treatment of headache after suboccippital acoustic tunour removal. Laryngoscope 1998; 108:1111–4.

Hagell P. Postoperative pain control after craniotomy. J Neuroscience Nursing 1999; 31:47–9.

Harner SG, Beatty CW, Ebersold MJ. Headache after acoustic neuroma excision. Am J Otol 1993; 14:552–5.

Harner SG, Beatty CW, Ebersold MJ. Impact of cranioplasty on headache after acoustic neuroma removal. Neurosurgery 1995; 36:1097–9.

Jackler R, Pitts L. Acoustic neuroma. Neurosurg Clin North Am 1990; 1:199–223.

Kaur A, Selwa L, Fromes G, Ross DA. Persistent headache after supratentorial craniotomy. Neurosurgery 2000; 47:633–6.

Koperer H, Deinsberger W, Jodicke A, Boker DK. Postoperative headache after suboccipital approach: craniotomy versus craniectomy. Minimally Invasive Neurosurgery 1999; 42:175–8.

Leith BA. Pharmacological management of pain after intracranial surgery. J Neurosci Nursing 1998; 30:220–4.

Leith BA. Pain assessment and management. Axone 1999; 21:4–9.

Lovely TJ, Lowry DW, Jannetta PJ. Functional outcome and the effect of cranioplasty after retromastoid craniectomy for microvascular decompression. Surg-Neurol 1999; 51:191–7.

Mosek AC, Dodick DW, Ebersold MJ, Swanson JW. Headache after resection of acoustic neuroma. Headache 1999; 39:89–94.

Nguyen A, Girard F, Boudreault D, Fugere F, Ruel M, Moumdjian R, Bouthilier A, Caron JL, Bojanowski MW and Girard DC. Scalp nerve blocks decrease the severity of pain after craniotomy. Anesthesia & Analgesia 2001; 93:1272–6.

Parving A, Mirko T, Thomsen J, Moller H, Buchwald C. Some aspect of life after surgery for acoustic neuroma. Arch Otolaryngol Head Neck Surg 1992; 118:1061–4.

Pedrosa CA, Ahern DK, McKenna MJ, Ojemann RG, Aquadro MA. Determinants and impact of headache after acoustic neuroma surgery. Am J Otol. 1994; 15:793–7.

Schessel DA, Nedzelski JM, Rowed D, Feghali JG. Pain after surgery for acoustic neuroma. Otolaryngol Head Neck surgery 1992: 107;424–9.

Schessel DA, Rowed D, Nedzelski JM, Feghali JG. Postoperative pain following excision of acoustic neuroma by the suboccipital approach: observations of possible cause and potential amelioration. Am J Otol 1993; 14:491–4.

Soumekh B, Levine SC, Haines SJ, Wulf JA. Retrospective study of postcraniotomy headaches in suboccipital approach: diagnosis and management. Am J Otol 1996; 17:617–9.

Tanskanen P, Kytta J, Randell T. Patient-controlled analgesia with oxycodone in the treatment of postcraniotomy pain. Acta Anaesthesiol Scand 1999; 43:42–5.

Vijayan N. Postoperative headache in acoustic neuroma. Headache 1995; 35:98–100.

Wazen JJ, Sisti M and Lam SM. Cranioplasty in acoustic neuroma surgery. Laryngoscope 2000; 110:1294–7.

Weigand DA, Ojemann RG, Fickel V. Surgical treatment of acoustic neuroma (vestibular schwannoma) in the United States: Report of the acoustic neuroma registry. Laryngoscope 1996; 106:58–66.

Wiegand DA, Fickel V. Acoustic neuroma – the patient's perspective: subjective assessment of symptoms, diagnosis, therapy, and outcome in 541 patients. Laryngoscope 1989; 99:179–87.

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
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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 Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL)
- 6.7.2 Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes (MELAS)
- 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[1] 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 relation 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 common 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 dysrythmia. 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 leptomeningeal angiomatosis may be a cause of symptomatic migraine, particularly of attacks with prolonged auras (possibly related to chronic oligaemia).

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 1 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 subform 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.

Bibliography

6.1.1 Headache attributed to ischaemic stroke

Ferro JM, Melo TP, Oliveira V, Salgado AV, Crespo M, Canhao P, Pinto AN. A multivariate study of headache associated with ischemic stroke. Headache 1995; 35:315–9.

Fisher CM. Headache in acute cerebrovascular disease. In: Vinken PH, Bruyn GW eds. Headache and cranial neuralgias. Handbook of clinical neurology. Amsterdam; Elsevier 1968; 5:124–56.

Gorelick PB, Hier DB, Caplan LR, Langenberg D. Headache in acute cerebrovascular disease. Neurology 1986; 36: 1445–50.

Portenoy RK, Abissi CJ, Lipton RB, Berger AR, Mebler MF, Baglivo J, Solomon S. Headache in cerebrovascular disease. Stroke 1984; 15:1009–12.

6.1.2 Headache attributed to transient ischaemic attack

Caplan LR. Migraine and vertebrobasilar ischemia. Neurology 1991; 41:55–61.

Ferro JM, Costa I, Melo TP, Canhao P, Oliveira V, Salgado AV, Crespo M, Pinto AN. Headache associated with transient ischemic attacks. Headache 1995; 35:544–8.

Fisher CM. Migraine accompaniments versus arteriosclerotic ischemia. Trans Am Neurol Assoc 1968; 93:211–3.

Fisher CM. Cerebral ischemia: less familiar types. Clin Neurosurg 1971; 18:267–336.

Fisher CM. Late-life migraine accompaniements as a cause of unexplained transient ischemic attacks. Can J Med Sci 1980; 7:9–17.

Martsen BH, Sorensen PS, Marquardsen J. Transient ischemic attacks in young patients: a thromboembolic or migrainous manifestation? A ten-year follow-up of 46 patients. J Neurol Neurosurg Psychiat 1990; 53:1029–33.

6.2.1 Headache attributed to intracerebral haemorrhage

Ferro JM, Melo TP, Guerreiro M. Headaches in intracerebral hemorrhage survivors. Neurology 1998; 50:203–7.

Gorelick PB, Hier DB, Caplan LR, Langenberg P. Headache in acute cerebrovascular disease. Neurology 1986; 36:1445–0.

Jensen TS, Gorrelick PB. Headache associated with stroke and intracranial hematoma. In: The Headaches. 2nd edition. J Olesen, P Telt-Hansen, KMA Welch eds. Philadelphia: Lippincott Williams and Wilkins 2000, pp 781–7.

Melo TP, Pinto AN, Ferro JM. Headache in intracerebral hematomas. Neurology 1996; 47:494–500.

Schuaib A, Metz L, and Hing T. Migraine and intra-cerebral hemorrhage. Cephalalgia 1989; 9:59–61.

6.2.2 Headache attributed to subarachnoid haemorrhage

Bassi P, Bandera R Loiero M, Togoni G, Mangoni. Warning signs in subarachnoid hemorrhage: a cooperative tudy. Acta Neurol Scand 1991; 84:277–81.

Edlow JA, Caplan LR. Avoiding pitfalls in the diagnosis of subarachnoid hemorrhage. NEJM 2000; 342:29–36.

Evans RW. Diagnostic testing for the evaluation of headaches. Neurology Clinics 1996; 14:1–26.

Linn FHH, Rinkel GJE, Algra A, van Gijn J. Headache characteristics in subarachnoid haemorrhage and benign thunderclap headache. J Neurol Neurosurg Psych 1998; 65: 791–3.

Mayberg MR, Batjer HH, Dacey R, Diringer M, Haley EC, Heros RC, Sternau LL, Torner J, Adams HP, Feinberg W, Thies W. Guidelines for the management of aneurysmal subarachnoid hemorrhage. A statement for healthcare professionals from a special writing group of the Stroke Council of the American Heart Association. Stroke 1994; 25:2315–28.

Ramirez-Lassepas M, Espinosa CE, Cicero JJ, Johnston KL, Cipolle RJ, Barber DL. Predictors of intracranial pathologic findings in patients who seek emergency care because of headache. Arch Neurol 1997; 54:1506–9.

Seymour JJ, Moscati RM, Jehle DV. Response of headaches to non-narcotic analgesics resulting in missed intracranial hemorrhage. Am J Emerg Med 1995; 13:43–5.

Sidman R, Vconnolly E, Lemke T. Subarachnoid hemorrhage diagnosis: lumbar puncture is still needed when the computed tomography scan is normal. Acad Emerg Med 1996; 3:827–31.

Van der Wee N, Rinkel GJE, Hasan D, van Gijn J. Detection of subarachnoid hemorrhage on early CT: is lumbar puncture still needed after a negative scan? J Neurol Neurosurg Psychiat 1995; 58:357–9.

Verweij RD, Wijdicks EFM, van Gijn J. Warning headache in aneurysmal subarachnoid hemorrhage. A case control study. Arch Neurol 1988; 45:1019–20.

Weir B. Diagnostic aspects of SAH. In: Weir B. Subarachnoid hemorrhage: causes and cures. New York: Oxford University Press 1998, pp 144–76.

6.3.1 Headache attributed to saccular aneurysm

Day JW, Raskin NH. Thunderclap headache: symptom of unruptured cerebral aneurysm. Lancet 1986; 2:1247–8.

Linn FHH, Wijdicks EFM, van der Graaf Y, Weerdesteyn-van Vliet FAC, Bartelds A, van Gijn J. Prospective study of sentinel headache in aneurysmal subarachnoid haemorrhage. Lancet 1994; 344:590–3.

Markus HS. A prospective follow-up of thunderclap headache mimicking subarachnoid haemorrhage. J Neurol Neurosurg Psych 1991; 54:1117–25.

Mas JL, Baron JC, Bousser MG, Chiras J. Stroke, migraine and inracranial aneurysm: a case report. Stroke 1986; 17:1019–21.

Ostergard JR, Ramadan N. Unruptured vascular malformations and subarachnoid hemorrhage. In The Headaches 2nd edition. J Olesen, P Tfelt-Hansen, KMA Welch eds. Philadelphia: Lipincott Williams and Wilkins 2000: pp 789–96.

Raps EC, Rogers JD, Galetta DL, Solomon RA, Lennihan L, Klebanoff LM, Fink ME. The clinical spectrum of unruptured intracranial aneurysms. Arch Neurol 1993; 50:265–8.

Schievink WI. Intracranial aneurysms. NEJM 1997; 336:28–40.

Wijdicks EFM, Kerkhoff H, van Gijn J. Long-term follow-up of 71 patients with thunderclap headache mimicking subarachnoid haemorrhage. Lancet 1988; ii:68–70.

6.3.2 Headache attributed to arteriovenous malformation

Bruyn GW. Intracranial arteriovenous malformation and migraine. Cephalalgia 1984; 4:191–207.

Haas DC. Arteriovenous malformations and migraine: case reports and an analysis of the relationship. Headache 1991; 31:509–13.

Troost BT, Mark LE, Maroon JC. Resolution of classic migraine after removal of an occipital lobe AVM. Ann Neurol 1979; 5:199–201.

6.3.3 Headache attributed to dural arterio-venous fistula

Malek AM, Halbach VV, Dowd CF, Higashida RT. Diagnosis and treatment of dural arteriovenous fistulas. Neuroimaging Clin N Am 1998; 8:445–68.

6.3.4 Headache attributed to cavernous angioma

De Benedittis G. SUNCT syndrome associated with cavernous angioma of the brain stem. Cephalalgia 1996; 16:503–6.

Epstein MA, Beerman PH, Schut L. Cavernous angioma presenting as atypical facial and head pain. J Child Neurol 1990; 5:27–30.

Robinson JR, Awad IA, Little JR. Natural history of the cavernous angioma. J Neurosurg 1991; 75:709–14.

6.3.5 Headache attributed to encephalotrigeminal angiomatosis (Sturge Weber syndrome)

Chabriat H, Pappata S, Traykov L, Kurtz A, Bousser MG. Angiomatose de Sturge Weber responsable d'une hémiplégie sans infarctus cérébral en fin de grossesse. Rev Neurol (Paris) 1996; 152:536–41.

Klapper J. Headache in Sturge-Weber syndrome. Headache 1994; 34:521–2.

6.4.1 Headache attributed to giant cell arteritis

Caselli RJ, Hunder GG. Neurologic aspects of giant cell (temporal) arteritis. Rheum Dis Clin North Am 1993; 19:941–53.

Hunder GG. Giant cell (temporal) arteritis. Rheum Dis Clin North Am 1990; 16:399–409.

Lee AG, Brazis PW. Temporal arteritis: a clinical approach. J Am Geriatr Soc 1999; 47:1364–70.

Solomon S, Cappa KG. The headache of temporal arteritis. J Am Geriatr Soc 1987; 35:163–5.

Swannell AJ. Polymyalgia rheumatica and temporal arteritis: diagnosis and management. BMJ 1997; 314:1329–32.

Thielen KR, Wydicks EFM, Nichols DA. Giant cell (temporal) arteritis: involvement of the vertebral and internal carotid arteries. Mayo Clin Proc 1998; 73:444–6.

6.4.2 Headache attributed to Primary CNS angiitis and 6.4.3 Headache attributed to Secondary CNS angiitis

Calabrese LH, Furlan AH, Gragg LA, Ropos TH. Primary angiitis of the central nervous system: diagnostic criteria and clinical approach. Cleve J Med 1992: 59:293–306.

Calabrese LH, Duna GF, Lie JT. Vasculitis in the central nervous system; Arthritis Rheum 1997; 40:1189–201.

Chu CT, Gray L, Goldstein LB, Hulette CM. Diagnosis of intracranial vasculitis: a multidisciplinary approach. J Neuropath Exp Neurol 1998; 57:30–8.

Hankey GJ. Necrotizing and granulomatous angiitis of the CNS. In Cerebrovascular disease. Pathophysiology, diagnosis and management, vol 2. MD Ginsberg, J Bogousslavsky eds. New York: Blackwell Science 1998: 1647–83.

Harris KG, Tran DD, Sickels WJ, Cornell SH, Yuh WTC. Diagnosing intracranial vasculitis: the roles or MR and angiography. Am J Neuroradiol 1994; 15:317–30.

Kumar R, Wijdicks EFM, Brown RD, Parisi JE, Hammond CA. Isolated angiitis of the CNS presenting as subarachnoid haemorrhage. J Neurol Neurosurg Psych 1997; 62:649–51.

Lie JT. Primary (granulomatous) angiitis of the central nervous system: a clinicopathologic analysis of 15 new cases and a review of the literature. Hum Pathol 1992; 23:164–71.

Moore PM. Vasculitis of the central nervous system. Semin Neurol 1994; 14: 313–9.

Savage COS, Harper L, Cockwell P, Adu D, Howie AJ. ABC of arterial and vascular disease: vasculitis. BMJ 2000; 320:1325–8.

6.5.1 Arterial dissection

Biousse V, D'Anglejan-Chatillon J, Massiou H, Bousser MG. Head pain in non traumatic artery dissection: a series of 65 patients. Cephalalgia 1994; 14:33–6.

Biousse V, D'Anglejan-Chatillon J, Touboul PJ, Amarenco P, Bousser MG. Time course of symptoms in extracranial carotid artery dissections. A series of 80 patients. Stroke 1995; 26:235–9.

Biousse V, Woimant F, Amarenco P, Touboul PJ, Bousser MG. Pain as the only manifestation of extracranial internal carotid artery dissection. Cephalalgia 1992; 12:314–7.

D'Anglejan Chatillon J, Ribeiro V, Mas JL, Youl BD, Bousser MG. Migraine – a risk factor for dissection of cervical arteries. Headache 1989; 29:560–1.

De la Sayette V, Leproux F, Letellier Ph. Cervical cord and dorsal medullary infarction presenting with retro-orbital pain. Neurology 1999; 53:632–4.

Duyff RF, Snidjers CJ, Vanneste JAL. Spontaneous bilateral internal carotid artery dissection and migraine: a potential diagnostic delay. Headache 1997; 37:109–12.

Fisher CM. The headache and pain of spontaneous carotid dissection. Headache 1982; 22:60–5.

Guillon B, Biousse V, Massiou H, Bousser MG. Orbital pain as an isolated sign of internal carotid artery dissection. A diagnostic pitfall. Cephalagia 1998, 18:222–4.

Guillon B, Lévy C, Bousser MG. Internal carotid artery dissection: an update. J Neurol Sci 1998; 153:146–58.

Nakatomi H, Nagata K, Kawamoto S, Shiokawa Y. Ruptured dissecting aneurysm as a cause of subarachnoid hemorrhage of unverified etiology. Stroke 1997; 28:1278–82.

Ramadan NM, Tietjen GE, Levine SR, Welch KMA. Scintillating scotomata associated with internal carotid artery dissection: report of three cases. Neurology 1991; 41:1084–7.

Silbert PL, Mokri B, Schievink WI. Headache and neck pain in spontaneous internal carotid and verterbral artery dissections. Neurology 1995; 45:1517–22.

Sturzenegger M. Headache and neck pain. The warning symptoms of vertebral artery dissection. Headache 1994; 34:187–93.

6.5.2 Post-endarterectomy headache

Breen JC, Caplan LR, DeWitt LD, Belkin M, Mackey WC, Donnell TP. Brain edema after carotid surgery. Neurology 1996; 46:175–81.

De Marinis M, Zaccaria A, Faraglia V, Fiorani P, Maira G, Agnoli A. Post endarterectomy headache and the role of the oculo-sympathetic system. J Neurol Neurosurg Psych 1991; 54:314–7.

Ille O, Woimant F, Pruna A, Corabianu O, Idatte JM, Haguenau M. Hypertensive encephalopathy after bilateral carotid endarterectomy. Stroke 1995; 26:488–91.

Leviton A, Caplan L, Salzman E. Severe headache after carotid endarterectomy. Headache 1975; 15:207–9.
Tehindrazanarivelo A, Lutz G, Petitjean C, Bousser MG. Headache following carotid endarterectomy: a prospective study. Cephalalgia 1991; 11(suppl 11):353.

6.5.3 Carotid angioplasty headache

Dietrich EB, Ndiaye M, Reid DB. Stenting in the carotid artery. Experience in 110 patients. J Endovasc Surg 1996; 3:42–62.
Gil-Peralta A, Mayol A, Gonzalez Marcos JR, Gonzalez A, Ruano J, Boza F, Duran F. Percutaneous transluminal angioplasty of the symptomatic atherosclerotic carotid arteries; Results, complications and follow-up. Stroke 1996; 27: 2271–3.
McCabe DJH, Brown MM, Clifton A. Fatal cerebral reperfusion hemorrhage after carotid stenting. Stroke 1999; 30:2483–6.
Munari LM, Belloni G, Moschini L, Mauro A, Pezzuoli G, Porta M. Carotid pain during percutaneous angioplasty. Pathophysiology and clinical features. Cephalalgia 1994; 14:127–31.
Schoser BG, Heesen C, Eckert B, Thie A. Cerebral hyperperfusion injury after percutaneous transluminal angioplasty of extracranial arteries. J Neurol 1997; 244:101–4.

6.5.4 Headache attributed to intracranial endovascular procedures

Martins IP, Baeta E, Paiva T, Campos J, Gomes L. Headaches during intracranial endovascular procedures: a possible model for vascular headache. Headache 1993; 23:227–33.
Nichols FT, Mawad M, Mohr JP, Hilal S, Adams RJ. Focal headache during balloon inflation in the vertebral and basilar arteries. Headache 1993; 33:87–9.
Nichols FT, Mawad M, Mohr JP, Stein B, Hilal S, Michelsen WJ. Focal headache during balloon inflation in the internal carotid and middle cerebral arteries. Stroke 1990; 21:555–9.

6.5.5 Angiography headache

Shuaib A, Hachinski VC. Migraine and the risks from angiogaphy. Arch Neurol 1988; 45:911–2.

6.6 Headache attributed to cerebral venous thrombosis

Aidi S, Chaunu MP, Biousse V, Bousser MG. Changing pattern of headache pointing to cerebral venous thrombosis after lumbar puncture and intra venous high dose corticosteroids. Headache 1999; 39:559–64.
Ameri A, Bousser MG. Cerebral venous thrombosis. Neurologic Clinics 1992; 10:87–111.
Biousse V, Ameri A, Bousser MG. Isolated intracranial hypertension as the only sign of cerebral venous thrombosis. Neurology 1999; 53:1537–42.
Bousser MG, Ross Russell R. Cerebral venous thrombosis. Major problems in neurology, vol 1. London: Saunders 1997.
Crassard I, Biousse V, Bousser MG, Meyer B, Marsot-Dupuch K. Hearing loss and headache revealing lateral sinus thrombosis in a patient with Factor V Leiden mutation. Stroke 1997; 28:876–7.
De Bruijn SFTM, Stam J, Kappelle LJ for CVST study group. Thunderclap headache as first symptom of cerebral venous sinus thrombosis. Lancet 1996; 348:1623–5.
Leker RR, Steiner I. Features of dural sinus thrombosis simulating pseudotumor cerebri. Eur J Neurol 1999; 6:601–4.
Newman DS, Levine SR, Curtis VL, Welch KMA: Migraine like visual phenomena associated with cerebral venous thrombosis. Headache 1989; 29:82–5.
Tehindrazanarivelo AD, Evrard S, Schaison M, Mas JL, Dormont D, Bousser MG: Prospective study of cereral sinus venous thrombosis in patients presenting with benign intracranial hypertension. Cerebrovasc Dis 1992; 2:22–7.

6.7.1 CADASIL

Chabriat H, Tournier-Lasserve E, Vahedi K, Leys D, Joutel A, Nibbio A, Escailla JP, Iba-Zizen MT, Bracard S, Tehindrazanarivelo A, Gastaut JL, Bousser MG. Autosomal dominant migraine with MRI white matter abnormalities mapping to the CADASIL locus. Neurology 1995; 45:1086–91.
Chabriat H, Vahedi K, Iba-Zizen MT, Joutel A, Nibbio A, Nagy TG, Krebs MO, Julien J, Dubois B, Ducrocq X, Levasseur M, Homeyer P, Mas JL, Lyon-Caen O, Tournier-Lasserve E, Bousser MG. Clinical spectrum of CADASIL: a study of 7 families. Lancet 1995; 346:934–9.
Joutel A, Corpechot C, Ducros A, Katayoun V, Chabriat H, Mouton P, Alamowitch S, Domenga V, Cécillion M, Maréchal E, Maciazek J, Vayssière C, Cruaud C, Cabanis EA, Ruchoux MM, Weissenbach J, Bacxh JF, Bousser MG, Tournier-Lasserve E. 'Notch 3' mutations in CADASIL, a hereditary adult-onset condition causing stroke and dementia. Nature 1996; 383:707–10.
Verin M, Rolland Y, Landgraf F Chabriat H, Bompais B, Michel A, Vahedi K, Martinet JP, Tournier-Lasserve E, Lemaitre MH, Edan G. New phenotype of the cerebral autosomal dominant arteriopathy mapped to chromosome 19 migraine as the prominent clinical feature. J Neurol Neurosurg Psychiat 1995; 59:579–85.

6.7.2 MELAS

Klopstock A, May P, Siebel E, Papagiannuli E, Diener NC, Heichmann H. Mitochondrial DNA in migraine with aura. Neurology 1996; 46:1735–8.
Koo B, Becker L, Chuang S, Merante F, Robinson BH, Mac Gregor D, Tein I, Ho VB, McGreal DA, Wherrett JR, Logan WJ. Mitochondrial encephalomyopathy, lactic acidosis, stroke-like episodes (MELAS): clinical, radiological, pathological and genetic observations. Ann Neurol 1993; 34:25–32.
Ojaimi J, Katsabanis S, Bower S, Quigley A, Byrne E. Mitochondrial DNA in stroke and migraine with aura. Cerebrovasc Dis 1998; 8:102–6.
Pavlakis SG, Phillips PC, Di Mauro S, De Vivo DC, Rowland P. Mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes: a distinct clinical syndrome. Ann Neurol 1984; 16:481–8.

6.7.3 Headache attributed to benign angiopathy of the CNS

Call GK, Fleming MC, Sealfon S, Levine H, Kistler JP, Fisher CM. Reversible cerebral segmental vasoconstriction. Stroke 1988; 19:1159–70.

Dodick DW, Brown RD, Britton JW, Huston J. Non aneurysmal thunderclap headache with diffuse, multifocal segmental and reversible vasospasm. Cephalalgia 1999; 19:118–213.

Lee KY, Sohn YH, Kim SH, Sunwoo IN. Basilar artery vasospasm in postpartum cerebral angiopathy. Neurology 2000; 54:2003–5.

McColl GJ, Fraser K. Pheochromocytoma and pseudovasculitis. J Rheumatol 1995; 22:1441–2.

Razavi M, Bendixen B, Maley JE, Schoaib M, Zargarian M, Razavi B, Adams HP. CNS pseudovasculitis in a patient with pheochromocytoma. Neurology 1999; 52:1088–90.

Serdaru M, Chiras J, Cujas M, Lhermitte F. Isolated benign cerebral vasculitis or migrainous vasospasm? J Neurol Neurosurg Psychiat 1984; 47:73–6.

6.7.4 Headache attributed to pituitary apoplexy

Carral F. Pituitary apoplexy. Arch Neurol 2001; 58:1143–4.

Chakeres DW, Curtin A, Ford G. Magnetic resonance imaging of pituitary and parasellar abnormalities. Radiol Clin North Am 1989; 27:265–81.

Da Motta LA, de Mello PA, de Lacerda CM, Neto AP, Da Motta LD, Filho MF. Pituitary apoplexy. Clinical course, endocrine evaluations and treatment analysis. J Neurosurg Sci 1991; 43:25–36.

Dodick DW, Wijdicks EFM. Pituitary apoplexy presenting as thunderclap headache. Neurology 1998; 50:1510–1.

Hernandez A, Angeles Del Real M, Aguirre M, Vaamonde J, Gudin M, Ibanez R. Pituitary apoplexy: a transient benign presentation mimicking with subarachnoid hemorrhage with negative angiography. Eur J Neurol 1998; 5:499–501.

Lee CC, Cho AS, Carter WA. Emergency department presentation of pituitary apoplexy. Am J Emerg Med 2000; 18: 328–31.

McFadzean RM, Doyle D, Rampling R, Teasdale E, Teasdale G. Pituitary apoplexy and its effect on vision. Neurosurgery 1991; 29:669–75.

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 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_2O in the non-obese, >250 mm H_2O 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_2O 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_2O in the non-obese, >250 mm H_2O 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

Comment:
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, sixth 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_2O in the non-obese or >250 mm H_2O 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[1]:
   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_2O 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, hypoliquorrhoeic 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_2O 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[1]
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[1]
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 hypophyseal 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[1]
B. Space-occupying intracranial tumour demonstrated by CT or MRI and causing hydrocephalus[2]
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 adrenocorticotropic 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[1]

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 *migralepsy* 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 'migralepsy' 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. ≥5 mm caudal descent of the cerebellar tonsils
   2. ≥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, disequilibrium, 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/μl) 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/μl), 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_20) 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

Bibliography and references

7.1 Headache attributed to high cerebrospinal fluid pressure

Corbett JJ, Mehta MP. Cerebrospinal fluid pressure in normal obese subjects and patients with pseudotumor cerebri. Neurology 1983; 33:1386–88.

Corbett JJ, Nerad JA, Tse DT, Anderson RL. Results of optic nerve sheath fenestration for pseudotumor cerebri. The lateral orbitotomy approach. Arch Ophthalmol 1988; 106:1391–7.

Corbett JJ, Thompson HS. The rational management of idiopathic intracranial hypertension. Arch Neurol 1989; 46:1049–51.

Digre KB, Corbett JJ. Pseudotumor cerebri in men. Arch Neurol 1988; 45:866–72.

Durcan FJ, Corbett JJ, Wall M. The incidence of pseudotumor cerebri: population studies in Iowa and Louisiana. Arch Neurol 1988; 45:875–7.

Eggenberger ER, Miller NR, Vitale S. Lumboperitoneal shunt for the treatment of pseudotumor cerebri. Neurology 1996; 46:1524–30.

Fishman RA. Cerebrospinal fluid in diseases of the nervous system, WB Saunders Company, Philadelphia. 1992.

Gamache FW, Patterson RH, Alksne JF. Headache associated with changes in intracranial pressure. In Wolff's headache and other head pain, (Dalessio DJ, ed.) Oxford University Press, New York. 1987; pp. 352–5.

Gardner K, Cox T, Digre K. Idiopathic intracranial hypertension associated with tetracycline use in fraternal twins: case report and review. Neurology 1995; 45:6–10.

Giuseffi V, Wall M, Siegal PZ, Rojas PB. Symptoms and disease associations in idiopathic intracranial hypertension (pseudotumor cerebri): A case control study. Neurology 1991; 41:239–44.

Griffin JP. A review of the literature on benign intracranial hypertension. Adverse Drug React Toxicol Rev 1992; 11:41–58.

Johnson LN, Krohel GB, Madsen RW, March GA. The role of weight loss and acetazolamide in the treatment of idiopathic intracranial hypertension (pseudotumor cerebri). Ophthalmol 1998; 105:2313–7.

Johnston I, Paterson A. Benign intracranial hypertension II. CSF pressure and circulation. Brain 1974; 97:301–12.

Karahalios DG, Rekate HL, Khayata MH, Apostolides PJ. Elevated intracranial venous pressure as a universal mechanism in pseudotumor cerebri of varying etiologies. Neurology 1996; 46:198–202.

Kelman SE, Heaps R, Wolf A, Elman MJ. Optic nerve decompression surgery improves visual function in patients with pseudotumor cerebri. Neurosurg 1992; 30:391–5.

Keltner J. Optic nerve sheath decompression: how does it work? Has its time come? Arch Ophthalmol 1988; 106:1378–83.

Konomi H, Imai M, Nihei K, Kamoshita S, Tada H. Indomethacin causing pseudotumor cerebri in Bartter's syndrome. N Eng J Med 1978; 298:855.

Kupersmith MJ, Gamell L, Turbin R, Peck V, Spiegel P, Wall M. Effects of weight loss on the course of idiopathic intracranial hypertension in women. Neurology 1998; 50:1094–8.

Marcelis J, Silberstein SD. Idiopathic intracranial hypertension without papilledema. Arch Neurol 1991; 48:392–9.

Mathew NT, Ravinshankar K, Sanin LC. Coexistence of migraine and idiopathic intracranial hypertension without papilledema. Neurology 1996; 46:1226–30.

Nicolas J, Ramadan NM. Idiopathic intracranial hypertension. In: Neurobase. La Jolla: Arbor Pubs 1998.

Radhkrishnan K, Ahlskog JE, Cross SA, Kurland LT, O'Fallon WM. Idiopathic intracranial hypertension (pseudotumor cerebri). Descriptive epidemiology in Rochester, Minnesota, 1976 to 1990. Arch Neurol 1993; 50:78–80.

Ramadan NM. Headache related to increased intracranial pressure and intracranial hypotension. Curr Opinion Neurol 1996; 9:214–8.

Sergott RC, Savino PJ, Bosley TM. Modified optic nerve sheath decompression provides long-term visual improvement for pseudotumor cerebri. Arch Ophthalmol 1988; 106:1384–90.

Spector RH, Carlisle J. Pseudotumor cerebri caused by a synthetic vitamin A preparation. Neurology 1984; 34:1509–11.

Spence JD, Amacher AL, Willis NR. Benign intracranial hypertension without papilledema: role of 24 hour cerebrospinal fluid pressure monitoring in diagnosis and management. Neurosurgery 1980; 7:326–36.

Sugerman HJ, Felton WL, Sismanis A, Kellum JM, DeMaria EJ, and Sugerman EL. Gastric surgery for pseudotumor cerebri associated with severe obesity. Ann Surg 1999; 229:634–40.

Wall M. The headache profile of idiopathic intracranial hypertension. Cephalalgia 1990; 10:331–5.

Wall M. Idiopathic intracranial hypertension. Neurol Clin 1991; 9:73–95.
Wall M, George D. Idiopathic intracranial hypertension. Brain 1991; 114:155–80.
Wall M, George D. Idiopathic intracranial hypertension: a prospective study of 50 patients. Brain 1991; 114:155–80.
Wall M, White WN. Asymmetric papilledema in idiopathic intracranial hypertension: prospective interocular comparison of sensory visual function. Inv Ophthalmol Vis Sci 1998; 39:134–42.
Wang SJ, Silberstein SD, Patterson S, Young WB. Idiopathic intracranial hypertension without papilledema: A case-control study in a headache center. Neurology 1998; 51:245–9.
Weisberg LA. Benign intracranial hypertension. Medicine 1975; 54:197–207.

7.2.1 Post-dural (post-lumbar) puncture headache

Bruera OC, Bonamico L, Giglio JA, Sinay V, Leston JA, Figuerola M de L. Intracranial hypotension: the non specific nature of MRI findings. Headache 2000; 40:848–52.
Camann WR, Murray S, Mushlin PS, Lambert DH. Effects of oral caffeine on postural puncture headache. A double-blind, placebo-controlled trial. Anesth Analg 1990; 70:181–4.
Christoforidis GA, Mehta BA, Landi JL, Czarnecki EJ, Piaskowski RA. Spontaneous intracranial hypotension: report of four cases and review of the literature. Neuroradiology 1998; 40:636–43.
Chung SJ, Kim JS, Lee M. Syndrome of cerebral fluid hypovolemia. Clinical and imaging features and outcome. Neurology 2000; 55:1321–7.
Connelly NR, Parker RK, Rahimi A, Gibson CS. Sumatriptan in patients in postdural puncture headache. Headache 2000; 40:316–9.
Dieterich M, Brandt T. Is obligatory bed rest after lumbar puncture obsolete. Europ Arch Psychiatr Neurol Sci 1985; 235:71–5.
Evans RW, Armon C, Frohman EM, Goodin GS. Assessment: prevention of post-lumbar puncture headaches. Report of the Therapeutics and Technology Assessment Subcommitee of the American Academy of Neurology. Neurology 2000; 55:909–14.
Fischman RA, Dillon WP. Dural enhancement and cerebral displacement secondary to intracranial hypotension. Neurology 1993; 43:609–11.
Flaatten H, Kräkenes J, Vedeler C. Post-dural puncture related complications after diagnostic lumbar puncture, myelography and spinal anaesthesia. Acta Neurol Scand 1998; 98: 445–51.
Hochman S, Naidich TP, Kobetz SA, Fernandez-Maitin A. Spontaneous intracranial hypotension with pachymeningeal enhancement on MRI. Neurology 1992; 42: 1628–30.
Kovanen J, Sulkava R. Duration of postural headache after lumbar puncture: effect of needle size. Headache 1986; 26: 224–6.
Levine DN, Rapalino O. The pathophysiology of lumbar puncture headache. J Neurol Sci 2001; 192:1–8.
Mokri B, Atkinson JLD, Piepgras DG. Absent headache despite CSF volume depletion (intraranial hypotension). Neurology 2000; 55:1722–4.
Mokri B, Maher CO, Sencavoka D. Spontaneous CSF leaks: underlying disorder of connective tissue. Neurology 2002; 58:814–6.
Mokri B, Parisi JE, Scheithauer BW, Piepgras DG, Miller GM. Meningeal biopsy in intracranial hypotension: Menigeal enhancement on MRI. Neurology 1995; 45:1801–7.
Mokri B, Posner JB. Spontaneous intracranial hypotension. The broadening spectrum of CSF leaks. Neurology 2000; 55:1771–2.
Mokri B. Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia – evolution of a concept. Mayo Clin Proc 1999; 74:1113–23.
Morrow JI. Failure of oblique needle insertion to prevent post-lumbar puncture headache. Arch Neurol 1987; 44:795–5.
O'Carroll CP, Brant-Zawadzki M The syndrome of spontaneous intracranial hypotension. Cephalalgia 1999; 19:80–7.
Olsen KS. Epidural blood patch in the treatment of post-lumbar puncture headache. Pain 1987; 30:293–301.
Rabin B, Roychowdhury S, Meyer JR, Cohen BA, LaPat KD, Russel EJ. Spontaneous intracranial hypotension: spinal MRI findings. AJNR Am J Neuroradiol 1998; 19:1034–9.
Rando TA, Fishman RA. Spontaneous intracranial hypotension: report of two cases and review of the literature. Neurology 1992; 42:481–7.
Raskin NH. Lumbar puncture headache: a review. Headache 1990; 30:197–200.
Schievink WI, Morreale VM, Atkinson JLD, Meyer FB, Piepgras DG, Ebersold MJ. Surgical treatment of spontaneous spinal cerebrospinal fluid leaks. J Neurosurg 1998; 88:243–6.
Schwalbe SS, Schiffmiller MW, Marx GF. Theophylline for post-dural puncture headache. Anaesthesiology 1991; 75:A1081.
Sencavoka D, Mokri B, McClelland RL. The efficacy of epidural blood patch in spontaneous CSF leaks. Neurology 2001; 57:1921–3.
Strupp M, Brandt T, Müller A. Incidence of post-lumbar puncture syndrome reduced by reinserting the stylet: a randomized prospective study of 600 patients. J Neurol 1998; 245:589–92.
Thomas SR, Jamieson DRS, Muir KW. Randomised controlled trial of atraumatic versus standard needles for diagnostic lumbar puncture. BMJ 2000; 321:986–90.
Vilming ST, Ellertsen B, Troland K, Schrader H, Monstad I. MMPI profiles in post-lumbar puncture headache. Acta Neurol Scand 1997; 95:184–8.
Vilming ST, Kloster R. Post-lumbar puncture headache: clinical features and suggestions for diagnostic criteria. Cephalalgia 1997; 17:778–84.
Vilming ST, Kloster R. The time course of post-lumbar puncture headache. Headache 1998; 18:97–100.

7.3.1 Headache attributed to neurosarcoidosis, 7.3.2 Headache attributed to aseptic (non-infectious) meningitis

Ainiala H, Loukkoa J, Peltola J, Korpela Migraine, Hietaharju A. The prevalence of neuropsychiatric syndromes in systemic lupus erythematosus. Neurology 2001; 57:496–500.
Al-Fahad SA, Al-Araji AH. Neuro-Behcet's disease in Iraq: a study of 40 patients. J Neurol Sci 1999; 170:105–11.
Bachmeyer C, de la Blanchardiere A, Lepercq J, Dhote R, Grateau G, Detilleux M, Tournaire M, Christoforov B.

Recurring episodes of meningitis (Mollaret's meningitis) with one showing an association with herpes simplex virus type 2. J Infect 1996; 32:247–8.
Bakheit AM, Kennedy PG, Graham DI, More JR. Idiopathic granulomatous meningitis. J Neurol Neurosurg Psychiat 1989; 52:1286–9.
Bartleson JD, Swanson JW, Whisnant JP. A migrainous syndrome with cerebrospinal fluid pleocytosis. Neurology 1981; 31:1257–62.
Berg MJ, Williams LS. The transient syndrome of headache with neurologic deficits and CSF lymphocytosis. Neurology 1995; 45:1648–54.
Brey RL, Gharavi AE, Lockshin MD. Neurologic complications of antiphospholipid antibodies. Rheum Dis Clin North Am 1993; 19:833–50.
Cooper SD, Brady MB, Williams JP, Pilgreen KL, Harp DL, Weissmann JR. Neurosarcoidosis: evaluation using computed tomography and magnetic resonance imaging. J Comput Tomogr 1988; 12:96–9.
Cohen BA, Rowley AH, Long CM. Herpes simplex type 2 in a patient with Mollaret's meningitis:meningitis:demonstration by polymerase chain reaction. Ann Neurol 1994; 35: 112–6.
Chapelon C, Ziza JM, Piette JC, Levy Y, Raguin G, Wechsler B *et al*. Neurosarcoidosis: signs, course and treatment in 35 confirmed cases. Medicine (Baltimore) 1990; 69:261–76.
Farah, Al-Shubaili A, Montaser A, Hussein JM, Malaviya AN, Mukhtar M, Al-Shayeb A, Khuraibet AJ, Khan R, Trontelj JV. Behcet's syndrome: a report of 41 patients with emphasis on neurological manifestations. J Neurol Neurosurg Psychiat 1998; 64:382–4.
Glanz BI, Venkatesan A, Schur PH, Lew RA, Khoshbin S. Prevalence of migraine in patients with systemic lupus erythematosus. Headache 2001; 41:285–9.
Gullapalli D, Phillips LH 2nd. Neurologic manifestations of sarcoidosis. Neurol Clin 2002; 20:59–83.
Hollinger P, Sturzenegger M. Mathis J, Schroth G, Hess CW. Acute disseminated encephalomyelitis in adults: a reappraisal of clinical CSF, EEG and MRI findings. J Neurol 2002; 249:320–9.
Jensenius M, Myrvang B, Storvold G, Bucher A, Hellum KB, Bruu Al. Herpes simplex virus type 2 DNA detected in cerebrospinal fluid of 9 patients with Mollaret's meningitis. Acta Neurol Scand 1998; 98:209–12.
Meng MV, St Lezin M. Trimethoprim-sulfamethoxazole induced recurrent aspetic meningitis. J Urol 2000; 164: 1664–5.
Moris G, Garcia-Monco JC. The challenge of drug-induced aseptic meningitis. Arch Intern Med 1999; 159:1185–94.
Nicolas J, Ramadan NM. Idiopathic intracranial hypertension. In: Neurobase. La Jolla: Arbor Pubs 1998.
Nowak DA, Widenka DC. Neurosarcoidosis: a review of its intracranial manifestation. J Neurol 2001; 248:363–72.
Omdal R, Waterloo K, Koldingsnes W, Husby G, Mellgren SI. Somatic and psychological features of headache in systemic lupus erythematosus. J Rheumatol 2001; 28:772–9.
Schwarz S, Mohr A, Knauth M, Wildemann B, Storch-Hagenlocher B. Acute disseminated encephalomyelitis; a follow-up study of 40 adult patients. Neurology 2001; 56:1313–8.
Seaton France AJ. Recurrent aseptic meningitis following nonsteroidal anti-inflammatory drugs – a reminder. Postgrad Med J 1999; 75:771–2.
Sharma OP. Neurosarcoidosis. Chest 1991; 100:301–2.
Sharma OP. Neurosarcoidosis: a personal perspective based on the study of 37 patient. Chest 1997; 112:220–8.
Stamboulis E, Spengos M, Rombos A, Haidemenos A. Aseptic inflammatory meningeal reaction manifesting as a migrainous syndrome. Headache 1987; 27:439–41.
Sylaja PN, Cherian PJ, Das CK, Raddhakrishnan W, Radhakrishnan K. Idiopathic hypertrophic cranial pachymeningitis. Neurol India 2002; 50:53–9.
Tselis A. Acute disseminated encephalomyelitis. Curr Treat Options Neurol 2001; 3:537–42.
Vinas FC, Rengachary S. Diagnosis and management of neurosarcoidosis. J Clin Neurosci 2001; 8:505–13.
Von Storch TJC, Merritt HH. The cerebrospinal fluid during and between attacks of migraine headaches. Am J Med Sci 1935; 190:226–31.
Walker A, Tyor W. Neurosarcoidosis. Curr Treat Options Neurol 2001; 3:529–35.

7.6 Headache attributed to epileptic seizure

Bernasconi A, Andermann F, Bernasconi N, Reutens DC, Dubeau F. Lateralizing value of peri-ictal headache: A study of 100 patients with partial epilepsy. Neurology 2000; 56:130–2.
Isler H, Wieser HG, Egli M. Hemicrania epileptica: synchronous ipsilateral ictal headache with migraine features. In: Andermann F, Lugaresi E, eds. Migraine and epilepsy. Boston, MA: Butterworth 1987; pp. 249–63.
Laplante P, Saint–Hilaire JM, Bouvier G. Headache as an epileptic manifestation. Neurology 1983; 33:1493–5.
Leniger T, Isbruch K, Von den Driesch S, Diener HC, Hufnagel A. Seizure-associated headache in epilepsy. Epilepsia 2001; 42:1176–9.
Lennox WG, Lennox MA. Epilepsy and related disorders. Boston: Little Brown 1960.
Marks DA, Ehrenberg BL. Migraine-related seizures in adults with epilepsy, with EEG correlation. Neurology 1993; 43:2476–83.
Panayiotopoulos CP, Ahmed Sharoqi I, Agathonikou A. Occipital seizures imitating migraine aura. J R Soc Med 1997; 90:255–7.
Schon F, Blau JN. Post-epileptic headache and migraine. J Neurol Neurosurg Psychiat 1987; 50:1148–52.
Sturzenegger MH, Meienberg O. Basilar artery migraine: a follow-up study of 82 cases. Headache 1985; 25:408–15.
Terzano MG, Parrino L, Pietrini V et al. Migraine-epilepsy syndrome: intercalated seizures in benign occipital epilepsy. In: Andermann F, Beaumanoir A, Mira L, eds. Occipital seizures and epilepsies in children. London: John Libbey 1993; pp. 93–9.
Young GB, Blume WT. Painful epileptic seizures. Brain 1983; 106:537–54.

7.7 Headache attributed to Chiari malformation type I (CM1)

Milhorat TH, Chou MW, Trinidad EM et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. Neurosurgery 1999; 44:1005–17.
Nohria V, Oakes WJ. Chiari I malformation: a review of 43 patients. Pediatr Neurosurg 1990–91; 16:222–7.

Nohria V, Oakes WJ. Chiari headaches. Neurology 1993; 43:1272.
Pascual J, Iglesias F, Oterino A et al. Cough, exertional, and sexual headaches: an analysis of 72 benign and symptomatic cases. Neurology 1996; 46:1520–4.
Pascual J, Oterino A, Berciano J. Headache in type I Chiari malformation. Neurology 1992; 42:1519–21.
Stovner LJ. Headache associated with the Chiari malformation. Headache 1993; 33:175–81.

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

Bartleson JD, Swanson JW, Whisnant JP. A migrainous syndrome with cerebrospinal fluid pleocytosis. Neurology 1981; 31:1257–62.
Berg MJ, Williams LS. The transient syndrome of headache with neurologic deficits and CSF lymphocytosis (review). Neurology 1995; 45:1648–54.
Gomez-Aranda F, Canadillas F, Marti-Masso JF, Diez-Tejedor E et al. Pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis: A report of fifty cases. Brain 1997; 120:1105–13.

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 induced by 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 medication-overuse headache
- 8.2.6 Headache attributed to other medication overuse
- 8.2.7 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 must occur if the diagnosis is to be definite. Prior to cessation, or pending improvement within 2 months after cessation, the diagnosis 8.2.7 *Probable medication-overuse headache* should be applied. If improve-

ment 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[1] and fulfilling criteria C and D

B. Absorption of a NO donor
C. Headache develops after NO is cleared from the blood[2]
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[1], 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[1]
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[1]
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[1] and fulfilling criteria C and D
B. Absorption of histamine
C. Headache develops after histamine is cleared from the blood[2]
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[1] and fulfilling criteria C and D
B. Absorption of CGRP
C. Headache develops after CGRP is cleared from the blood[2]
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 induced by 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

Introduction

This and the following section deal with headache disorders associated with chronic substance use or exposure.

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 ≥15 days per month and of a mixed picture of migraine-like and tension-type-like headaches on ≥15 days per month is overuse of symptomatic migraine drugs and/or analgesics. In general, overuse is defined in terms of treatment days per month. What is crucial is that treatment occurs both frequently and regularly, *ie*, on several days each week. For example, if the diagnostic criterion is use on ≥10 days per month, this translates into 2–3 treatment days every week. Bunching of treatment days with long periods without medication intake, practised by some patients, is much less likely to cause medication-overuse headache.

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*. Furthermore, the headache associated with medication overuse often has a peculiar pattern shifting, even within the same day, from having migraine-like characteristics to having those of tension-type headache (*ie*, a new type of 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 present on >15 days/month with at least one of the following characteristics and fulfilling criteria C and D:
   1. bilateral
   2. pressing/tightening quality
   3. mild or moderate intensity
B. Ergotamine intake on ≥10 days/month on a regular basis for ≥3 months
C. Headache has developed or markedly worsened during ergotamine overuse
D. Headache resolves or reverts to its previous pattern within 2 months after discontinuation of ergotamine

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

8.2.2 Triptan-overuse headache

Diagnostic criteria:
A. Headache present on >15 days/month with at least one of the following characteristics and fulfilling criteria C and D:
   1. predominantly unilateral
   2. pulsating quality
   3. moderate or severe intensity
   4. aggravated by or causing avoidance of routine physical activity (*eg*, walking or climbing stairs)
   5. associated with at least one of the following:
      a) nausea and/or vomiting
      b) photophobia and phonophobia
B. Triptan intake (any formulation) on ≥10 days/month on a regular basis for ≥3 months
C. Headache frequency has markedly increased during triptan overuse
D. Headache reverts to its previous pattern within 2 months after discontinuation of triptan

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 present on >15 days/month with at least one of the following characteristics and fulfilling criteria C and D:

1. bilateral
2. pressing/tightening (non-pulsating) quality
3. mild or moderate intensity

B. Intake of simple analgesics on ≥15 days/month[1] for >3 months
C. Headache has developed or markedly worsened during analgesic overuse
D. Headache resolves or reverts to its previous pattern within 2 months after discontinuation of analgesics

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

8.2.4 Opioid-overuse headache

Diagnostic criteria:
A. Headache present on >15 days/month fulfilling criteria C and D
B. Opioid intake on ≥10 days/month for >3 months
C. Headache has developed or markedly worsened during opioid overuse
D. Headache resolves or reverts to its previous pattern within 2 months after discontinuation of opioid

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

8.2.5 Combination medication-overuse headache

Diagnostic criteria:
A. Headache present on >15 days/month with at least one of the following characteristics and fulfilling criteria C and D:
   1. bilateral
   2. pressing/tightening (non-pulsating) quality
   3. mild or moderate intensity
B. Intake of combination medications[1] on ≥10 days/month for >3 months
C. Headache has developed or markedly worsened during combination medication overuse
D. Headache resolves or reverts to its previous pattern within 2 months after discontinuation of combination medication

Note:
1. Combination medications typically implicated are those containing simple analgesics combined with opioids, butalbital and/or caffeine

8.2.6 Headache attributed to other medication overuse

Diagnostic criteria:
A. Headache present on >15 days/month fulfilling criteria C and D
B. Regular overuse[1] for >3 months of a medication other than those described above
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

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

8.2.7 Probable medication-overuse headache

Diagnostic criteria:
A. Headache fulfilling criteria A–C for any one of the subforms 8.2.1 to 8.2.6 above
B. 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 8.2.7 *Probable medication-overuse headache* are 8.2.7.1 *Probable ergotamine-overuse headache*, 8.2.7.2 *Probable triptan-overuse headache*, 8.2.7.3 *Probable analgesic-overuse headache*, 8.2.7.4 *Probable opioid-overuse headache*, 8.2.7.5 *Probable combination medication-overuse headache* and 8.2.7.6 *Headache probably attributed to other medication overuse*.

Many patients fulfilling the criteria for 8.2.7 *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[1] for any therapeutic indication
C. Headache develops during medication
D. Headache resolves after discontinuation of medication[2]

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 pharmacological 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 ≥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 ≥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).

Bibliography

8.1 Headache induced by acute substance use or exposure

Altura BM, Altura BT, Gebrewold A. Alcohol induced spasm of cerebral blood vessels. J Mental Sci 2000; 104:972–99.

Armstrong PJ, Bersten A. Normeperidine toxicity. Anesth Analg 1986; 65:536–8.

Ashina M, Bendtsen L, Jensen R, Olesen J. Nitric oxide-induced headache in patients with chronic tension-type headache. Brain 2000; 123:1830–7.

Askew GL, Finelli L, Genese CA, Sorhage FE, Sosin DM, Spitalny KC. Bouillabaisse: an outbreak of methemoglobinemia in New Jersey in 1992. Pediatrics 1994; 94:381–4.

Askmark H, Lundberg PO, Olsson S. Drug related headache. Headache 1989; 29:441–4.

Atkins FM. A critical evaluation of clinical trials in adverse reactions to foods in adults. J Allergy Clin Immunol 1986; 78:174–82.

Beck HG, Schulze WH, Suter GM. Carbon monoxide – a domestic hazard. JAMA 1940; 115:1.

Bonnet GF, Nepveux P. Migraine due to tyramine. Sem Hop 1971; 47:2441–5.

Brewerton TD, Murphy DL, Lesem MD, Brandt HA, Jimerson DC. Headache responses following m-chlorophenylpiperazine in bulimics and controls. Headache 1992; 32:217–22.

Catalano G, Catalano MC, Rodriguez R. Dystonia associated with crack cocaine use. South Med J 1997; 90:1050–2.

Cleophas TJ, Niemeyer MG, vanderWall EE, vanderMeulen J. Nitrate-induced headache in patients with stable angina pectoris: beneficial effect of starting on a low dose. Angiology 1996; 47:679–85.

Council of Scientific Affairs. Aspartame: review of safety issues. JAMA 1985; 254:400–2.

Cregler LL, Mark H. Medical complications of cocaine abuse. N Engl J Med 1986; 315:1495–501.

Dhopesh V, Maany I, Herring C. The relationship of cocaine to headache in polysubstance abusers. Headache 1991; 31:17–9.

Dhuna A, Pascual-Leone A, Belgrade M. Cocaine-related vascular headaches. J Neurol Neurosurg Psychiat 1991;54:803–6.

De Marinis M, Janiri L, Agnoli A. Headache in the use and withdrawal of opiates and other associated substances of abuse. Headache 1991; 31:159–63.

Ekbom K. Nitroglycerin as a provocative agent in cluster headache. Arch Neurol 1968; 19:487–93.

el-Mallakh RS. Marijuana and migraine. Headache 1987; 27:442–3.

el-Mallakh RS, Kranzler HR, Kamanitz JR. Headaches and psychoactive substance use. Headache 1991; 31:584–7.

Forbes HS, Cobb S, Fremont-smith F. Cerebral edema and headache following carbon monoxide asphyxia. Arch Neurol Psychiatr 1924; 11:164.

Gawin FH. Cocaine addiction: psychology and neurophysiology. Science 1991; 251:1580–6.

Ghose K, Carrol JD. Mechanisms of tyramine-induced migraine: similarities with dopamine and interactions with disulfiram and propranolol. Neuropsychiobiol 1984; 12:122–6.

Gordon CR, Mankuta D, Shupak A, Spitzer O, Doweck I. Recurrent classic migraine attacks following transdermal scopolamine intoxication. Headache 1991; 31:172–4.

Gore ME, Salmon PR. Chinese restaurant syndrome: fact or fiction. Lancet 1980; 318:251–2.

Hanington E, Harper AM. The role of tyramine in the etiology of migraine and related studies on the cerebral and intracerebral circulations. Headache 1968; 8:84–97.

Hansen HJ, Drewes VM. The nitroglycerine ointment test – a double-blind examination. Dan Med Bull 1970; 7:226–9.

Heckerling PS, Leikin JB, Maturen A, Perkins JT. Predictors of occult carbon monoxide poisoning in patients with headache and dizziness. Ann Int Med 1987; 107:174–6.

Henderson WR, Raskin NH. 'Hot dog' headache: individual susceptibility to nitrite. Lancet 1972; ii:1162–3.

Hirsch AR, Rankin KM, Panelli PP. Trichloroethylene exposure and headache. Headache Quarterly 1996; 7:126–38.

Horowitz LD, Herman MV, Gorlin R. Clinical response to nitroglycerine as a diagnostic test for coronary artery disease. Am J Cardiol 1972; 29:149–53.

Iversen HK, Nielsen TM, Olesen J, Tfelt-Hansen P. Intravenous nitroglycerin as an experimental model of vascular headache. Basic characteristics. Pain 1989; 38:17–24.

Kenney RA, Tidball CS. Human susceptibility to oral monosodium 1-glutamate. Am J Clin Nutr 1972; 25:140–6.

Kerr GR, Lee MW, elLozy M, McGandy R, Stare F. Prevalence of the 'Chinese restaurant syndrome'. J Am Diet Assoc 1979; 75:29–33.

Krabbe AA, Olesen J. Headache provocation by continuous intravenous infusion of histamine, clinical results and receptor mechanisms. Pain 1980; 8:253–9.

Larkin JM, Brahos GJ, Moylin JA. Treatment of carbon monoxide poisoning: prognostic factors. J Trauma 1976; 16:111.

Lassen LH, Thomsen LL, Olesen J. Histamine induces migraine via the H receptor. Support for the NO-hypothesis of migraine. Neuroreport 1995; 6: 1475–9.

Leon AS, Hunninghake DB, Bell C, Rassin DK, Tephly TR. Safety of long-term doses of aspartame. Arch Int Med 1989; 149:2318–24.

Leone M, Attanasio A, Croci D, Filippini G, D'Amico D, Grazzi L, Nespolo A, Bussone G. The serotonergic agent m-chlorophenylpiperazine induced migraine attacks: a controlled study. Neurology 2000; 55:136–9.

Levine SR, Brust JC, Futrell N, et al. Cerebrovascular complications of the use of the crack form of alkaloidal cocaine. N Engl J Med 1990; 323:699–704.

Lipton RB, Kwong CM, Solomon S. Headaches in hospitalized cocaine users. Headache 1989; 29:225–8.

Lowenstein DH, Massa SM, Rowbotham MC et al. Acute neurologic and psychiatric complications associated with cocaine abuse. Am J Med 1987; 83:841–6.

Luthy J, Schlatter C. Biogenic amines in food: effects of histamine, tyramine and phenylethylamine in the human. Z Lebensm Unters Forsch 1983; 177:439–43.

McCullock J, Harper AM. Phenylethylamine and the cerebral circulation. In *Current concepts in migraine research* (McCullock J, Harper AM, eds.). Raven Press, New York. 1978, pp. 85–8.

Meigs JL, Hughes JP. Acute carbon monoxide poisoning: an analysis of 105 cases. AMA Arch Ind Hygiene Occupat Med 1952; 6:344–56.

Merkel PA, Koroshetz WJ, Irizarry MC, Cudkovicz ME. Cocaine-associated cerebral vasculitis. Semin Arthritis Rheum 1995; 25:172–83.

Merrit JE, Williams PB. Vasospasm contributes to monosodium glutamate-induced headache. Headache 1990; 30:575–80.

Mitchell JD. Clinical neurotoxicology: an introduction. In *Handbook of Clinical Neurology* (DeWolff FA, ed.). Elsevier Science, 1995, pp. 1–22.

Moffet AM, Swash M, Scott DF. Effect of chocolate in migraine: a double-blind study. J Neurol Neurosurg Psychiatry 1974; 37:445–8.

Monteiro JM. Headache associated with single use of substances. In *The headaches* (Olesen J, Tfelt-Hansen P, Welch KMA, eds.). Raven Press, Limited, New York. 1993, pp. 715–20.

Monteiro JM, Dahlof CG. Single use of substances. In *The Headaches* (Olesen J, Tfelt-Hansen P, Welch KM, eds.). Lippincott Williams & Wilkins, Philadelphia. 2000, pp. 861–9.

Murphree AB, Greenberg LA, Carrol RB. Neuropharmacologic effects of substances other than ethanol in alcoholic beverages. Fed Proc 1967; 26:1468–73.

Norton S. Toxicology of the central nervous system. In *Toxicology* (Casarett LJ, Doult J, eds.). MacMillan, New York. 1975, pp. 1019–35.

Ogata S, Hosoi T, Saji H. Studies on acute alcohol intoxication. Japanese Journal of Studies of Alcohol 1966; 1:67–79.

Peters GA. Migraine: diagnosis and treatment with emphasis on the migraine-tension headache, provocative tests and use of rectal suppositories. Proc Mayo Clin 1953; 28:673–86.

Reif-Lehrer L. A questionnaire study of the prevalence of Chinese restaurant syndrome. Fed Proc 1977; 36:1617–23.

Sabatini U, Rascol O, Rascol A, Montastruc JL. Migraine attacks induced by subcutaneous apomorphine in two migrainous Parkinson Ian patients. Clin Neuropharmacol 1990; 13:264–7.

Satel SL, Gawin FH. Migraine-like headache and cocaine use. JAMA 1989; 261:2995–6.

Schamburg HH, Byck R, Gerstl R, Mashman JH. Monosodium L-glutamate: its pharmacology and role in the Chinese restaurant syndrome. Science 1969; 163:826–8.

Scher W, Scher BM. A possible role for nitric oxide in glutamate (MSG)-induced Chinese restaurant syndrome, glutamate induced asthma, 'hot-dog headache', pugilistic Alzheimer's disease, and other disorders. Med Hypotheses 1992; 38:185–8.

Schiffmann SS, Buckley CE, Sampson HA et al. Aspartame and susceptibility to headache. N Engl J Med 1987; 317:1181–5.

Schnitker MT, Schnitker MA. Clinical notes, suggestions and new instruments. JAMA 1947; 135:89.

Schwartz AM. The cause, relief and prevention of headache arising from contact with dynamite. N Engl J Med 1946; 235:541–4.

Scopp AL. MSG and hydrolyzed vegetable protein induced headache review and case studies. Headache 1991; 31:107–10.

Seltzer S. Foods and drug combinations, responsible for head and neck pain. Cephalalgia 1982; 2:111–24.

Shaw SW, Johnson RH, Keogh HG. Oral tyramine in dietary migraine sufferers. In *Current concepts in migraine research* (Shaw SW, Johnson RH, Keogh HG, eds.). Raven Press, New York. 1978, pp. 31–9.

Shively M, Riegel B. Effect of nitroglycerin ointment placement on headache and flushing in health subjects. Int J Nurs Stud 1991; 28:153–61.

Sicuteri F, Bene ED, Poggioni M, Bonazzi A. Unmasking latent dysnociception in healthy subjects. Headache 1987; 27:180–5.

Smith I, Kellow AH, Hanington E. Clinical and biochemical correlation between tyramine and migraine headache. Headache 1970; 10:43–52.

Tarasoff L, Kelly MF. Monosodium L-glutamate: a double-blind study and review. Food Chem Toxicol 1993; 31:1019–35.

Thomsen LL, Kruse C, Iversen HK, Olesen J. A Nitric oxide donor triggers genuine migraine attacks. Eur J Neurol 1994; 1:71–80.

Towers CV, Pircon RA, Nageotte MP, Porto M, Garite TJ. Cocaine intoxication presenting as preeclampsia and eclampsia. Ob Gyn 1993; 81:545–7.

Trelles L, Jeri R. Central nervous system stimulants: cocaine, amphetamine, nicotine. In *Handbook of clinical neurology* (DeWolff FA, ed.). New York: Elsevier Science, 1995, pp. 251–7.

Wallgreen H, Barry A. Drug actions in relation to alcohol effects. In: Action of alcohol. New York: Elsevier, 1970, pp. 621–714.

Wilson J. Cyanogenic glycosides. Ch. 3 in *Handbook of clinical neurology*, vol. 65 (DeWolff FA, ed.). New York: Elsevier Science, 1995, pp. 25–34.

Varon J, Marik PE, Fromm RE, Gueler A. Carbon Monoxide Poisoning: a review for clinicians. J Emerg Med 1999; 17:87–93.

Yang WH, Drouin MA, Herbert M, Mao Y, Kursh J. The monosodium glutamate symptom complex: assessment in a double blind, placebo controlled, randomized study. J Allergy Clin Immunol 1997; 99:757–62.

8.2 Medication-overuse headache

Ala-Hurula V, Myllyla V, Hokkanen E. Ergotamine abuse: results of ergotamine discontinuation with special reference to the plasma concentrations. Cephalalgia 1982; 2:189–95.

Ala-Hurula V, Myllyla V, Hokkanen E, Tokola O. Tolfenamic acid and ergotamine abuse. Headache 1981; 21:240–2.

Allgulander C. History and current status of sedative-hypnotic drug use and abuse. Acta Psychiatr Scand 1986; 73:465–78.

Andersson PG. Ergotamine headache. Headache 1975; 15:118–21.

Baar HA. Treatment for headache: a four-step standardized withdrawal program for analgesic abusers. Pain Clin 1990; 3:173–7 (Abstract).

Bennett WM, DeBroe ME. Analgesic nephropathy: a preventable renal disease. N Eng J Med 1989; 320:1269–71.

Bowdler I, Killian J, Gänsslen-Blumberg S. The association between analgesic abuse and headache – coincidental or causal. Headache 1988; 28:494.

Braithwaite RA. The toxicity of tricyclic and newer antidepressants. In *Handbook of Clinical Neurology* (DeWolff FA, ed.). New York: Elsevier Science, 1995, pp. 311–20.

Brust JC. Opiate addiction and toxicity. Ch. 16 in *Handbook of clinical neurology*, vol. 65 (DeWolff FA, ed.). New York: Elsevier Science, 1995, pp. 356–61.

Catarci T, Fiacco F, Argentino C. Ergotamine-induced headache can be sustained by sumatriptan daily intake. Cephalalgia 1994; 14:374–5.

Centonze V, Polite BM, diBari M, Caporaletti P, Albano O. Vascular injuries in ergotamine abuse: a case report. Funct Neurol 1993; 8:265–70.

Dalquen P, Fasel J, Mihatsch MJ, Rist M, Rutishauser G. Phenacetinabusus IV. Sind zytologische harnuntersuchungen in der tumorvorsorge bei phenacetinabusern erfolgversprechend und anwendbar. Schweizerische Medizinische Wochenschrift 1980; 110:302–6.

DeBroe ME, Elseviers MM. Analgesic nephropathy – still a problem? Nephron 1993; 64:505–13.

deMarinis M, Janiri L, Agnoli A. Headache in the use and withdrawal of opiates and other associated substances of abuse. Headache 1991; 31:159–63.

Diamond S, Dalessio DJ. Drug abuse in headache. In *The practicing physician's approach to headache* (Diamond S, Dalessio DJ, eds.). Williams & Wilkins, Baltimore. 1982, pp. 114–21.

Dichgans J, Diener HC. Clinical manifestations of excessive use of analgesic medication. In *Drug – induced headache* (Diener HC, Wilkinson M, eds.). Springer-Verlag, Berlin. 1988, pp. 8–15.

Dichgans J, Diener HD, Gerber WD et al. Analgetika-induzierter dauerkopfschmerz. Dtsch Med Wschr 1984, 109:369–73.

Diener HC. A personal view of the classification and definition of drug dependence headache. Cephalalgia 1993; 13:68–71.

Diener HC, Dahlof CG. Headache associated with chronic use of substances. In *The headaches* (Olesen J, Tfelt-Hansen P, Welch KMA, eds.). Lippincott, Williams & Wilkins, Philadelphia. 1999, pp. 871–8.

Diener HC, Dichgans J, Scholz E, Geiselhart S, Gerber WD, Bille A. Analgesic-induced chronic headache: long-term results of withdrawal therapy. J Neurol 1989; 236:9–14.

Diener HC, Haab J, Peters C, Ried S, Dichgans J, Pilgrim A. Subcutaneous sumatriptan in the treatment of headache during withdrawal from drug-induced headache. Headache 1991; 31:205–9.

Diener HC, Pfaffenrath V, Soyka D, Gerber WD. Therapie des medikamenten-induzierten dauerkopfschmerzes. Münch Med Wschr 1992; 134:159–62.

Diener HC, Tfelt-Hansen P. Headache associated with chronic use of substances. In *The headaches* (Olesen J, Tfelt-Hansen P, Welch KMA, eds.). Raven press LTD, New York. 1993, pp. 721–7.

Dige-Petersen H, Lassen NA, Noer J, Toennesen KH, Olesen J. Subclinical ergotism. Lancet 1977; i:65–6.

Drucker P, Tepper S. Daily sumatriptan for detoxification from rebound. Headache 1998; 38:687–90.

Dubach UC, Rosner B, Pfister E. Epidemiologic study of abuse of analgesics containing phenacetin. Renal morbidity and mortality (1968–1979). N Engl J Med 1983; 308:357–62.

Elkind AH. Drug abuse in headache patients. Clin J Pain 1989; 5:111–20.

Elkind AH. Drug abuse and headache. Med Clin N Am 1991; 75:717–32.

Evers S, Gralow I, Bauer B, Suhr B, Buchheister A, Husstedt IW, Ringelstein EB. Sumatriptan and ergotamine overuse and drug-induced headache: a clinicoepidemiologic study. Clin Neuropharmacol 1999; 22:201–6.

Fanciullaci M, Alessandri M, Pietrini U, Briccolani-Bandini E, Beatrice S. Long-term ergotamine abuse: effect on adrenergically induced mydriasis. Clin Pharm Ther 1992; 51:302–7.

Fincham JE. Over-the-counter drug use and misuse by the ambulatory elderly: a review of the literature. J Ger Drug Ther 1987; 1:3–21.

Fincham RW, Perdue Z, Dunn VD. Bilateral focal cortical atrophy and chronic ergotamine abuse. Neurology 1985; 35:720–2.

Fisher CM. Analgesic rebound headache refuted. Headache 1988; 28:666.

Friedman AP, Brazil P, vonStorch TJ. Ergotamine tolerance in patients with migraine. JAMA 1955; 157:881–4.

Gaist D, Hallas J, Sindrup SH, Gram LF. Is overuse of sumatriptan a problem? A population-based study. Eur J Clin Pharmacol 1996; 50:161–5.

Gaist D, Tsiropoulus I, Sindrup SH, Hallas J, Rasmussen BK, Kragstrup J. Inappropriate use of sumatriptan: population based register and interview study. Br J Med 1998; 316:1352–3.

Granella F, Farina S, Malferrari G, Manzoni GC. Drug abuse in chronic headache: a clinicoepidemiologic study. Cephalalgia 1987; 7:15–9.

Gutzwiller F, Zemp E. Der analgetikakonsum in der bevölkerung und socioökonomische aspekte des analgetikaabusus. In *Das analgetikasyndrom* (Mihatsch MJ, ed.). Thieme, Stuttgart. 1986, pp. 197–205.

Hering R, Steiner TJ. Abrupt outpatient withdrawal from medication in analgesic-abusing migraineurs. Lancet 1991; 337:1442–3.

Hokkanen E, Waltimo O, Kallanranta T. Toxic effects of ergotamine used for migraine. Headache 1978; 18:95–8.

Horowski R, Ziegler A. Possible pharmacological mechanisms of chronic abuse of analgesics and other antimigraine drugs. In *Drug-induced headache* (Diener HC, Wilkinson M, eds.). Springer-Verlag, Berlin. 1988, pp. 95–104.

Horton BT, Peters GA. Clinical manifestations of excessive use of ergotamine preparations and management of withdrawal effect: report of 52 cases. Headache 1963; 3:214–26.

Isler H. Migraine treatment as a cause of chronic migraine. In *Advances in migraine research and therapy* (Rose FC, ed.). Raven Press, New York. 1982, pp. 159–64.

Jaffe JH. Drug addiction and drug abuse. In *The pharmacological basis of therapeutics* (Gilman AG, Rall TW, Nies AS, Taylor P, eds.). Pergamon Press, New York. 1985, pp. 522–73.

Katsarava Z, Fritsche G, Muessig M, Diener HC, Limmroth V. Clinical features of withdrawal headache following overuse of triptans and other headache drugs. Neurology 2001; 57:1694–8.

Kaube H, May A, Diener HC, Pfaffenrath V. Sumatriptan misuse in daily chronic headache. Br Med J 1994; 308:1573.

Kielholz P, Ladewig D. Probleme des medikamentenmi betabrauches. Schweiz Arztezeitung 1981; 62:2866–9.

Klapper JA. Rebound headache: definition, symptomatology, treatment, and prevention. Headache Quarterly 1992; 3:398–402.

Kouyanou K, Pither CE, Rabe-Hesketh S, Wessely S. A comparative study of iatrogenesis, medication abuse, and psychiatric morbidity in chronic pain patients with and without medically explained symptoms. Pain 1998; 76:417–26.

Kudrow L. Paradoxical effects of frequent analgesic use. Adv Neurol 1982; 33:335–41.

Lader M. Hypnotics and sedatives. In *Handbook of clinical neurology* (DeWolff FA, ed.). New York: Elsevier Science, 1995, pp. 329–55.

Lance F, Parkes C, Wilkinson M. Does analgesic abuse cause headache de novo? Headache 1988; 28:61–2.

Lance JW. A concept of migraine and the search for the ideal headache drug. Headache 1990; 30:17–23.

Limmroth V, Kazarawa S, Fritsche G, Diener HC. Headache after frequent use of new 5-HT agonists zolmitriptan and naratriptan. Lancet 1999; 353:378.

Limmroth V, Katsarava Z, Fritsche G, Przywara S, Diener HC. Features of medication overuse headache following overuse of different acute headache drugs. Neurology 2002; 59:1011–14.

Lucas RN, Falkowski W. Ergotamine and methysergide abuse in patients with migraine. Br J Psychiatry 1973; 122:199–203.

Ludolph AC, Husstedt IW, Schlake HP, Grotemeyer KH, Brune GG. Chronic ergotamine abuse: evidence of functional impairment of long ascending spinal tracts. Eur Neurol 1988; 28:311–6.

MacGregor EA, Vorah C, Wilkinson M. Analgesic use: a study of treatments used by patients for migraine prior to attending the City of London migraine clinic. Headache 1990; 30:634–8.

Manzoni GC, Micieli G, Granella F, Sandrini G, Zanferrari C, Nappi G. Therapeutic approach to drug abuse in headache patients. In *Drug-induced headache* (Diener HC, Wilkinson M, eds.). Springer-Verlag, Berlin. 1988, pp. 143–9.

Marks V. Reactive (rebound) hypoglycemia. In *Hypoglycemia* (Marks V, Rose CF, eds.). Blackwell, Oxford. 1981, pp. 179–217.

Mathew NT. Amelioration of ergotamine withdrawal symptoms with naproxen. Headache 1987; 27:130–3.

Mathew NT, Kurman R, Perez F. Drug induced refractory headache – clinical features and management. Headache 1990; 30:634–8.

Michultka DM, Blanchard EB, Appelbaum KA, Jaccard J, Dentinger MP. The refractory headache patient-2. High medication consumption (analgesic rebound) headache. Behav Res Ther 1989; 27:411–20.

Micieli G, Manzoni GC, Granella F, Martignoni E, Malferrari G, Nappi G. Clinical and epidemiological observations on drug abuse in headache patients. In *Drug-Induced Headache* (Diener HC, Wilkinson M, eds.). Springer-Verlag, Berlin. 1988, pp. 20–8.

Nicolodi M, DelBianco PL, Sicuteri F. The way to serotonergic use and abuse in migraine. Int J Clin Pharmacol Res 1997; 17:79–84.

Page H. Rebound headache from ergotamine withdrawal. JAMA 1981; 246:719.

Peters G, Horton BT. Headache: with special reference to the excessive use of ergotamine preparations and withdrawal effects. Proc Mayo Clin 1951; 26:153–61.

Pini LA, Trenti T. Case report: does chronic use of sumatriptan induce dependence? Headache 1994; 34:600–1.

Pradalier A, Dry S, Baron JF. Cephalée induite par l'abuse de tartrate d'ergotamine chez les migrainieux. Concours Méd 1984; 106:106–10.

Rahman A, Segasothy M, Samad SA, Zulfiqar A, Rani M. Analgesic use and chronic renal disease in patients with headache. Headache 1993; 33:442–5.

Rapoport A, Stang P, Gutterman DL, Cady R, Markley H, Weeks R, Saiers J, Fox AW. Analgesic rebound headache in clinical practice: data from a physician survey. Headache 1996; 36:14–9.

Rapoport AM. Analgesic rebound headache. Headache 1988; 28:662–5.

Rapoport AM, Weeks RE. Characteristics and treatment of analgesic rebound headache. In *Drug-induced headache* (Diener HC, Wilkinson M, eds.). Springer-Verlag, Berlin. 1988, pp. 162–7.

Roswell AR, Neylan C, Wilkinson M. Ergotamine induced headache in migrainous patients. Headache 1973; 13:65–7.

Sandler DP, Smith JC, Weinberg CR, Buckalew VM, Dennis VW, Blythe WB, Burgess WP. Analgesic use and chronic renal disease. N Engl J Med 1989; 320:1238–43.

Saper JR. Drug abuse among headache patients. In *Headache disorders* (Saper JR, ed.). PSG Publishers, Boston. 1983, pp. 263–78.

Saper JR. Drug overuse among patients with headache. Neurol Clin 1983; 1:465–77.

Saper JR. Daily chronic headaches. Neurol Clin N Amer 1990; 8:891–902.

Saper JR, Jones JM. Ergotamine tartrate dependency: features and possible mechanisms. Clin Neuropharmacol 1986; 9:244–56.

Schnider P, Aull S, Baumgartner C et al. Long-term outcome of patients with headache and drug abuse after inpatient withdrawal: five-year followup. Cephalalgia 1996; 16:481–5.

Schnider P, Aull S, Feucht M. Use and abuse of analgesics in tension-type headache. Cephalalgia 1994; 14:162–7.

Schnider P, Maly J, Grunberger J, Aull S, Zeiler K, Wessely P. Improvement of decreased critical flicker frequency in headache patients with drug abuse after successful withdrawal. Headache 1995; 35:269–72.

Schoenen J, Lenarduzzi P, Sianard-Gainko J. Chronic headaches associated with analgesics and/or ergotamine abuse: a clinical survey of 434 consecutive outpatients. In *New advances in headache research* (Rose FD, ed.). Smith-Gordon, London. 1989, pp. 29–43.

Seller EM, Busto UE, Kaplan HL, Somer G, Baylon GJ. Comparative abuse liability of codeine and naratriptan. Clin Pharmacol Ther 1998; 63:121.

Shakir RA. Vitamin toxicity. In *Handbook of clinical neurology*, vol. 65 (DeWolff FA, ed.). New York: Elsevier Science, 1995, pp. 567–76.

Silberstein SD, Lipton RB. Chronic daily headache. In *Headache* (Goadsby PJ, Silberstein SD, eds.). Butterworth-Heinemann, Newton. 1997, pp. 201–25.

Silberstein SD, Lipton RB, Solomon S, Mathew NT. Classification of daily and near daily headaches: proposed revisions to the IHS classification. Headache 1994; 34:1–7.

Silberstein SD, Silberstein JR. Chronic daily headache: prognosis following inpatient treatment with repetitive IV DHE. Headache 1992; 32:439–45.
Stewart JH. Analgesic abuse and renal failure in Australia. Kidney International 1978; 13:72–8.
Sullivan JT, Sellers EM. Treatment of the barbiturate abstinence syndrome. Med J Aust 1986; 145:456–8.
Taschner KL, Wiesbeck GA. Psychiatric aspects of drug addiction of the barbiturate-alcohol type. In *Drug-induced headache* (Diener HC, Wilkinson M, eds.). Berlin, Springer-Verlag. 1988, pp. 80–4.
Tfelt-Hansen P. Ergotamine headache. In *Updating in headache* (Pfaffenrath V, Lundberg P, Sjaastad O, eds.). Springer, Berlin. 1985, pp. 169–72.
Tfelt-Hansen P. The effect of ergotamine on the arterial system in man. Acta Pharmacol Toxicol 1986; 59:1–29.
Tfelt-Hansen P, Krabbe AA. Ergotamine. Do patients benefit from withdrawal? Cephalalgia 1981; 1:29–32.
Tfelt-Hansen P, Olesen J. Arterial response to ergotamine tartrate in abusing and non-abusing migraine patients. Acta Physiol Scand 1981; 48:69–72.
Tfelt-Hansen P, Paalzow L. Intramuscular ergotamine: plasma levels and dynamic activity. Clin Pharmacol Ther 1985; 37:29–35.
Tfelt-Hansen P, Saxena PR, Ferrari MD. Ergot alkaloids. In *Handbook of clinical neurology* (DeWolff FA, ed.). New York: Elsevier Science. 1995, pp. 61–7.
Timmings PL, Richens A. Neurotoxicology of antiepileptic drugs. In *Handbook of clinical neurology* (DeWolff FA, ed.). New York: Elsevier Science. 1995, pp. 495–513.
Vasconcellos E, Pina-Garza JE, Millan EJ, Warner JS. Analgesic rebound headache in children and adolescents. J Child Neurol 1998; 13:443–7.
Verhoeff NP, Visser WH, Ferrari MD, Saxena PR, vonRoyen EA. Dopamine D2 receptor imaging with 123-I-iodobenzamide SPECT in migraine patients abusing ergotamine: does ergotamine cross the blood brain barrier. Cephalalgia 1993; 13:325–9.
VonKorff M, Galer BS, Stang P. Chronic use of symptomatic headache medications. Pain 1995; 62:179–86.
Walker J, Parisi S, Olive D. Analgesic rebound headache: experience in a community hospital. Southern Med J 1993; 86:1202–5.
Warner JS. Rebound headaches: a review. Headache Quarterly 1999; 10:207–19.
Zed PJ, Loewen PS, Robinson G. Medication-induced headache: overview and systematic review of therapeutic approaches. Ann Pharmacother 1999; 33:61–72.
Ziegler DK. Opiate and opioid use in patients with refractory headache. Cephalalgia 2000; 14:5–10.

8.3 Headache as an adverse event attributed to chronic medication

Dalton K. Migraine and oral contraceptives. Headache 1976; 15: 247–51.
de Lignières B, Silberstein SD. Pharmacodynamics of oestrogens and progestagens. Cephalalgia 2000; 20:200–7.
Magos AL, Brewster E, Singh R, O'Dowd T, Brincat M, Studd JWW. The effects of norethisterone in postmenopausal women on oestrogen replacement therapy: a model for the premenstrual syndrome. Br J Obstet Gynaecol 1986; 93:1290–6.
Nappi RE, Cagnacci A, Granella F, Piccinini F, Polatti F, Facchinetti F. Course of primary headaches during hormone replacement therapy. Maturitas 2001; 38:157–63.
Silberstein SD. Hormone-related headache. M Clin N Am 2001; 85:1017–35.
Silberstein SD, de Lignières B. Migraine, menopause and hormonal replacement therapy. Cephalalgia 2000; 20:214–2.

8.4 Headache attributed to substance withdrawal

Abbott PJ. Caffeine: a toxicological overview. Med J Aust 1986; 145:518–21.
Baumgartner GR, Rowen RC. Transdermal clonidine versus chlordiazepoxide in alcohol withdrawal: a randomized, controlled clinical trial. South Med J 1991; 84:312–21.
Dalessio DJ. (1980). *Wolff's headache and other head pain*, Oxford University Press, Oxford.
Epstein MT, Hockaday JM, Hockaday TDR. Migraine and reproductive hormones through the menstrual cycle. Lancet 1975; i:543–8.
Greden JF, Fontaine M, Lubetsky M, Chamberlin K. Anxiety and depression associated with caffeinism among psychiatric inpatients. Am J Psychiatr 1978; 135:963–6.
Laska EM, Sunshine A, Mueller F, Elvers WB, Siegel C, Rubin A. Caffeine as an analgesic adjuvant. JAMA 1984; 251:1711–8.
Lichten E, Lichten J, Whitty A, Pieper D. The confirmation of a biochemical marker for women's hormonal migraine: the depo-oestradiol challenge test. Headache 1996; 36: 367–71.
Raskin NH, Appenzeller O. *Headache*, Saunders, Philadelphia. 1980.
Silverman K, Evans SM, Strain EC, Griffiths RR. Withdrawal syndrome after the double-blind cessation of caffeine consumption. N Eng J Med 1992; 327:1109–14.
Somerville B. Estrogen withdrawal migraine. Neurology 1975; 25:239–50.
vanDusseldorp M, Katan MB. Headache caused by caffeine withdrawal among moderate coffee drinkers switched from ordinary to decaffeinated coffee: a 12 week double blind trial. Br Med J 1990; 300:1558–9.
White BC, Lincoln CA, Pearcz NW, Reeb R, Vaida C. Anxiety and muscle tension as consequence of caffeine withdrawal. Science 1980; 209:1547–8.

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[1]
C. Headache develops in close temporal relation to meningitis
D. Headache resolves within 3 months[2] 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[1]
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[1] 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[2], by neuroimaging, CSF examination, EEG and/or 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 C*omments*.

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.

Bibliography and reference

9.1.1 Headache attributed to bacterial meningitis

Drexler ED. Severe headache: when to worry, what to do. Postgrad Med 1990; 87:164–70, 173–80.

Francke E. The many causes of meningitis. Postgrad Med 1987; 82:175–8, 181–3, 187–8.

Gedde-Dahl TW, Lettenstrom GS, Bovre K. Coverage for meningococcal disease in the Norwegian morbidity and mortality statistics. NIPH Ann 1980; 3(2):31–5.

Jones HR, Siekert RG. Neurological manifestation of infective endocarditis. Brain 1989; 112:1295–315.

Tonjum T. Nilsson F, Bruun JH, Hanebeg B. The early phase of meningococcal disease. NIPH Ann 1983; 6:175–81.

Zhang SR, Zhang YS, Zhao XD. Tuberculous meningitis with hydrocephalus: a clinical and CT study. Chung Hua Nei Ko Tsa Chih 1989; 28:202–4.

9.1.2 Headache attributed to lymphocytic meningitis

Cochius JI, Burns RJ, Willoughby JO. CNS cryptococcosis: unusual aspects. Clin Exp Neurol 1989; 26:183–91.

Dalton M, Newton RW. Aseptic meningitis. Dev Med Child Neurol 1991; 33:446–58.

Gomez-Arada F, Canadillas F, Marti-Masso FJ et al. Pseudomigraine with temporary neurological symptoms and lymphocytic pleocytosis. Brain 1997; 120:1105–13.
Mak SC, Jeng JE, Jong JY, Chiang CH, Chou LC. Clinical observations and virological study of aseptic meningitis in the Kaohsinug area. Taiwan I Hsueh Hui Twa Chih 1990; 89:868–72.
Pachner AR, Steere AC. Neurological findings of Lyme disease. Yale Biol Med 1984; 57:481–3.
Pachner AR, Steere AC. The triad of neurologic manifestations of Lyme disease: meningitis, cranial neuritis, and radiculoneuritis. Neurology 1985; 35:47–53.
Singer JI, Maur PR, Riley JP, Smith PB. Management of central nervous system infections during an epidemic of enteroviral aseptic meningitis. J Pediatr 1980; 96:559–63.

9.1.3 Headache attributed to encephalitis

Brooks RG, Licitra CM, Peacock MG. Encephalitis caused by Coxiella burnetii. Ann Neurol 1986; 20:91–3.
Davis LE, McLaren LC. Relapsing herpes simplex encephalitis following antiviral therapy. Ann Neurol 1983; 13:192–5.
Domachowske JB, Cunningham CK, Cummings DL, Crosley CJ, Hannan WP, Weiner LB. Acute manifestations and neurologic sequelae of Epstein-Barr virus encephalitis in children. Pediatr Infect Dis J 1996; 15:871–5.
Kennedy PG. Retrospective analsys of 46 cases of simplex encephalitis seen in Glasgow between 1962 and 1985. OJM 1988; 86:533–40.
Kennedy PG, Adams IH, Graham DI, Clements GB. A clinicopathological study of herpes simplex encephalitis. Neuropathol Appl Neurobiol 1998; 14:395–415.
Poneprasert B. Japanese encephalitis in children in northern Thailand. Southeast Asian J Trop Med Public health 1989; 20:599–603.
Saged JI, Weinstein Mo, Miller DC. Chronic encephalitis possibly due to herpes simplex virus: two cases. Neurology 1985; 35:1470–2.

9.1.4 Headache attributed to brain abscess

Chalstrey S, Pfleiderer AG, Moffat DA. Persisting incidence and mortality of sinogenic cerebral abscess: a continuing reflection of late clinical diagnosis. J R Soc Med 1991; 84:193–5.
Chun CH, Johnson JD, Hofstetter M, Raff MJ. Brain abscess: a study of 45 consecutive cases. Medicine 1986; 65:415–31.
Harris LF, Maccubbin DA, Triplett JN, Haws FB. Brain abscess: recent experience at a community hospital. South Med J 1985; 78:704–7.
Kulay A, Ozatik N, Topucu I. Otogenic intracranial abscesses. Acta Neurochir (Wien) 1990; 107:140–6.
Yen PT, Chan ST, Huang TS. Brain abscess: with spcial reference to otolaryngologic sources of infection. Otolaryngol Head Neck Surg 1995; 113:15–22.

9.1.5 Headache attributed to subdural empyema

Hodges J, Anslow P, Gillet G. Subdural empyema: continuing diagnostic problems in the CT scan era. QJM 1986; 59:387–93.
McIntyre PB, Lavercombe PS, Kemp RJ, McCormack JG. Subdural and epidural empyema: diagnostic and therapeutic problems. Med J Aust 1991; 154:653–7.
Sellik JA. Epidural abscess and subdural empyema. J Am Osteopath Assoc 1989; 89:806–10.

9.2 Headache attributed to systemic infection

De Marinis M, Welch KM, Headache associated with noncephalic infections: classification and mechanisms. Cephalalgia 1992; 12:197–201.

9.3 Headache attributed to HIV/AIDS

Brew BJ, Miller J. Human immunodeficiency virus-related headache. Neurology 1993; 43:1098–100.
Denning DW. The neurological features of HIV infection. Biomed Pharmacother 1988; 42:11–4.
Evers S, Wibbeke B, Reichelt D, Suhr B, Brilla R, Husstedt IW. The impact of HIV infection on primary headache. Unexpected findings from retrospective, cross-sectional, and prospective analyses. Pain 2000; 85:191–200.
Hollander H, Strimgari S. Human immunodeficiency virus-associated meningitis. Clinical course and correlations. Am J Med 1987; 83:813–6.
Rinaldi R, Manfredi R, Azzimondi G et al. Recurrent 'migrainelike' episodes in patients with HIV disease. Headache 1007; 37:443–8.
Weinke T, Rogler G, Sixt C et al. Cryptococcosis in AIDS patients: observations concerning CNS involvement. J Neurol 1989; 236:38–42.

9.4 Chronic post infection headache

Bohr V, Hansen B, Kjersen H, Rasmussen N, Johnsen N, Kristensen HS, Jessen O. Sequelae from bacterial meningitis and their relation to the clinical condition during acute illness, based on 667 questionnaire returns. Part II of a three part series. J Infect 1983; 7:102–10.

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 phaeochromocytoma
- 10.3.2 Headache attributed to hypertensive crisis without hypertensive encephalopathy
- 10.3.3 Headache attributed to hypertensive encephalopathy
- 10.3.4 Headache attributed to pre-eclampsia
- 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 cephalalgia
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_2 <70 mm Hg or in chronically hypoxic patients with PaO_2 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_2 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_2

Comments:
Hypercapnia (arterial PCO_2 >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_2 production more than 10-fold, resulting in a transient elevation of PCO_2 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 ≥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. The 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

Comments:
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 phaeochromocytoma

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. Phaeochromocytoma 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 phaeochromocytoma. 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 phaeochromocytoma has not yet been made, and hypertensive encephalopathy is not present, patients may meet the diagnostic criteria for 10.3.2 *Headache attributed to hypertensive crisis without hypertensive encephalopathy*.

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 mmHg. 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 7 days postpartum), 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 postpartum), 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 cephalalgia

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
D. 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 cephalalgia* 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 homoeostasis

Diagnostic criteria:

A. Headache fulfilling criteria C and D
B. Evidence of a disorder of homoeostasis 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 homoeostasis

Bibliography

10.1.1 High-altitude headache; 10.1.3 Sleep apnoea headache

Aldrich MS, Chauncey JB. Are morning headaches part of obstructive sleep apnea syndrome? Arch Intern Med 1990; 150:1265–7.

Appenzeller O. Altitude Headache. Headache 1972; 12:126–9.

Ginsberg MD. Carbon monoxide intoxication: clinical features, neuropathology and mechanisms of injury. J Toxicol Clin Toxicol 1985; 23:281–8.

Heckerling PS, Leikiin JB, Maturen A, Perkins JT. Predictors of occult carbon monoxide poisoning in patients with headache and dizziness. Ann Intern Med 1987; 107:174–6.

Jozefowicz RF. Neurologic manifestations of pulmonary disease. Neurologic Clinics 1989; 7:605–16.

Lipton RB, Mazer C, Newman LC, Solomon S. Sumatriptan relieves migraine-like headaches associated with carbon monoxide exposure. Headache 1997; 37:392–5.

Loh NK, Dinner DS, Foldvary DO, Skobieranda F, Yew WW. Do patients with obstructive sleep apnea wake up with headaches? Arch Intern Med 1999; 159:1765–8.

Poceta JS, Dalessio DJ. Identification and treatment of sleep apnea in patients with chronic headache. Headache 1995; 35:586–9.

Porcelli J, Gugelchuk G. A trek to the top: A review of acute mountain sickness. J Amer Osteopath Assoc 1995; 95:718–20.

Silber E, Sonnenberg P, Collier DJ, Pollard A, Murdoch DR, Goadsby PJ. Clinical features of headache at altitude: a prospective study. Neurology 2003; 60:1167–71.

10.1.2 Diving headache

Cheshire WP, Ott MC Jr. Headache in divers. Headache 2001; 41:235–47.

Edmonds RC, Greene ER, Schoene RB et al. Diving and subaquative medicine. 3rd Ed. Oxford: Butterworth-Heinemann;1992: pp 404–6.

Sliwka U, Kransney JA, Simon SG et al. Effects of sustained low-level elevations of carbon dioxide on cerebral blood flow and autoregulation of the intracerebral arteries in humans. Aviat Space Environ Med 1998; 69:299–306.

10.2 Dialysis headache

Antoniazzi AL, Bigal ME, Bordini CA, Speciali JG. Headache associated with dialysis. The IHS criteria revisited. Cephalalgia 2003; 23:146–9.

Jameson MD, Wiegmann TB. Principles, uses, and complications of hemodialysis. Medical Clinics of North America 1990; 74:945–60.

10.3 Headache attributed to arterial hypertension

Dodick DW. Recurrent short-lasting headache associated with paroxysmal hypertension: a clonidine-responsive syndrome. Cephalalgia 2000; 20:509–14.

Gus M, Fuchs FD, Pimentel M, Rosa D, Melo AG, Moreira LB. Behavior of ambulatory blood pressure surrounding episodes of headache in mildly hypertensive patients. Arch Intern Med 2001; 161:252–5.

Kruszewski P, Bieniaszewski L, Neubauer J, Krupa-Wojciechowska B. Headache in patients with mild to moderate hypertension is generally not associated with simultaneous blood pressure elevation. J Hypertension 2000; 18:437–44.

Lance JW, Hinterberger H. Symptom of pheochromocytoma with particular reference to headache, correlated with catecholamine production. Arch Neurol 1976; 33:281–8.

Land SH, Donovan T. Pre-eclampsia and eclampsia headache: classification recommendation [letter]. Cephalalgia 1999; 19:67–9.
Loh KC, Shlossberg AH, Abbott EC, Salisbury SR, Tan MH. Phaeochromocytoma: a ten-year survey. Quart J Med 1997; 90:51–60.
Mannelli M, Ianni L, Cilotti A, Conti A. Pheochromocytoma in Italy: A multicentric retrospective study. Eur J Endocrinol 1999; 141:619–624.
Thomas JE, Rooke ED, Kvale WF. The neurologists experience with pheochromocytoma. JAMA 1966; 197:754–58.
Vaughan CJ, Delanty N. Hypertensive emergencies. Lancet 2000; 356:411–17.
Walker JJ. Pre-eclampsia. Lancet 2000; 56:1260–65.
Weiss NS. Relation of high blood pressure to headache, epistaxis, and selected other symptoms. The United States Health Examination Survey of Adults. N Engl J Med. 1972; 287:631–3.
Zampaglione B, Pascale C, Marchisio M, Cavallo-Perin P. Hypertensive urgencies and emergencies. Prevalence and clinical presentation. Hypertension 1996; 27:144–7.

10.4 Headache attributed to hypothyroidism

Airaghi L, Catania A. Endocrine headache. In: Seminars in headache management. Neuroendocrinological aspects of headache, vol 4, number 4. B.C.Decker Inc, 1999: pp 1–15.
Amy JR. Tests of thyroid function in chronic headache patients. Headache 1987; 27:351–3.
Arafah BM, Prunty D, Ybarra J, Hlavin ML, Selman WR. The dominant role of increased intrasellar pressure in the pathogenesis hypopituitarism, hyperprolactinemia, and headache in patients with pituitary adenomas. J Clin Endocrinol Metab 2000; 85:1789–93.
Fenichel NM. Chronic headache due to masked hypothyroidism. Ann Intern Med 1948; 29:456–60.
Moreau T. Headache in hypothyroidism. Prevalence and outcome under thyroid hormone therapy. Cephalalgia 1988; 18:687–9.

10.5 Headache attributed to fasting

Dalton K. Food intake prior to migraine attacks. Study of 2,313 spontaneous attacks. Headache 1975; 15:188–93.
Dexter JD, Roberts J, Byer JA. The five hour glucose tolerance test and effect of low sucrose diet in migraine. Headache 1978; 18:91–4.
Malouf R, Brust JCM. Hypoglycemia: causes, neurological manifestations, and outcome. Ann Neurol 1985; 17:421–30.
Mosek AC, Korczyn AD. Yom Kippur Headache. Neurology 1995;45:1953–5.
Pearce J. Insulin induced hypoglycaemia in migraine. J Neurol Neurosurg Psychiat 1971; 34:154–6.
Service FJ. Hypoglycemic disorders. In: Wyngaarden JB, Smith LH, Bennett JC, eds. Cecil's textbook of medicine, 18th ed. Philadelphia: WB Saunders, 1992:1310–7.

10.6 Cardiac cephalalgia

Blacky RA, Rittlemeyer JT, Wallace MR. Headache angina. Am J Cardiol 1987; 60:730.
Bowen J, Oppenheimer G. Headache as a presentation of angina: reproduction of symptoms during angioplasty. Headache 1993; 33:238–239.
Fleetcroft R, Maddocks JL. Headache due to ischaemic heart disease. J R Soc Med 1985; 78:676.
Grace A, Horgan J, Breathnach K, Staunton H. Anginal headache and its basis. Cephalalgia 1997; 17:195–6.
Lefkowitz D, Biller J. Bregmatic headache as a manifestation of myocardial ischemia. Arch Neurol 1982; 39:130.
Lipton RB, Lowenkopf T, Bajwa ZH, Leckie RS, Ribeiro S, Newman LC, Greenberg MA. Cardiac cephalgia: a treatable form of exertional headache. Neurology 1997; 49:813–6.
Vernay D, Deffond D, Fraysse P, Dordain G. Walk headache: an unusual manifestation of ischemic heart disease. Headache 1989; 29:350–1.

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 3 months, 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[1]
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 *Headache attributed to retropharyngeal tendonitis* or 11.2.3 *Headache attributed to 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[1]
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[2]
   2. abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls[3]
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 ≥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

Comments:
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[1,2]
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

Bibliography

11.1 Headache attributed to disorder of cranial bone

Bhatoe HS, Deshpande GU. Primary cranial Ewing's sarcoma. Br J Neurosurg 1998; 12:165–9.

Hayashi T, Kuroshima Y, Yoshida K, Kawase T, Ikeda E, Mukai M. Primary osteosarcoma of the sphenoid bone with extensive periosteal extension – case report. Neurol Med Chir (Tokyo) 2000; 40:419–22.

Scherer A, Engelbrecht V, Nawatny J, Messing-Junger M, Reifenberger G, Modder U. MRI of the cerebellopontine angle in patients with cleidocranial dysostosis. Rofo Fortschr Geb Rontgenstr Neuen Bildgeb Verfahr 2001; 173:315–8.

11.2 Headache attributed to disorder of neck

Antonaci F, Fredriksen TA, Sjaastad O. Cervicogenic headache: clinical presentation, diagnostic criteria, and differential diagnosis. Curr Pain Headache Rep 2001; 5:387–92.

Antonaci F, Ghirmai S, Bono G, Sandrini G, Nappi G. Cervicogenic headache: evaluation of the original diagnostic criteria. Cephalalgia 2001; 21:573–83.

Bogduk N, Corrigan B, Kelly P, Schneider G, Farr R. Cervical Headache. Med J Aust 1985; 143:202–207.

Bogduk N. Cervicogenic headache: anatomic basis and pathophysiologic mechanisms. Curr Pain Headache Rep 2001; 5:382–6.

Bogduk N. Headache and the neck. In: Goadsby PJ, Silberstein SD (eds): Headache. Boston, Butterworth-Heinemann, 1997, pp. 369–381.

Fredriksen TA, Sjaastad O. Cervicogenic headache: current concepts of pathogenesis related to anatomical structure. Clin Exp Rheumatol 2000;18(2 Suppl 19):S16–8.

Göbel H, Edmeads JG. Disorders of the skull and cervical spine. In: Olesen J, Tfelt-Hansen P, Welch KMA. The Headaches. 2nd edition. Lippincott Williams & Wilkins, Philadelphia; 2000: pp. 891–898.

Göbel H. Die Kopfschmerzen. Ursachen, Mechanismen, Diagnostik und Therapie in der Praxis. Springer Verlag, Berlin, Heidelberg, New York, London, Paris, Tokyo, Hong Kong, Barcelona, Budapest; 1997: pp. 1–901.

Lance JW, Anthony M. Neck-tongue syndrome on sudden turning of the head. J Neurol Neurosurg Psychiat 1980; 43:97–101.

Leone M, D'Amico D, Grazzi L, Attanasia A, Bussone G. Cervicogenic headache: a critical review of the current diagnostic criteria. Pain 1998; 78:1–5.

Leone M, D'Amico D, Moschiano F, Farinotti M, Fillipini G, Bussone G. Possible identification of cervicogenic headache among patients with migraine: an analysis of 374 headaches. Headache 1995, 35:461–4.

Lord S, Barnsley L, Wallis B, Bogduk N. Third occipital headache: a prevalence study. J Neurol Neurosurg Psychiat 1994, 57:1187–90.

Lord SM, Bogduk N. The cervical synovial joints as sources of post-traumatic headache. J Musculoskel Pain 1996, 4:81–94.

Poughias L, Kruszewski P, Inan L. Cervicogenic headache: a clinical review with special emphasis on therapy. Funct Neurol 1997; 12:305–17.

Sjaastad O, Fredriksen TA, Stolt-Nielsen A, Salvesen R, Jansen J, Pareja JA, Sjaastad O, Fredriksen TA. Cervicogenic headache: the importance of sticking to the criteria. Funct Neurol 2002; 17:35–6.

11.2.2 Retropharyngeal tendonitis

Eastwood JD, Hudgins PA, Malone D. Retropharyngeal effusion in acute calcific prevertebral tendonitis: diagnosis with CT and MR imaging. Am J Neuroradiol 1998; 19:1789–92.

Ekbom K, Torhall J, Annell K, Traff J. Magnetic resonance imaging in retropharyngeal tendonitis. Cephalalgia 1994; 14:266–9.

Pearce JM. Longus cervicis colli 'myositis' (syn: retropharyngeal tendonitis). J Neurol Neurosurg Psychiat 1996; 61:324–9.

Sarkozi J, Fam AG. Acute calcific retropharyngeal tendonitis: an unusual cause of neck pain. Arthritis Rheum 1984; 27:708–10.

11.2.3 Craniocervical dystonia

Csala B, Deuschl G. Craniocervical dystonia. Pragmatic general concept or nosologic entity? Nervenarzt 1994; 65:75–94.

Friedman J, Standaert DG. Dystonia and its disorders. Neurol Clin 2001; 19:681–705.

Göbel H, Deuschl G. Dauerkontraktionen kranialer oder zervikaler Muskeln. Münchener Medizinische Wochenschrift 1997; 139:456–8.

Göbel H, Heinze A, Heinze-Kuhn K, Austermann K. Botulinum toxin A in the treatment of headache syndromes and pericranial pain syndromes. Pain 2001; 91:195–9.

Markham CH. The dystonias. Curr Opin Neurol Neurosurg 1992; 5:301–7.

11.3 Headache attributed to disorder of eyes

Daroff RB. Ocular causes of headache. Headache 1998; 38:661–7.

Daum KM, Good G, Tijerina L. Symptoms in video display terminal operators and the presence of small refractive errors. J Am Optom Assoc 1988; 59:691–7.

Gerling J, Janknecht P, Kommerell G. Orbital pain in optic neuritis and anterior ischemic optic neuropathy. Neuro-Ophthalmology 1998; 19:93–99.

Göbel H, Martin TJ. Ocular disorders. In: Olesen J, Tfelt-Hansen P, Welch KMA. The Headaches. 2nd edition. Lippincott Williams & Wilkins, Philadelphia; 2000: pp. 899–904.

Gordon GE, Chronicle EP, Rolan P. Why do we still not know whether refractive error causes headaches? Towards a framework for evidence based practice. Ophthalmic Physiol Opt 2001; 21:45–50.

Lewis J, Fourman S. Subacute angle-closure glaucoma as a cause of headache in the presence of a white eye. Headache 1998; 38:684–6.

McCluskey PJ, Lightman S, Watson PG et al. Posterior scleritis. Clinical features, systemic associations, and outcome in a large series of patients. Ophthalmology 1999; 106:2380–6.

11.4 Headache attributed to disorder of ears; 11.5 Headache attributed to rhinosinusitis

Abu-Bakra M, Jones NS. Prevalence of nasal mucosal contact points in patients with facial pain compared with patients without facial pain. J Laryngol Otol 2001; 115:629–32.

Blumenthal HJ. Headache and sinus disease. Headache 2001; 41:883–8.

Boes CJ, Swanson JW, Dodick DW. Chronic paroxysmal hemicrania presenting as otalgia with a sensation of external acoustic meatus obstruction: two cases and a pathophysiologic hypothesis. Headache 1998; 38:787–91.

Close LG, Aviv J. Headaches and disease of the nose and paranasal sinuses. Semin Neurol 1997; 17:351–4.

De Vuyst D, De Schepper AM, Parizel PM. Chronic cocaine abuse. JBR-BTR 2001; 84:60.

Göbel H, Baloh RW. Disorders of ear, nose, and sinus. In: Olesen J, Tfelt-Hansen P, Welch KMA. The Headaches. 2nd edition. Lippincott Williams & Wilkins, Philadelphia; 2000: pp. 905–12.

Kenny TJ, Duncavage J, Bracikowski J, Yildirim A, Murray JJ, Tanner SB. Prospective analysis of sinus symptoms and correlation with paranasal computed tomography scan. Otolaryngol Head Neck Surg 2001; 125:40–3.

Lam DK, Lawrence HP, Tenenbaum HC. Aural symptoms in temporomandibular disorder patients attending a craniofacial pain unit. J Orofac Pain 2001; 15:146–57.

Lanza DC, Kennedy DW. Adult rhinosinusitis defined. Report of the Rhinosinusitis Task Force Committee of the American Academy of Otolaryngology Head and Neck Surgery. Otolaryngol Head Neck Surg 1997; 117:S1–S7.

Levine HL. Patients with headache and visual disturbance: a differentiation between migraine and sinus headache. Arch Otolaryngol Head Neck Surg 2000: 126:234–35.

Murphy E, Merrill RL. Non-odontogenic toothache. J Ir Dent Assoc 2001; 47(2):46–58.

Pinto A, De Rossi SS, McQuone S, Sollecito TP. Nasal mucosal headache presenting as orofacial pain: a review of the literature and a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2001; 92:180–3.

Sandstrom M, Wilen J, Oftedal G, Hansson Mild K. Mobile phone use and subjective symptoms. Comparison of symptoms experienced by users of analogue and digital mobile phones. Occup Med (Lond) 2001; 51:25–35.

Seiden AM, Martin VT. Headache and the frontal sinus. Otolaryngol Clin North Am 2001; 34:227–41.

Sydbom A, Blomberg A, Parnia S, Stenfors N, Sandstrom T, Dahlen SE. Health effects of diesel exhaust emissions. Eur Respir J 2001; 17:733–46.

Tosun F, Gerek M, Ozkaptan Y. Nasal surgery for contact point headaches. Headache 2000; 40:237–40.

West B, Jones NS. Endoscopy-negative, computed tomography-negative facial pain in a nasal clinic. Laryngoscope 2001; 111(4 Pt 1):581–6.

11.6 Headache attributed to disorder of teeth, jaws or related structures

Allen DT, Voytovich MC, Allen JC. Painful chewing and blindness: signs and symptoms of temporal arteritis. J Am Dent Assoc. 2000; 131:1738–41.

Ciancaglini R, Radaelli G. The relationship between headache and symptoms of temporomandibular disorder in the general population. J Dent 2001; 29:93–8.

Egermark I, Carlsson GE, Magnusson T. A 20-year longitudinal study of subjective symptoms of temporomandibular disorders from childhood to adulthood. Acta Odontol Scand 2001; 59:40–8.

Epstein JB, Caldwell J, Black G. The utility of panoramic imaging of the temporomandibular joint in patients with temporomandibular disorders. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2001; 92:236–9.

Henrikson T, Ekberg EC, Nilner M. Symptoms and signs of temporomandibular disorders in girls with normal occlusion and Class II malocclusion. Acta Odontol Scand 1997; 55:229–35.

Ivanhoe CB, Lai JM, Francisco GE. Bruxism after brain injury: successful treatment with botulinum toxin-A. Arch Phys Med Rehabil 1997; 78:1272–3.

Kirveskari P. Prediction of demand for treatment of temporomandibular disorders. J Oral Rehabil 2001; 28:572–5.

Magnusson T, Egermark I, Carlsson GE. A longitudinal epidemiologic study of signs and symptoms of temporomandibular disorders from 15 to 35 years of age. J Orofac Pain 2000; 14:310–9.

Marcusson A, List T, Paulin G, Dworkin S. Temporomandibular disorders in adults with repaired cleft lip and palate: a comparison with controls. EOS 2001; 23:193–204.

Sonnesen L, Bakke M, Solow B. Malocclusion traits and symptoms and signs of temporomandibular disorders in children with severe malocclusion. Eur J Orthod 1998; 20:543–59.

11.7 Temporomandibular joint disorder

List T, Wahlund K, Larsson B. Psychosocial functioning and dental factors in adolescents with temporomandibular disorders: a case-control study. J Orofac Pain 2001; 15:218–27.

Ciancaglini R, Radaelli G. The relationship between headache and symptoms of temporomandibular disorder in the general population. J Dent 2001; 29:93–8.

Molina OF, dos Santos Junior J, Nelson SJ, Nowlin T. Profile of TMD and Bruxer compared to TMD and nonbruxer patients regarding chief complaint, previous consultations, modes of therapy, and chronicity. Cranio 2000; 18:205–19.

Ogus H. Degenerative disease of the temporomandibular joint and pain-dysfunction syndrome. J Roy Soc Med 1978; 71:748–54.

Jacome D. Primary yawning headache. Cephalalgia 2001; 21:697–9.

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[1] 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.

Bibliography and reference

Curioso EP, Young WB, Shecter AL, Kaiser R. Psychiatric comorbidity predicts outcome in chronic daily headache patients. Neurology 1999; 52 (Suppl 2): A471.

Canestri P, Galli F, Guidetti V, Tomaciello A. Chronic Daily Headache in children and adolescents: a two years follow-up. Cephalalgia 2001; 21:288.

Guidetti V, Galli F, Fabrizi P, Napoli L, Giannantoni AS, Bruni O, Trillo S. Headache and psychiatric comorbidity: clinical aspects and outcome in an 8-year follow-up study. Cephalalgia 1998; 18:455–62.

Lake A. Behavioral and nonpharmacologic treatments of headache. Med Clin North Am 2001; 85:1055–75.

Marazzitti D, Toni C, Pedri S, Bonucelli U et al. Prevalence of headache syndromes in panic disorder. Int Clin Psychopharmacol 1999; 14:247–251.

Mitsikostas DD, Thomas AM. Comorbidity of headache and depressive disorders. Cephalalgia 1999; 19:211–217.

Pakalnis A, Greenberg G, Drake ME, Paolich J. Pediatric migraine prophylaxis with divalproex. J Child Neurol 2001; 16:731–4.

Radat F, Sakh D, Lutz G, el Amrani M, Ferreri M, Bousser MG. Psychiatric comorbidity is related to headache induced by chronic substance use in migraineurs. Headache 1999; 39:477–80.

Radat F, Psychopathology and headache. Rev Neurol 2000; 156 (Suppl 4):4S62–7.

Yutzy S. Somatoform disorders. In: Tasman A, Kay J, Lieberman JA. Psychiatry, 2nd ed. Chichester: John Wiley and Sons 2003: pp. 1419–20.

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
- 13.2.2 Symptomatic glossopharyngeal neuralgia

13.3 Nervus intermedius neuralgia
13.4 Superior laryngeal neuralgia
13.5 Nasociliary neuralgia
13.6 Supraorbital neuralgia
13.7 Other terminal branch neuralgias
13.8 Occipital neuralgia
13.9 Neck-tongue syndrome
13.10 External compression headache
13.11 Cold-stimulus headache
- 13.11.1 Headache attributed to external application of a cold stimulus
- 13.11.2 Headache attributed to ingestion or inhalation of a cold stimulus

13.12 Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions
13.13 Optic neuritis
13.14 Ocular diabetic neuropathy
13.15 Head or facial pain attributed to herpes zoster
- 13.15.1 Head or facial pain attributed to acute herpes zoster
- 13.15.2 Post-herpetic neuralgia

13.16 Tolosa-Hunt syndrome
13.17 Ophthalmoplegic 'migraine'
13.18 Central causes of facial pain
- 13.18.1 Anaesthesia dolorosa
- 13.18.2 Central post-stroke pain
- 13.18.3 Facial pain attributed to multiple sclerosis
- 13.18.4 Persistent idiopathic facial pain
- 13.18.5 Burning mouth syndrome

13.19 Other cranial neuralgia or other centrally mediated facial pain

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[1]

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[1]

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[1]

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 tempera-

ture 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[1] 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[1]
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[1]
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[1]
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 ≥3 months after the onset of 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 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[1]

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 and 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[1], 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.

Bibliography and reference

13.1 Trigeminal neuralgia

Barker FG II, Jannetta PJ, Bissonette DJ, Larkins MV, Jho HD. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Engl J Med 1997; 334:1077–83.

Taha JM, Tew JM Jr. Treatment of trigeminal neuralgia by percutaneous radiofrequency rhizotomy. Neurosurgery Clinics of North America 1997; 8:31–9.

Terrence CF, Jensen TS. Trigeminal neuralgia and other facial neuralgias. In: Olesen J, Tfelt-Hansen P, Welch KMA eds The Headaches, 2nd ed. Philadelphia, Lippincott Williams & Wilkins 2000: 929–38.

Zakrzewska JM. Trigeminal neuralgia. In: Zakrzewska JM, Harrison SD, eds. Assessment and management of orofacial pain. Pain Research and Clinical Management, Amsterdam, Elsevier 2002; 14:263–366.

13.2 Glossopharyngeal neuralgia

Ekbom KA, Westerberg CE. Carbamazepine in glossopharyngeal neuralgia. Arch Neurol 1966; 14:595–6.

Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. J Neurosurg 1977; 47:316–20.

Minagor A, Sheremata WA. Glossopharyngeal neuralgia and MS. Neurology 2000; 54:1368–70.

Rushton JG, Stevens JC, Miller RH. Glossopharyngeal (vagoglossopharyngeal) neuralgia. A study of 217 cases. Arch Neurol 1981; 38:201–5.

13.3 Nervus intermedius neuralgia

Bruyn GW. Nervus intermedius neuralgia (Hunt). In: Rose FC ed Headache. Handbook of Clinical Neurology. Amsterdam: Elsevier 1986; 4(48):487–94.

13.4 Superior laryngeal neuralgia

Bruyn GW. Superior laryngeal neuralgia. In: Rose FC ed. Headache. Handbook of Clinical Neurology. Amsterdam: Elsevier 1986; 4(48):495–500.

13.5 Nasociliary neuralgia

Bruyn GW. Charlin's neuralgia. In: Rose FC ed. Headache. Handbook of Clinical Neurology, Amsterdam: Elsevier 1986; 4(48):483–6.

13.6 Supraorbital neuralgia

Sjaastad O, Stolt-Nielsen A, Pareja JA, Vincent M. Supraorbital neuralgia. The clinical manifestation and a possible therapeutic approach. Headache 1999; 39:204–12.

13.7 Infraorbital neuralgia

de Vries N, Smelt WL. Local anaesthetic block of posttraumatic neuralgia of the infraorbital nerve. Rhinology 1990; 28:103–106.

13.8 Occipital neuralgia

Bogduk N. Greater occipital neuralgia. In: Long DM ed. Current Therapy in Neurological Surgery. Toronto, Philadelphia: BC Decker Inc 1985: 175–180.

Hammond SR, Danta A. Occipital neuralgia. Clin Exp Neurol 1978; 15:258–270.

13.9 Neck-tongue syndrome

Bertoft ES, Westerberg CE. Further observations on the neck-tongue syndrome. Cephalalgia 1985; 5(Suppl 3):312–3.

Bogduk N. An anatomical explanation for the neck-tongue syndrome. J Neurol Neurosurg Psychiat 1981; 44:202–8.

Lance JW, Anthony M. Neck-tongue syndrome on sudden turning of the head. J Neurol Neurosurg Psychiat 1980; 43:97–101.

13.10 External compression headache

Pestronk A, Pestronk S. Goggle migraine. N Engl J Med 1983; 308:226.

13.11 Cold stimulus headache

Drummond PD, Lance JW. Neurovascular disturbances in headache patients. Clin Exp Neurol 1984; 20:93–9.

Odell-Smith R. Ice cream headache. In: Vinken PJ, Bruyn GW eds Handbook of Clinical Neurology. Amsterdam: Elsevier 1968; 5:188–91.

Raskin NH. Ice cream, ice-pick and chemical headaches. In: Rose FC ed Headache. Handbook of Clinical Neurology. Amsterdam: Elsevier 1986; 4(48):441–8.

Raskin NH, Knittle SC. Ice cream headache and orthostatic symptoms in patients with migraine. Headache 1976; 16:222–5.

Wolf S, Hardy JD. Studies on pain. Observations on pain due to local cooling and on factors involved in the 'cold pressor' effect. J Clin Invest 1941; 20:521–33.

13.12 Constant pain caused by compression, irritation or distortion of cranial nerves

Adams RD, Victor M, Ropper AH. Principles of Neurology 6th ed. New York: McGraw-Hill 1997: pp. 187–193, 1370–85.

Capobianco DJ. Facial pain as a symptom of nonmetastatic lung cancer. Headache 1995; 35:581–5.

Dalessio DJ ed. Wolff's Headache and other head pain. 6th edn New York: Oxford University Press 1993:345–64.

Mokri B. Raeder's paratrigeminal syndrome. Arch Neurol 1982; 39:395–99.

Schoenen J, Broux R, Moonen G. Unilateral facial pain as the first symptom of lung cancer: are there diagnostic clues? Cephalalgia 1992; 12:178–9.

13.13 Optic neuritis

Adams RD, Victor M, Ropper AH. Principles of Neurology 6th edn. New York, McGraw-Hill 1997: pp. 910–11.

Kaufman DI, Trobe JD, Eggenberger ER, Whitaker JN. Practice parameter: the role of corticosteroids in the management of acute monosymptomatic optic neuritis. Neurology 2000; 54:2039–44.

Shults WT. Diseases of the visual pathways I. In Swash M, Oxbury J, eds. Clinical Neurology. Edinburgh, Churchill Livingstone 1991:410–13.

13.14 Ocular diabetic neuropathy

Kennard C. Disorders of eye movements I. In Swash M, Oxbury J, eds. Clinical Neurology. Edinburgh. Churchill Livingstone 1991: pp. 446–7.

13.15 Head or facial pain attributed to herpes zoster

Bowsher D. The management of postherpetic neuralgia. Postgrad Med J 1997; 73:623–9.

Dworkin RH, Portenoy RK. Pain and its persistence in herpes zoster. Pain 1996; 67:241–52.

Ragozzino MW, Melton LJ, Kerland LT, Chu CP, Perry HO. Population-based study of Herpes Zoster and its sequelae. Medicine 1982; 61:310–6.

13.16 Tolosa-Hunt syndrome

de Arcaya AA, Cerezal L, Canga A, Polo JM, Berciano J, Pascual J. Neuroimaging diagnosis of Tolosa-Hunt syndrome. MRI contribution. Headache 1999; 39:321–5.

Forderreuther S, Straube A. The criteria of the International Headache Society for Tolosa-Hunt syndrome need to be revised. J Neurol 1999; 246:371–7.

Goadsby PJ, Lance JW. Clinicopathological correlation in a case of painful ophthalmoplegia: Tolosa-Hunt syndrome. J Neurol Neurosurg Psychiat 1989; 52:1290–3.

Hannerz J. Pain characteristics of painful ophthalmoplegia (the Tolosa-Hunt syndrome). Cephalalgia 1985; 5:103–6.

Hunt WE. Tolosa-Hunt syndrome: one cause of painful ophthalmoplegia. J Neurosurg 1976; 44:544–9.

Tolosa E. Periarteritic lesions of the carotid siphon with the clinical features of a carotid infraclinoidal aneurysm. J Neurol Neurosurg Psychiatry 1954; 17:300–2.

13.17 Ophthalmoplegic 'migraine'

Lance JW, Zagami AS. Ophthalmoplegic migraine: a recurrent demyelinating neuropathy? Cephalalgia 2001; 21:84–9.

Mark AS, Casselman J, Brown D, Sanchez J, Kolsky M, Larsen TC III, Lavin P, Ferraraccio B. Ophthalmoplegic migraine: reversible enhancement and thickening of the cisternal segment of the oculomotor nerve on contrast-enhanced MR images. Am J Neuroradiol 1998; 19:1887–91.

13.18.1 Anaesthesia dolorosa

Illingworth R. Trigeminal neuralgia: surgical aspects. In: Rose RD ed Headache. Handbook of Clinical Neurology. Amsterdam: Elsevier 1986; 4 (48):449–58.

Pagni CA. Central and painful anaesthesia. Pathophysiology and treatment of sensory deprivation syndromes due to

central and peripheral nervous system lesions. Progr Neurol Surg 1977; 2:132–257.

13.18.2 Central post-stroke pain

Bowsher D. Central pain: clinical and physiological characteristics. J Neurol Neurosurg Psychiat 1996; 61:62–9.

Bowsher D, Leijon G, Thuomas KA. Central poststroke pain. Correlation of MRI with clinical pain characteristics and sensory abnormalities. Neurology 1998; 51:1352–8.

Fitzek S, Baumgartner U, Fitzek C et al. Mechanisms and Predictors of chronic facial pain in lateral medullary infarction. Ann Neurol 2001; 49:493–500.

13.18.3 Facial pain attributed to multiple sclerosis

Jensen TS, Rasmussen P, Reske-Nielsen E. Association of trigeminal neuralgia with multiple sclerosis: clinical pathological features. Acta Neurol Scand 1982; 65:182–9.

13.18.4 Persistent idiopathic facial pain

Boivie J, Casey KL. Central pain in the face and head. In Olesen J, Tfelt-Hansen P, Welch KMA eds. The Headaches, 2nd ed. Philadelphia, Lippincott, Williams & Wilkins 2000:939–45.

Gouda JJ, Brown JA. Atypical facial pain and other pain syndromes. Differential diagnosis and treatment. Neurosurgery Clinics of North America 1997; 1:87–99.

Harrison SD. Atypical facial pain and atypical odontalgia in Zakrzewska JM, Harrison SD, eds. Assessment and management of orofacial pain. Pain Research and Clinical Management, Amsterdam: Elsevier 2002; 14:251–62.

13.18.5 Burning mouth syndrome

Zakrzewska J. Burning mouth. In Zakrzewska JM, Harrison SD, eds. Assessment and management of orofacial pain. Pain Research and Clinical Management. Amsterdam. Elsevier 2002; 14:367–80.

13.19 Other cranial neuralgia or other centrally mediated facial pain

Montalbetti L, Ferrandi D, Pergami P, Savoldi F. Elongated styloid process and Eagle's syndrome. Cephalalgia 1995; 15:80–93.

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 or had 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 entitities 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)
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 ± 2 (*ie*, days –2 to +3)[1] of menstruation[2] 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 ± 2 (*ie*, days –2 to +3)[1] of menstruation[2] 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[1]

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 without aura* and A1.1.2 *Menstrually-related migraine without aura* 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 ≥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[1]
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.

Bibliography

MacGregor EA, Chia H, Vohrah RC, Wilkinson M. Migraine and menstruation: a pilot study. Cephalalgia 1990; 10:305–10.

MacGregor EA. 'Menstrual' migraine: towards a definition. Cephalalgia 1996; 16:11–21.
Somerville B.The role of estradiol withdrawal in the etiology of menstrual migraine. Neurology 1972; 22:355–65.

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 another disorder[1,2]

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 8.2.7 *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 8.2.7 *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 subtype 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 ≥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 1 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 ≥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

Bibliography

Goadsby PJ, Matharu MS, Boes CJ. SUNCT syndrome or trigeminal neuralgia with lacrimation. Cephalalgia 2001; 21:82–83.

Sjaastad O, Kruszewski P. Trigeminal neuralgia and 'SUNCT' syndrome: similarities and differences in the clinical picture. An overview. Functional Neurology 1992; 7:103–107.

Sjaastad O, Pareja JA, Zukerman E, Jansen J, Kruszewski P. Trigeminal neuralgia. Clinical manifestations of first division involvement. Headache 1997; 37:346–357.

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 *Headache or facial or neck pain attributed to 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

Bibliography

Biousse V, Bousser MG. The myth of carotidynia. Neurology 1994; 44:993–5.

Burton BS, Syms MJ, Petermann GW, Burgess LPA. MR imaging of patients with carotidynia. AJNR 2000; 21:766–9.

Fay T. Atypical neuralgia. Arch Neurol Psychiat 1927; 18:309–15.

Forwith KD, Tami TA. Carotidynia: symptom or diagnosis? Curr Opin Otolaryngol Head Neck Surg 1999; 7:150–4.

Hill LM, Hastings G. Carotidynia: a pain syndrome. J Fam Pract 1994; 39:71–5.

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

Bibliography and references

A7.9.1 Post-radiosurgery headache

Kondziolka D, Lundsford LD, Flickinger JC. Gamma knife stereotactic radiosurgery for cerebral vascular malformations. In: Alexander E III, Loeffler JS, Lundsford LD eds. Stereotactic Radiosurgery. New York: McGraw Hill Inc 1993: pp. 136–145.

Lundsford LD, Flickinger JC, Coffee RJ. Stereotactic gamma knife radiosurgery. Initial North American experience in 207 patients. Arch Neurol 1990; 47:169–75.

Rozen TD, Swanson JW. Post-gamma knife headache: A new headache syndrome? Headache 1997; 37:180–3.

A7.9.2 Post-electroconvulsive therapy (ECT) headache

DeBattista C, Mueller K. Sumatriptan prophylaxis for post-electroconvulsive therapy headaches. Headache 1995; 35:502–3.

Folkerts H. Migraine after electroconvulsive therapy. Convulsive Therapy 1995; 11:212–5.

Ghoname EA, Craig WF, White PF. The use of percutaneous electrical nerve stimulation (PENS) for treating ECT-induced headaches. Headache 1999; 39:502–5.

Hawken ER, Delva NJ, Lawson JS. Successful use of propranolol in migraine associated with electroconvulsive therapy. Headache 2001; 41:92–6.

Markowitz JS, Kellner CH, DeVane CL, Beale MD, Folk J, Burns C, Liston HL. Intranasal sumatriptan in post-ECT headache: results of an open-label trial. Journal of ECT 2001; 17:280–3.

Oms A, Miro E, Rojo JE. Sumatriptan was effective in electroconvulsive therapy (ECT) headache. Anesthesiology 1998; 89:1291–2.

Weiner SJ, Ward TN, Ravaris CL. Headache and electroconvulsive therapy. Headache 1994; 34:155–9.

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 lists medications reported to cause headache during therapeutic use.

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[1] 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 trema-

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

todes (*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 be proposed only 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

Westerink MA, Amsterdam D, Petell RJ, Stram MN, Apricella MA. Septicemia due to DF-2. Cause of a false-positive cryptococcal latex agglutination result. Am J Med 1987; 83:155–8.

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, 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 homoeostasis 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[1]
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-craniocervical disorder headache

Diagnostic criteria:
A. Headache, no typical characteristics known, fulfilling criteria C and D
B. A craniocervical disorder has been present but has been effectively treated or has remitted spontaneously
C. Headache has been attributed to the craniocervical disorder
D. Headache persists for >3 months after effective treatment or spontaneous remission of the craniocervical 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 ≥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. persistent and excessive fear or reluctance 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

Pareja JA, Caminero AB, Serra J, Barriga FJ, Dobato JL, Barón M, Vela L, Sánchez del Río M. Numular headache: a coin-shaped cephalgia. Neurology 2002; 58:1678–9.

Definition of Terms

Accompanying symptoms: Symptoms that typically accompany rather than precede or follow headache. In migraine, for example, the most frequent are nausea, vomiting, photophobia and phonophobia; osmophobia, diarrhoea and other symptoms occur more rarely.

Anorexia: Lack of appetite and dislike for food to a mild degree.

Attack of headache (or pain): Headache (or pain) that builds up, remains at a certain level for minutes to 72 hours, then wanes until it is gone completely.

Aura: Early symptoms of an attack of migraine with aura, being the manifestations of focal cerebral dysfunction. The aura typically lasts 20–30 minutes and precedes the headache. See also: *focal symptoms*, *prodrome, premonitory symptoms* and *warning 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, 2nd 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*).