

Introduction

When you get this rather extensive document in your hands, please do not be overwhelmed. It is big, complicated, but not supposed to be learned by heart. The primary use is for research, but over the course of years it will probably influence the way we diagnose patients in our daily work.

The book contains a hierarchically constructed classification and operational diagnostic criteria for all headache disorders. The hierarchical system coding with up to four digits makes it possible to use the classification at different levels of sophistication. In routine practice, diagnosis will be made at the one or two digit level. In specialized centers, diagnosis will be made at the fourth digit level. We provide so-called “short descriptions” of most disorders. These short descriptions are less precise than the operational diagnostic criteria, but easier to remember and may be used in textbooks, for reading purposes, etc. Finally, comments and references are provided.

Is all this really necessary? Have we not been happy with the existing system? Do we really know enough about headache to introduce operational diagnostic criteria? These and many other questions have been raised again and again from colleagues, who have not been actively involved in the classification work. The classification of headache by the Ad Hoc Committee of the National Institute of Health served us well initially, but has been outdated for several years. Waters, the distinguished British epidemiologist, wrote in 1980: “Current migraine definitions are just descriptions rather than explicit definitions. The various features characteristic of migraine are said to be ‘commonly’, ‘often’ or ‘frequently’ present, but it is not precisely stated whether they have to be present in order to establish the diagnosis and, if so, how many of the features have to be present”. The same is even more true about the definition of tension headache. To state that a patient fulfils the criteria for migraine or tension headache of the NIH Ad Hoc Committee does not characterize the patient precisely, but is almost synonymous with stating that the patient has one or the other diagnosis according to the opinion of the investigator. In other fields of medicine operational diagnostic criteria are being introduced at a more or less advanced stage because this is the only way to ensure reasonably low inter-observer diagnostic variability. The process of creating and introducing such criteria is thought stimulating and makes clear what we do not know nosographically. Last, but not least, operational criteria can be proved or disproved, and they are easy to modify according to new developments in our knowledge.

Therefore, however tedious and irritating it may be, operational diagnostic criteria must be introduced if headache research is to accomplish significant advance in the future.

The late chief of neuroepidemiology of the National Institute of Health in the United States of America, Bruce S. Schoenberg, formulated the basic demands to a classification system and to diagnostic criteria as follows: “Any form of headache in a particular patient must fit one set of criteria and only one (but a patient may very well have more than one form of headache). Each set of diagnostic criteria should be as specific and as sensitive as possible”. In other words, only patients who really have the disease should have the diagnosis, but on the other hand all patients who really have the disease should also fulfil the diagnostic criteria. Specificity is achieved by rigorous criteria, which on the other hand may exclude too many patients.

Therefore, the chosen criteria for a particular diagnosis always represent a compromise between sensitivity and specificity. As elements in constructing a set of diagnostic criteria can serve only unambiguous parameters, words such as “often”, “sometimes” or “usually” are banned. Constructing the criteria presented here has involved careful weighing of every single word.

To classify and define diseases is always a difficult task, and the field of headache poses particular problems. Most fundamental of all is the scarcity of pathophysiological knowledge reflected in complete absence of laboratory tests which can be used as diagnostic criteria for any of the primary headache forms. Although typical and pure syndromes exist, there are many transitional forms. The headache of an individual patient may change over a lifetime, not only quantitatively but also qualitatively, e.g. migraine with aura may change into migraine aura without headache. One patient frequently has more than one form of headache, e.g. migraine without aura and episodic tension-type headache. At one point in a patient's life one form may predominate, but later it may be the other. It is a consequence of these problems that it has not been possible to classify patients, only to classify headaches.

To better understand this problem let us look at current practice. Patients have been categorized as having either classic migraine or common migraine in published scientific studies. Many patients, however, suffer both “classic” and “common” attacks. Some authors have classified patients as “classic” if they have just one attack with aura in their lifetime, some have required more. This results in very large diagnostic variability, and, furthermore, a patient could be classified as “common” in one trial and as “classic” in the next if in the meantime one or more attacks with aura had occurred or vice versa. In the new classification system the patient receives a diagnosis for each distinct headache form, i.e. migraine with aura and migraine without aura, which eliminates these problems.

That we cannot classify patients but only headaches does, however, introduce other problems. It is not possible to classify all headache episodes in every patient; most patients have too many, cannot remember them sufficiently well, have taken treatment, etc. The idea is to classify the most important form of headache or perhaps one or two more forms. Patients always have a number of attacks which for the above mentioned reasons cannot be formally classified. The patient can usually identify such episodes as abortive migraine attacks or tension-type headaches. Even with operational diagnostic criteria sound judgement and common sense are necessary.

The quantitative aspect of headache diagnosis should also be taken into account. It is therefore required that each diagnosis should be followed by the estimated number of headache days per year with that particular form of headache given in brackets. Further instructions are given under “General rules for use of the headache classification”. It is absolutely necessary to know these rules in order to use the classification correctly and, as an exception, this small part of the document should be learned by heart.