Chapter 2

Classification of Headache

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Undoubtedly, most physicians consider disease classification to be boring, difficult to learn, and clinically not very useful. In practice, most physicians do not memorize diagnostic criteria or take the time to look the criteria up except in special cases. However, clinical diagnosis always depends on a classification system and we all use formal or informal diagnostic criteria, often without knowing it.

A new classification system often finds its initial application in research. But, after it has been used for some years, all new research results in the particular field have been gathered using that system. To apply that information in practice, physicians must diagnose their patients in ways that are compatible with the new system. Through this process, the evidence-based medicine links a new classification system to clinical practice. Classification systems also have more direct effects. For example, the explicit diagnostic criteria for migraine without aura (7) request knowledge only about duration of attack, severity of pain, unilaterality, pulsating or no pulsating quality, and aggravation or no aggravation of pain by physical activity as well as knowledge about the associated symptoms of nausea, vomiting, and photo- and phonophobia. This serves as a clear guideline to the clinical interview and makes it unnecessary in most cases to ask many questions that physicians used in the past to characterize a headache.

The headache field has enjoyed a systematic hierarchical classification system and associated explicit (previously called *operational*), diagnostic criteria for more than 15 years after the publication of the first edition at the *Classification of the International Headache Society* (IHS) (ICHD-1) in 1988 (8). Building on ICHD-1, the second edition of the *International Classification of Headache Disorders* (ICHD-2) has recently been published (7). Like its predecessor, it will have a dominating influence on research and clinical practice for at least the next decade (Figs. 2-1 and 2-2).

GENERAL PRINCIPLES OF DISEASE CLASSIFICATION AND DIAGNOSTIC CRITERIA

To classify disease means to decide how many different entities that should be recognized and to order them in a logical system. There are two diametrically opposed forces in classification: the tendency to lump and the tendency to split. Lumping headache disorders leads in the extreme to one category: headache. To physicians interested in headache this sounds as a highly theoretical option, but it is the way many general physicians and the general population classify headache disorders. If they are a little bit more sophisticated, they recognize the difference between migraine and other headaches. When professionals talk about headache disorders, laypersons often believe that it does not include migraine and it is often useful for politicians and other decision makers to talk about "migraine and other headaches." For extreme splitters, it is possible to diagnose an almost endless variety of headaches. Taken to the extreme this leads to the attitude that "There are no diseases, only patients." Each patient has special characteristics and therefore merits his or her own category of subdiagnosis. Successful classifications balance these opposing tendencies by grouping similar patients together in ways that are useful for clinical practice. These similarities can be based on epidemiology, symptom profile, disease mechanism, and sometimes treatment response. Even sophisticated studies of brain blood flow, biochemistry, or genetics can be taken into account when constructing the classification system. Diagnostic criteria should, on the other hand, include only parameters that are available to the physician when diagnosing patients.

It is often suggested that there should be two different classifications: One for research, which can be detailed, and another for clinical practice, which should be simple and very easy to apply. It is absolutely hopeless, however, to have different classifications for research and clinical

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FIGURE 2-1. The Chairman of the First and Second Headache Classification Committee together with the secretary Shiela M. Westh, who did all the practical work on the first edition of the International Headache Classification. This photo was taken in 1988.

practice because research results obtained with one kind of classification would be difficult to apply to patients diagnosed according to another system. The ICHD-1 and -2 use hierarchical classification to solve this problem (7,8). Patients can be diagnosed in groups and subgroups with various levels of diagnostic refinement. With such a system, patients can be diagnosed according to the first or second digit in general practice, and special clinics and researchers may diagnose to third or fourth digit. Ideally, a classification system should be based on etiology, but very

FIGURE 2-2. Second International Headache Classification Committee, at its last meeting in Copenhagen, March 2003. From left to right: Top row: Peter J. Goadsby, Guiseppe Nappi, Giorgio Sandrini, Tim Steiner. Middle row: Peer Tfelt-Hansen, James W. Lance, Miguel J.A. Lainez, Michael First. Front row: Hartmut Göbel, Fumihiko Sakai, Rigmor Jensen, Jes Olesen, Marie G. Bousser, Hans Christoph Diener. Missing from this picture are David Dodick, Richard B. Lipton, Jean Schoenen, and Stephen D. Silberstein, who were unable to attend the last meeting, but all did an equally good job to those present. Of these Peer Tfelt-Hansen, Giorgio Sandrini, and Rigmor Jensen were very active in subcommittees, but not members of the main committee.

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often the etiology is not known. The classification system then has to use clinical features and the results of laboratory tests, and thus is based on phenomenology. The latter applies to the primary headaches; etiology is used to classify the secondary headaches in ICHD-2.

THE HISTORY OF HEADACHE CLASSIFICATION

The first attempt at creating a headache classification was made by an ad hoc committee of the National Institutes of Health in America. It published a paper entitled "Classification of Headache" in 1962 (1). This classification became quite widely used in North America and parts of Europe, but never gained true international acceptance or broad use. It recognized only a few headache disorders and its so-called definitions of different headache disorders were wide open for individual interpretation. Migraine, for example, was defined as "recurrent attacks of headache, widely varied in intensity, frequency, and duration. The attacks are commonly unilateral in onset; are usually associated with anorexia, and sometimes with nausea and vomiting; and some are proceeded by, or associated with, conspicuous sensory, motor, and mood disturbances; and are often familial" (1, p. XX). Some attempts were made to operationalize the diagnostic criteria for migraine (16,19). These criteria were never internationally accepted, but formed a valuable basis for the subsequent classification work. In 1985, the IHS formed a headache classification committee, which in 1988 published the first international classification of headache disorders (ICHD-1) (8). It contained operational (now called *explicit*) diagnostic criteria for all headache disorders. The system was endorsed by all national headache societies that were members of the IHS and by the World Federation of Neurology. The classification was translated into more than 20 different languages and has been used throughout the world as the only internationally accepted headache classification system. The World Health Organization (WHO) accepted the major principles of ICHD-1. After 15 years of service the ICHD-1 has now been replaced by ICHD-2, published in January 2004 following more than 4 years of intense preparations by a new international headache classification committee. The classification is available on the IHS web site (9). A slide kit is also available for downloading. A short version of the classification has been developed and is available on the IHS web site and as a printed pocket folder (10).

ICHD-2 Classification and Terminology

Like it predecessor, ICHD-2 is hierarchical using up to four digits to code for all varieties of headache disorders. These are now organized in 14 major groups. Groups 1 to 4 cover the primary headaches (Table 2-1), groups 4 to 12 the secondary headaches, group 13 the cranial neuralgias and facial pain, and group 14 other headaches, cranial neuralgias, or primary facial pain (Table 2-2). New groups in ICHD-2 are group 10 headache, attributed to disorder of homeostasis, and group 13 headache, attributed to psychiatric disorder. Also new is the subdivision of group 14 into headache not elsewhere classified and headache where insufficient information is available to classify. New entities have been included within most groups but particularly so in Chapter 4, which was previously called miscellaneous primary headaches now "Other Primary Headaches." Major reorganization has also taken place within the chapter on headache attributed to substance use, especially the clear recognition of medication overuse headache. An appendix has been added that includes research criteria for new types of headache not yet sufficiently validated.

The taxonomy of headache disorders was quite markedly changed in ICHD-1, notably by the introduction of the *migraine with aura* and *migraine without aura* to replace *common migraine* and *classical migraine,* respectively, as well as *tension-type headache* replacing *tension headache or muscle contraction headache.* This change of taxonomy has largely been successful. Therefore, few changes have been made to the taxonomy of ICHD-2. Previously used terms are presented whenever such terms have existed, just like in ICHD-1.

ICHD-2 Diagnostic Criteria

The obvious difficulty in constructing diagnostic criteria for primary headache disorders is the lack of a definitive gold standard for diagnosis. For the primary headaches, there is no clear biochemical or diagnostic marker, no valid external diagnostic touchstone. ICHD-1 criteria were therefore constructed using clinical features and easily available results of laboratory investigations. ICHD-2 has continued this tradition. The diagnostic criteria previously called *operational* are now called *explicit* diagnostic criteria. This means that terms such as sometimes, usually, and often are avoided and instead numerical figures are given. Sometimes criteria are monothetic, requiring the presence or absence of a single characteristic. At other times the criteria are polythetic, requiring for example two out of four characteristics. These kinds of criteria allow use of characteristics that occur in, for example, 50% of the patients, such as the pain criteria of migraine without aura (see Table 2-2). The basic system from DSM-4 (3) used in ICHD-1 is still used in ICHD-2. Thus, the criteria contain several letter headings: A, B, and C, and each letter heading has to be fulfilled to come to a diagnosis.

ICHD-2 and WHO Classifications

The *International Classification of Diseases,* 9th edition (ICD-9) contained only a few headache entities and very important syndromes such as tension-type headache

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TABLE 2-1 Classification of Headache (First Level, With Selected Disorders at Second and Third Levels)

ICH D-II Code	*ICD-10NA Code*	*Diagnosis*
1.	**[G43]**	**Migraine**
1.1	[G43.0]	Migraine without aura
1.2	[G43.1]	Migraine with aura
1.5.1	[G43.3]	Chronic migraine
2.	**[G44.2]**	**Tension-type headache (TTH)**
2.1	[G44.2]	Infrequent episodic tension-type headache
2.2	[G44.2]	Frequent episodic tension-type headache
2.3	[G44.2]	Chronic tension-type headache
3.	**[G44.0]**	**Cluster headache and other trigeminal autonomic cephalalgias**
3.1	[G44.0]	Cluster headache
3.2	[G44.03]	Paroxysmal hemicrania
4.	**[G44.80]**	**Other primary headaches**
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
5.2	[G44.3]	Chronic post-traumatic headache
5.3	[G44.841]	Acute headache attributed to whiplash injury [S13.4]
5.4	[G44.841]	Chronic headache attributed to whiplash injury [S13.4]
6.	**[G44.81]**	**Headache attributed to cranial or cervical vascular disorder**
6.2.2	[G44.810]	Headache attributed to subarachnoid hemorrhage (SAH) [I60]
6.4.1	[G44.812]	Headache attributed to giant cell arteritis (GCA) [M31.6]
7.	**[G44.82]**	**Headache attributed to non-vascular intracranial disorder**
7.1.1	[G44.820]	Headache attributed to idiopathic intracranial hypertension (IIH) [G93.2]
7.2.1	[G44.820]	Post-dural puncture headache [G97.0]
7.4	[G44.822]	Headache attributed to intracranial neoplasm [C00-D48]
7.6.2	[G44.82]	Post-seizure (post-ictal) headache [G40.x or G41.x to specify seizure type]
8.	**[G44.4 or G44.83]**	**Headache attributed to a substance or its withdrawal**
8.2	[G44.41 or G44.83]	Medication-overuse headache (MOH)
9.		**Headache attributed to infection**
9.1.1	[G44.821]	Headache attributed to bacterial meningitis [G00.9]
9.4.1	[G44.821]	Chronic post-bacterial meningitis headache [G00.9]
10.	**[G44.882]**	**Headache attributed to disorder of homeostasis**
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**
11.2.1	[G44.841]	Cervicogenic headache [M99]
12.	**[R51]**	**Headache attributed to psychiatric disorder**
12.1	[R51]	Headache attributed to somatization disorder [F45.0]
12.2	[R51]	Headache attributed to psychotic disorder [code to specify etiology]
13.	**[G44.847, G44.848 or G44.85]**	**Cranial neuralgias and central causes of facial pain**
13.1	[G44.847]	Trigeminal neuralgia
14.	**[R51]**	**Other headache, cranial neuralgia, central or primary facial pain**
14.1	[R51]	Headache not elsewhere classified
14.2	[R51]	Headache unspecified

were not included under neurology. However, the much more detailed classification and the diagnostic criteria of ICHD-1 were largely accepted by the WHO and included in ICD-10 (21). This meant that all primary headache syndromes were now grouped under neurology. Unfortunately, the United States has not yet adopted the ICD-10, which has greatly hampered a reasonable reimbursement of headache experts and therefore also the development of services to headache patients. From ICD-10 a neurologic adaptation (ICD-10 NA) was developed (22), which is more detailed and includes most of the headache disorders classified in ICHD-1. A further document, "ICD-10

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TABLE 2-2 Diagnostic Criteria for Migraine Without Aura

Diagnostic criteria
A. At least 5 attacks[1] fulfilling B–D.
B. Headache attacks lasting 4–72 hours[2,3] and occurring <15 days per month[4] (untreated or unsuccessfully treated).
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 (i.e., 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]

[1]Differentiating between migraine without aura and episodic tension-type headache may be difficult. Therefore at least five attacks are required. Individuals who otherwise meet criteria for migraine without aura but have fewer than five attacks should be coded 1.6.
[2]If the patient falls asleep during migraine and wakes up without it, duration of the attack is until time of awakening.
[3]In children, attacks may last 1–72 hours. (The evidence for untreated durations less than 2 hours in children should be corroborated by prospective diary studies).
[4]If attack frequency ≥15 days per month and if there is no medication overuse code 1.1 and 1.5.1 chronic migraine.
[5]Migraine headache is often bilateral in young children; an adult pattern of unilateral pain often 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 at rest or with movement.
[8]In young children, photophobia and phonophobia may be inferred from behavior.
[9]History, physical, and neurologic examinations do not suggest one of the disorders listed in groups 5–12, or history and/or physical and/or neurologic examinations do suggest such disorder, but it is ruled out by appropriate investigations, or such disorder is present, but migraine attacks do not occur for the first time in close temporal relation to the disorder.

guide for headaches," was published to provide a cross-way between ICD-10 (11), ICD NA, and ICHD-1. In ICHD-2, it was decided to include both the IHS and ICD-10 NA code numbers. The latter system is less detailed. Different ICHD diagnoses may therefore often receive the same ICD-10 NA diagnosis. Table 2-1 gives ICHD-2 codes and diagnoses as well as ICD-10 NA codes.

IMPORTANT NEW FEATURES OF ICHD-2

The Primary Headaches

The criteria for migraine without aura remain unchanged, except that childhood migraine may sometimes last as little as 1 hour. The criteria for migraine with aura have been rather extensively changed to allow for the coding of migraine aura with migraine headache, with nonmigraine headache and without headache. Although criteria are now more explicit and should have a higher interobserver reliability, they diagnose the same patients as the criteria under ICHD-1. Thus, the new criteria have a high sensitivity and specificity when tested in a large material of patients diagnosed with ICHD-1 criteria (4). The childhood syndromes that are precursors of migraine have been extensively revised, as has the section on complications of migraine. Most important is the addition of chronic migraine. Chronic migraine is diagnosed in patients having migraine on 15 days per month or more in the absence of medication overuse. If medication overuse is present, only probable chronic migraine can be diagnosed. The diagnosis should be revised 1 month after medication overuse has been stopped, when it is clear whether the patient has improved or not.

Distinction is now made between infrequent and frequent episodic tension-type headache. The idea is to single out the infrequent tension-type headaches, which are really not a significant health problem; frequent tension-type headache may be a health problem and chronic tension-type headache always is. Cluster headache and related trigeminal autonomic cephalalgias have been minimally changed and now include Short-lasting Unilateral Neuralgiform pain with Conjunctival injection and Tearing (SUNCT). In the chapter on other primary headaches (previously called miscellaneous primary headaches), substantial changes have been made. The diagnostic criteria are now more explicit and several new entities have been added: hypnic headache, primary thunderclap headache, hemicrania continuat, and new daily persistent headache. These are syndromes that have either been described for the first time or have been validated after the publication of ICHD-1.

The Secondary Headaches

There are more changes in the secondary than in the primary headaches. When ICHD-1 called the secondary headaches "associated with," for example, head trauma, ICHD-2 now says "attributed to," indicating a better documented causality. The explicit diagnostic criteria for the secondary headaches now have a standard format (Table 2-3). They always contain four-letter headings, all of which have to be fulfilled. Under A is given as many headache characteristics as are known for that particular type of secondary headache. Typically it is written: "Headache has at least *x* of the following characteristics: . . ."

Under B is described the presumed causative disorder and how it is diagnosed. Under C is given the reasons why this disorder is assumed to cause the headache, usually a very close temporal relationship, sometimes plus one or more other reasons. Finally, D specifies that the headache gets better or disappears after successful treatment or spontaneous remission of the causative disease. Another important feature is that it is now allowed to use

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TABLE 2-3 Standard Diagnostic Criteria for Secondary Headache

A. Headache has at least one of the following characteristics.[1,2]
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 disappears within 3 months (this may be shorter for some disorders) after successful treatment or spontaneous remission of the causative disorder.[3]

[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 establish causality. However, criteria B, C, and D usually effectively establish causality. 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 to stimulate research. This is why the formulation of criterion A now allows to mention a number of features in order of their frequency. 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 conditions cannot be treated or do not remit. In such cases criterion D may be replaced by: "Other causes ruled out by appropriate investigations."

the diagnosis of a secondary headache even if the headache phenomenologically has the character of a preexisting primary headache. In other words, if a patient has rare migraine attacks but after a head trauma starts having frequent severe attacks that dramatically change the impact of migraine, then the patient can receive two diagnoses: Migraine and Chronic posttraumatic headache. In the past it was only possible to diagnose a posttraumatic headache if this type of headache occurred de novo. Other major changes in the secondary headaches include a chapter, which now contains all infections; in the first edition, intracranial infections were in the chapter on intracranial disorders. A new chapter contains all the headaches caused by a disturbance of equilibrium, which means all systemic disorders such as arterial hypertension, hypoxia, and hypoglycemia. Finally, a new chapter has been added on headache attributed to psychiatric disorder. This chapter contains only two very rare entities, which are considered to be proven beyond doubt. It was the opinion of the experts that a number of psychiatric disorders may occasionally cause headache, but this has never been studied scientifically. To facilitate research, explicit criteria are given for a number of headaches attributed to different psychiatric disorders in the appendix. The appendix is also a new feature in ICHD-2. It contains, in addition to the psychiatric disorders, a number of disorders presumed to cause headache but not supported by sufficient scientific data. The appendix also contains entities that are considered to be nonexistent, but are given a last chance of study before being deleted completely. Alternative criteria to those in the main body of the classification are given when some committee members felt that these criteria would be superior, but insufficient scientific evidence was available to support this.

Scientific Evaluation of ICHD Classifications

Classifications have to be measured against some kind of standard. For headache disorders, no gold standard exists and the scientific evaluation and improvement must be an iterative process with advances taken in many small steps. The criteria for migraine without aura of ICHD-1 and ICHD-2 have been validated by the advent of the triptans. When these drugs are given by injection, the response rate is approximately 80 to 90% (2,20), indicating that the criteria of delineate a fairly homogenous group of patients. Unfortunately, there are no biochemical or imaging parameters that can serve as external validators for the headache classification. Likewise, the sensitivity and specificity of the diagnostic criteria of ICHD-1 could only be compared to the diagnosis of expert physicians (12), but not to an objective external standard. Because the first edition has now been in use for more than 15 years and has worked well, ICHD-2 diagnosis can be compared to ICHD-1 diagnoses of the same patients (5,6).

An important feature of a classification system is that is should be generalizable, that is, applicable in diverse clinical settings ranging from general practice to highly specialized headache centers. The classification should also be exhaustive, which means that there should be a diagnosis for every patient and it should be reliable with a low interrater diagnostic variability. Several studies in the general population have shown ICHD-1 to be exhaustive (17,18); the reliability and validity of the ICHD-1 have also been fairly good (13–15). ICHD-2 is so young that there have still not been studies on its reliability, validity, and exhaustiveness.

Research Opportunities Offered by ICHD-2

Like its predecessor, ICHD-2 is eminently suitable for scientific studies. Because the diagnostic criteria are so explicit, they can be tested in a variety of clinical settings. This is a good example of low-technology research, which can be done in almost any department that has a significant flow of patients. The secondary headaches are the least studied of all. It remains largely unknown how the characteristics are of the different types of secondary headache. For almost every one, any large series with a careful prospective clinical description of the headache would be valuable. Also criterion D, which specifies that headache gets better or disappears after treatment of the

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cause of the headache, is virtually unstudied. Is it really true that headache caused by a meningioma disappears after successful removal of the meningioma? Or do a significant number of patients continue to have the headache? In the latter case, a diagnosis of chronic postmeningioma headache would be documented. The same question can be asked for a large number of the secondary headaches. Is it true that headache can be caused by depression? Or are migraine and depression comorbid disorders? The number of questions that can be asked is almost endless. Finally, as the genetics of migraine and other headache disorders becomes better understood and genotype–phenotype correlation studies appear, that may lead to significant changes in our concepts and to future revisions of the classification.

Use of ICHD-2 and a Standardized Format of Chapters in the Present Book

In the present book, headache disorders are presented in exactly the same sequence as in ICHD-2. This is also true within each individual chapter. To facilitate overview, each chapter begins with the relevant section of the classification according to ICHD-2. Also the taxonomy and the explicit diagnostic criteria of ICHD-2 are used throughout.

The material for each disease is presented in exactly the same sequence: ICHD-2 and WHO (ICD-10 NA) diagnosis and code number; short description; other terms; epidemiology; genetics; anatomy, physiology, and pathophysiology; explicit diagnostic criteria and clinical features; prognosis; and management.

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