

## Proposal for an International Classification of the Epilepsies

On 19 July, 1968, the World Health Organization informed its experts who were working on a glossary of terms used in epileptology, of its wish that an International Classification of the Epilepsies be published to supplement the International Classification of Epileptic Seizures already established by the Commission on Terminology of the International League against Epilepsy.

Since the International League against Epilepsy had for several years expressed the same wish, the Secretary General of the International League and expert epileptologist of the WHO, Dr. H. Gastaut, considered it logical to ask the members of the above mentioned Commission on Terminology to prepare a draft of an International Classification of the Epilepsies, to be submitted for discussion at the quadrennial meeting of the League in New York on 27 September, 1969.

He proceeded in the same way as had been used in preparing the International Classification of Epileptic Seizures; that is to say he prepared a draft Classification of the Epilepsies which was submitted in August and November, 1968, to all the officers of the League and to the members of its Commission on Terminology and to the experts of the World Federation of Neurology and the International Federation of Societies for Electroencephalography and Clinical Neurophysiology attached to this Commission.

As it appeared to be impossible to call a meeting of the Commission on Terminology at a sufficiently early date, Prof. Gastaut decided to submit to the New York meeting his draft (which with some reservations was approved by one-third of the members of the Commission on Terminology), together with the criticisms made. *Epilepsia* has kindly undertaken to publish these documents in advance as a Supplement in order to facilitate a discussion (10: S14–S21). In the same Supplement, comments on the Classification of Epilepsy and a simplified Classification by Prof. R. L. Masland have been published (S22–S28).

Preceding the New York Congress the president of the International League against Epilepsy, Dr. J. K. Merlis, convened a meeting of a group of specialists including representatives of the World Federation of Neurology, the World Federation of Neurosurgical Societies, the International League against Epilepsy and the International Federation of Societies for Electroencephalography and Clinical Neurophysiology.

Discussions of these proposals during a full day and at a meeting of a sub-committee a few days later led to consensus on a Proposal for a Classification of the Epilepsies. This proposal was presented to the General Assembly by Dr. Merlis with the following Introduction.

## INTRODUCTORY REMARKS

In the supplement to *Epilepsia* which contains his proposed Classification of the Epilepsies, Dr. Gastaut has detailed the events that led to this publication. In his words "it appeared to be impossible to call a meeting of the Commission on Terminology at a sufficiently early date, and I decided to submit to the New York meeting my own draft (which with some reservations was approved by one-third of the members of the Commission on Terminology), together with the criticisms made".

It was obvious that detailed discussion at this meeting would be impossible, due to the strictures of time. In the belief that some preliminary discussions were essential if what appeared to be irreconcilable differences of approach and viewpoint were to be resolved, I, as President of our League, attempted the impossible. With the cooperation of the Presidents of the International Federation of Societies for Electroencephalography and Clinical Neurophysiology, the World Federation of Neurology and the World Federation of Neurosurgical Societies, an International Commission for Classification of the Epilepsies was appointed and arrangements were made to meet here, in New York, on 20 September, one week ago.

The officially designated members of this commission are as follows: for the IFSECN: L. Dondey\* (Paris), R. Hess\* (Zurich), C. Loeb\* (Genoa)—for the WFN: D. Janz\* (Heidelberg), S. Refsum\* (Oslo), D. Williams\* (London)—for the WFNS: H. Diemath (Salzburg), M. Falconer\* (London), C. Villavicencio\* (Santiago de Chile)—for the ILAE: H. Gastaut\* (Marseilles), K. Inanaga (Kurume), F. McNaughton\* (Montreal), O. Magnus\* (Wassenaar), R. Masland\* (New York), J. Merlis\*, Chairman (Baltimore).

The meeting was held as scheduled, with all but two of the members present. Dr. R. Broughton of Montreal participated and kindly served as a most efficient secretary.

The discussions occupied the whole of the day, last Saturday. The initial period could be described as chaotic, with many divergent approaches being presented and with frequent interruptions of speakers by, sometimes, rather warm disagreement. Nevertheless, as time went on, a core of agreement on certain basic principles seemed to emerge and, once this became apparent, the discussions proceeded with surprising ease. I think I can say that, despite being hoarse and tired at the end of the day, many, if not all, of the participants felt that it had been an instructive, productive and exhilarating experience.

It was unanimously agreed that our goal was to develop a classification of the epilepsies which could be used, and hopefully, would be used by all physicians, not only by those with special knowledge of, and interest in, the epilepsies. It was realized that care must be exercised not to intermix classification of seizures and classification of epilepsies in an inconsistent fashion, that the criteria for each class must be defined as clearly as possible, and that these criteria must be those which are necessary and

\* Participated in work-shop.

sufficient for classifying any given case. The approach, then, is operational; we asked, in effect, "what does a physician do when he sees a patient with epilepsy?"; and we answered that he takes a history and does a physical examination and perhaps other laboratory studies such as radiological examination. He pays careful attention to seizure description and other clinical features such as age of onset, the presence or absence of neurologic or psychologic evidence of brain disease or dysfunction, etc. Electroencephalographic study provides further crucial information and it is the combination and correlation of these two kinds of information, the clinical and the electrographic, which should enable him to classify a patient with epilepsy, regardless of the system of classification used.

We ask then, first, does the patient have a generalized epilepsy or does he have a partial (or focal) epilepsy? If he has a generalized epilepsy, is it primary or secondary?

I must emphasize that what we have called the primary form of generalized epilepsy is considered to be determined largely by positive clinical and EEG criteria, and is not a waste-basket, simply by exclusion, as the basket "idiopathic" so often is.

Some will be unhappy with the terms "primary" and "secondary". In a large sense, all the epilepsies are "secondary". Yet it was generally felt that, at the present, two groups of generalized epilepsies can be distinguished by clinical and EEG criteria. We use the term "secondary" to apply to those with evidence of underlying brain pathology, and it seems logical, then, to use the term "primary" for the other group. The Commission would be happy to learn of another pair of simple terms which could be used in similar fashion and which would be more satisfactory. We have included a third category of generalized seizures, *i.e.*, "undetermined", for those cases with generalized seizures, for whom data are inadequate for further classification into primary or secondary groups. You will note that we have adopted the following format in defining the criteria for each group:

(A) CLINICAL CRITERIA

- (1) Seizure form
- (2) Presence of neurologic or psychologic evidence of brain pathology
- (3) Age of onset
- (4) Etiology

(B) EEG CRITERIA

- (1) Interictal
- (2) Ictal

Finally, I would emphasize that this is a phenomenological approach, an attempt to provide a basic skeleton upon which can be engrafted the flesh and blood of seizure types, etiologies, etc. It rests on the combination of clinical and EEG data which, again, are necessary and sufficient. Other features of these nosologic groups may be of significance, *i.e.*, the response to different types of medication, the genetic factors, the presumed pathophysiology, etc., but these features are not necessarily crucial in classifying any given case. In some cases, however, they may contribute significantly. We did not think it useful to define all possible etiologies, nor indeed, to define rigidly, the epilepsies, although there was tacit agreement that we were concerned with those

conditions of chronically recurring seizures of primary cerebral origin, and not of seizures associated with systemic disease.

We believe that this classification can serve as a useful instrument to facilitate communication and clinical and epidemiological investigation. Indeed, it may encourage further research, for we do not, in the least, consider it fixed for all time and, hopefully, not even for a long time. As our knowledge of the epilepsies increases, it will undoubtedly require modification, not only as to criteria, but in fact, in its whole organization.

I urge you to give this classification your careful consideration. Time will not permit detailed discussion here, but you can and, in fact, are urged to, send comments and criticisms to me for presentation to the Commission.

J. K. MERLIS  
President of the ILAE

## Proposal for an International Classification of the Epilepsies

### I. GENERALIZED EPILEPSIES

#### 1. PRIMARY GENERALIZED EPILEPSIES

##### (A) *Clinical criteria*

##### (1) Seizures

Seizures which are generalized from the onset in the form of absences, bilateral myoclonus, and tonic-clonic seizures. One or more of these types of seizures can occur in the same patient

##### (2) Neurological status

The usual absence of neurological or psychological evidence of cerebral abnormality

##### (3) Age of onset

Onset in childhood and adolescence, although they are liable to persist to, and may even begin at, any age

##### (4) Etiology

Lack of any clear etiology

##### (B) *Electroencephalographic criteria*

##### (1) Interictal EEG

The presence (usually) of bilaterally synchronous spikes, polyspikes, spike-and-wave, or polyspike-wave complexes. These may occur singly or rhythmically, at about 3/s. They are spontaneous or are induced by hyperventilation, intermittent photic stimulation, or sleep

## (2) Ictal EEG

The occurrence (usually) of synchronous and symmetrical discharges with a given type\* of seizure (rhythmic at about 3/s spike-and-wave complexes during absences; polyspike-wave complexes during bilateral myoclonus; “recruiting” rhythms at about 10/s followed by rhythmic polyspike-waves during tonic-clonic seizures)

## 2. SECONDARY GENERALIZED EPILEPSIES

## A. Clinical criteria

## (1) Seizures

Seizures which are generalized *from the onset* in the form of absences; bilateral myoclonus; tonic or atonic seizures; or tonic-clonic seizures. One or more of these seizures can occur in a single patient

## (2) Neurological status

The presence (usually) of neurological or psychological signs (*i.e.* mental deficiency or deterioration), or both, indicative of diffuse cerebral pathology

## (3) Age of onset

Onset at any age; most frequent in childhood

## (4) Etiology

May be ascribed (usually) to diffuse or multifocal cerebral lesions

## B. Electroencephalographic criteria

## (1) Interictal EEG

Slow background activity with sharp and slow wave complexes usually symmetrical and synchronous, or asymmetrical or even asynchronous. These are less frequently induced by hyperventilation and photic stimulation. Sleep may be effective

## (2) Ictal EEG

Ictal patterns which may contain diminution in amplitude of background EEG activities, a low voltage rapid discharge, a “recruiting” rhythm at about 10/s, sharp and slow wave discharges at about 2/s, and spike-and-wave or polyspike-wave discharges. The sharp and slow wave discharges are less synchronous and symmetrical and are more variable in topographical distribution than the other discharge types and than those in primary generalized epilepsy. The correlation between these ictal EEG patterns and the seizure types is not as good as in primary generalized epilepsy\*\*

## 3. UNDETERMINED GENERALIZED EPILEPSIES

Using the criteria above, the information available concerning a given patient with a generalized epilepsy may not be adequate to determine whether it is primary or secondary. The patient will then be classified as generalized epilepsy, undetermined.

\* The precise ictal patterns of the various types of epileptic seizures are described in detail in the International Classification of Epileptic Seizures (*Epilepsia*, 1970, 11: 102–113).

\*\* The most usual correlates are described in the International Classification of Epileptic Seizures.

## II. PARTIAL (FOCAL, LOCAL) EPILEPSIES

A. *Clinical criteria*

## (1) Seizures

Partial seizures (of local onset) with or without generalization whose manifestations (chiefly initial) are of many forms as detailed in The International Classification of Epileptic Seizures. Postictal focal neurological deficit may be present

## (2) Neurological status

The presence (frequently) of neurological signs related to the epileptogenic lesion.

## (3) Age of onset

Onset at any age

## (4) Etiology

Associated (usually) with brain damage

B. *Electroencephalographic criteria*

## (1) Interictal EEG

Occurrence of local spikes or spike-and-wave complexes (usually). Sleep, hyperventilation and photic stimulation are less effective activators than in other types of epilepsies. The site of the epileptogenic focus should correspond to the clinical symptoms of the seizures

## (2) Ictal EEG

Local discharges related to the lesion. In many cases, these may be diffuse; they may even be absent. Postictal focal abnormalities may be present.

## III. UNCLASSIFIABLE EPILEPSIES

This group comprises all those epilepsies which cannot be classified in one of the above mentioned generalized or partial groups, either because they are atypical or because data are insufficient. The epilepsy with "erratic seizures" in the newborn and the unilateral seizures of childhood may have to be included in this category.

The above *groups* may differ significantly in cyclic characteristics, response to medication, prognosis and so forth. As our understanding of the basic mechanisms of the epilepsies increases, modification of this classification will undoubtedly be necessary.