

# Proposal for Classification of Epilepsies and Epileptic Syndromes

\*Commission on Classification and Terminology of the  
International League Against Epilepsy

## Preface

The Commission on Classification and Terminology of the International League Against Epilepsy (ILAE) herewith presents a proposal for an International Classification of Epilepsies and Epileptic Syndromes (ICE). This proposal is to be presented for a vote by the General Assembly of the ILAE in Hamburg, F.R.G., on September 9, 1985.

On December 28, 1981, Dr. Mogens Dam, the President of ILAE, charged the Commission to develop a classification of the epilepsies, get it approved by the active chapters of the ILAE and other pertinent international societies, develop a current dictionary of the epilepsies, and promote its use throughout the world.

The purpose of the ICE is to supplement the International Classification of Epileptic Seizures, the revised form of which was accepted by the General Assembly of the ILAE in September, 1981.

The Commission began by examining the Proposal for International Classification of Epilepsies first propounded in 1970 (Merlis). It rapidly became apparent that the problems for the ICE were different from those of the seizure classification and would not be solved in the same objective manner, namely, by excluding everything on which no video documentation existed. In the past, a major obstacle for the creation of an ICE had been the apparently irreconcilable opinions of different schools about syndromic classifications and terminology. Merlis graphically described this problem, which at that time was solved by the publication of the minimum statement on which consensus could be reached.

The present Commission realized that no international classification would be acceptable if it canonized one point of view. Because science is essentially pluralistic, a scientifically valid classification would have to reflect this pluralism. The Commission therefore repeatedly

sought the advice of other epileptologists. In July, 1983, a symposium was organized in Marseille, France, and the proceedings of this symposium have recently been published (Roger et al, 1985). In March, 1984, a workshop was held in Bethesda, MD, U.S.A., hosted by Dr. Roger Porter and the Epilepsy Branch of the National Institute of Neurological and Communicative Disorders and Stroke. Contributors were an advisory group that included Drs. A. V. Delgado-Escueta (Los Angeles, CA, U.S.A.), R. H. Mattson (West Haven, CT, U.S.A.), T. A. Pedley (New York, NY, U.S.A.), J. K. Penry (Winston-Salem, NC, U.S.A.), L. P. Quesney (Montreal, Canada), and H. G. Wieser (Zurich, Switzerland). These consultants contributed and discussed video-documented data of epilepsies in defined loci studied with depth electrodes.

The Commission met at the Epilepsy International Symposia of 1982 and 1983, held in London, England and Washington, DC, U.S.A., respectively, as well as during the Marseille and Bethesda meetings. On two occasions, both early and late in the proceedings, drafts were mailed to the national branches of the ILAE with a call for comments. Much valuable input resulted, and the present proposal is the result of these deliberations, which culminated with a Commission meeting in Berlin in March 1985. This proposal is the result of the elimination of superfluous or excessively controversial items. Many of these controversial issues are addressed in the two appendices. The aim of the ICE was to provide a scheme that would be compatible with the view of the majority of international epileptologists, and thus suitable for mutual exchange of ideas. The Commission hopes that the proposed ICE represents such compatibility and will be manageable for practical use in clinical and research situations.

## PROPOSAL FOR AN INTERNATIONAL CLASSIFICATION

### Introduction

In the past three decades, increasing international discussion and cooperation in the field of epilepsy have

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resulted in an increasing realization of the need for a common international terminology and classification. This is the precondition for comparability of results in research and therapy, and for a meaningful exchange of ideas.

The ILAE has acknowledged its responsibility for this by appointing a Commission on Classification and Terminology. At an early stage of this Commission's work, it was decided that two separate systems of classification were needed: one for epileptic seizures, and one for epilepsies and epileptic syndromes.

An International Classification of Epileptic Seizures (ICES) was accepted by the ILAE in 1969 and revised in 1981. An outline of an ICE was proposed in 1970, but never elaborated in detail. The proposal presented here, a result of the efforts of the present Commission, is based on that outline; it takes into account some problems with the terminology suggested in 1970 and refers to the revised ICES.

While the term "epilepsies"—used in the heading of this classification—may suggest a classification of disease, in fact, at this time, only a few epileptic disease entities, such as Lafora's disease, have been identified. Thus, at our present stage of knowledge a classification of epilepsies will, by necessity, represent mainly a classification of syndromes rather than diseases.

An epileptic syndrome is defined as an epileptic disorder characterized by a cluster of signs and symptoms customarily occurring together. The signs and symptoms may be clinical (e.g., case history, seizure type, modes of seizure recurrence, and neurological and psychological findings) or findings detected by ancillary studies (e.g., EEG, x-ray, CT, and NMR). In contradistinction to a disease, a syndrome does not necessarily have a common etiology and prognosis. Some epileptic syndromes, however, are of great prognostic importance.

Syndromes may belong to different orders. Some represent rather broad concepts, others are much more specific. Syndromes of high specificity may include syndromes of lesser specificity. Overlapped syndromes and inclusion of one syndrome within another occur frequently. Thus, patients with a highly specific syndrome, such as juvenile myoclonic epilepsy, may also demonstrate epilepsy with grand mal on awakening, juvenile absence epilepsy, and photosensitivity. A syndrome of low specificity is sleep grand mal, which is defined by two criteria: the occurrence of generalized convulsive seizures, and restriction of these seizures to the state of sleep. Many of these patients can be classed more precisely if the syndrome is elaborated by the presence of other seizure types, but many others cannot be classed so easily. It may even be impossible to decide whether a patient's seizures are of focal or generalized onset. Classing these seizures as sleep grand mal epilepsy is perhaps not totally satisfac-

tory, but it is surely preferable to regarding them as unclassifiable.

Some of the epileptic disorders included in this classification are diseases, and in some disorders, considered syndromes at the moment, a common etiology may still be discovered. For the sake of convenience, all these disorders are included in one ICE.

Two dichotomies are widely used to shape the major classes: The first separates epilepsies with generalized seizures (generalized epilepsies) from epilepsies with partial or focal seizures (localization-related partial or focal epilepsies). The other separates epilepsies of known etiology (symptomatic or "secondary" epilepsies) from those that are idiopathic (primary) or cryptogenic.

The present proposal maintains these dichotomies, which formed the base of the 1970 proposal. However, it deviates from some of the terms suggested in 1970, such as "primary" and "secondary." The meaning of primary and secondary has been frequently misunderstood. In accordance with the ICES, many writers label all epilepsies with seizures that have to be classed as generalized, "primary generalized epilepsies," and the term "secondary generalized" is used for epilepsies with generalized [mostly generalized tonic-clonic seizures (GTCS)] seizures of partial or focal onset. The terms "idiopathic" and "symptomatic," which are almost exact synonyms of "primary" and "secondary" in their intended meaning, are therefore preferred here for clarity. The term "partial epilepsy"—which ignores the fundamental problem of talking of "partial" diseases—creates difficulties with the classification of patients whose only seizure type is simple partial (focal, local) evolving to generalized tonic-clonic seizures.

Should the patient who often experiences only a short and almost imperceptible aura before the GTCS be considered to suffer from "partial" epilepsy, there would be little hope that this could be understood by other than a small circle of epileptologists. Moreover, the term "focal," which would be appropriate for these cases, cannot be used everywhere without reservation, because in cases classed as focal epilepsy many epileptologists envision a well-defined constant epileptic focus. This idea is belied by many children suffering from idiopathic epilepsy with focal seizures of shifting site and side of onset.

Thus, the Commission ventured to propose "localization-related epilepsies" as the most appropriate term. We realize that some epileptologists prefer the term "focal" and others "partial." Because of the lack of unanimity all three terms are proffered here.

The present proposal recognizes that the dichotomy of generalized and localization-related epilepsies is not applicable in all cases because occasionally both generalized and focal features may present in one patient, or neither of these features may be evident. Therefore, a

third category of epilepsies, undetermined as to whether focal or generalized, was added.

Some syndromes defy classification as either symptomatic or idiopathic, because there may be both idiopathic and symptomatic cases occurring in this syndrome, or there may be insufficient evidence for definitive placement.

Some authors would even advocate the demise of the terms "idiopathic" and "symptomatic," because they feel that an interaction of these factors is the rule. However, the differentiation of idiopathic versus symptomatic syndromes is a tradition that will be difficult to abandon. The concept of age-related syndromes might prove more suitable than the above terms, especially for the classification of generalized epilepsies, and is therefore included in this classification.

The proposed classification includes a category of special syndromes. This allows the handling of data for patients with specific, identifiable features that qualify their epilepsy—for example, a patient with musicogenic complex partial seizures—so that these features are not lost in the general store of complex partial seizures.

### International classification of epilepsies and epileptic syndromes

#### 1. Localization-related (focal, local, partial) epilepsies and syndromes.

##### 1.1 Idiopathic with age-related onset.

At present, two syndromes are established, but more may be identified in the future:

- Benign childhood epilepsy with centrotemporal spike
- Childhood epilepsy with occipital paroxysms

##### 1.2 Symptomatic.

This category comprises syndromes of great individual variability, which will mainly be based on anatomical localization, clinical features, seizure types, and etiological factors (if known). Major examples and descriptions of varieties subsumed under this heading are given in Appendix I.

#### 2. Generalized epilepsies and syndromes.

##### 2.1 Idiopathic, with age-related onset, listed in order of age.

- Benign neonatal familial convulsions
- Benign neonatal convulsions
- Benign myoclonic epilepsy in infancy
- Childhood absence epilepsy (pyknolepsy)
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy (impulsive petit mal)
- Epilepsy with grand mal seizures (GTCS) on awakening

Other generalized idiopathic epilepsies, if they do not belong to one of the above syndromes,

can still be classified as generalized idiopathic epilepsies.

##### 2.2 Idiopathic and/or symptomatic, in order of age of appearance.

- West syndrome (infantile spasms, Blitz-Nick-Salaam Krämpfe)
- Lennox-Gastaut syndrome
- Epilepsy with myoclonic-astatic seizures
- Epilepsy with myoclonic absences

##### 2.3 Symptomatic.

###### 2.3.1 Nonspecific etiology.

- Early myoclonic encephalopathy

###### 2.3.2 Specific syndromes.

- Epileptic seizures may complicate many disease states.

Included under this heading are those diseases in which seizures are a presenting or predominant feature. These are detailed in Appendix II.

#### 3. Epilepsies and syndromes undetermined as to whether they are focal or generalized.

##### 3.1 With both generalized and focal seizures.

- Neonatal seizures
- Severe myoclonic epilepsy in infancy
- Epilepsy with continuous spike-waves during slow wave sleep
- Acquired epileptic aphasia (Landau-Kleffner syndrome)

##### 3.2 Without unequivocal generalized or focal features.

This heading covers all cases with GTCS where clinical and EEG findings do not permit classification as clearly generalized or localization-related, such as in many cases of sleep grand mal.

#### 4. Special syndromes.

##### 4.1 Situation-related seizures (Gelegenheitsanfälle).

- Febrile convulsions
- Seizures related to other identifiable situations such as stress, hormonal changes, drugs, alcohol, or sleep deprivation

##### 4.2 Isolated, apparently unprovoked epileptic events.

##### 4.3 Epilepsies characterized by specific modes of seizure precipitation.

##### 4.4 Chronic progressive epilepsy partialis continua of childhood.

### Definitions

#### *Localization-related (focal, local, partial) epilepsies and syndromes*

These are epileptic disorders where seizure semiology or findings at investigation disclose a localized origin of the seizures. This includes patients with small circumscribed constant epileptogenic lesions (anatomical or

functional), i.e., true focal epilepsies, but also patients with less well-defined lesion, whose seizures may originate from variable loci. In most symptomatic localization-related epilepsies, the epileptogenic lesion can be traced to one part of one cerebral hemisphere, but in idiopathic age-related epilepsies with focal seizures, corresponding regions of both hemispheres may be functionally involved.

#### *Generalized epilepsies and syndromes*

According to ICES, generalized epilepsies and syndromes are epileptic disorders with generalized seizures, i.e. "seizures in which the first clinical changes indicate initial involvement of both hemispheres... The ictal encephalographic patterns initially are bilateral."

#### *Epilepsies and syndromes undetermined as to whether they are focal or generalized*

There may be two different reasons why this cannot be determined: (1) the patient has both focal and generalized seizures together or in succession (e.g., partial seizures plus absences), likewise for both focal and generalized EEG seizure discharges (e.g., temporal spike focus plus independently bilateral spike-wave); and (2) there are no positive signs of either focal or generalized seizure onset. The most common reasons for this are that the seizures occur during sleep, the patient recalls no aura, and ancillary investigations including EEG are unrevealing.

#### *Idiopathic epilepsies and syndromes*

The term "idiopathic" comes from the Greek *idios*, meaning self, own, or personal. Idiopathic epilepsies and syndromes are described as disorders "not preceded or occasioned by another," according to the Oxford English Dictionary. They have no known or suspected etiology, other than possible hereditary predisposition.

#### *Symptomatic epilepsies and syndromes*

These epilepsies are considered to be the consequence of a known or suspected disorder of the central nervous system.

#### *Idiopathic localization-related epilepsies*

These are childhood epilepsies with partial seizures and focal EEG abnormalities. They are age-related, without demonstrable anatomical lesions, and subject to spontaneous remission. Clinically, patients have neither neurological and intellectual deficit nor a history of antecedent illness, but frequently have a family history of benign epilepsy. The seizures are usually brief and rare, but may be frequent early in the course. The seizure patterns may vary from case to case, but usually remain constant in the same child. The EEG is characterized by

normal background activity and localized high-voltage repetitive spikes, which are sometimes independently multifocal. Brief bursts of generalized spike-waves can occur. Focal abnormalities are increased by sleep, and are without change in morphology.

**Benign childhood epilepsy with centro-temporal spikes.** This is a syndrome of brief, simple, partial, hemifacial motor seizures, frequently having associated somatosensory symptoms, which have a tendency to evolve into GTCS. Both seizure types are often related to sleep. Onset is between 3 and 13 years of age (peak, 9–10), and recovery before ages 15–16. Genetic predisposition is frequent, and there is male predominance. The EEG has blunt high-voltage centro-temporal spikes, often followed by slow waves that are activated by sleep and tend to spread or shift from side to side.

**Childhood epilepsy with occipital paroxysms.** This syndrome is, in general respects, similar to the previous one. The seizures start with visual symptoms (amaurosis, phosphenes, illusions, or hallucinations), and are often followed by a hemiclonic seizure or automatism. In a quarter of the cases, the seizures are immediately followed by migrainous headache. The EEG has paroxysms of high-amplitude spike-waves or sharp waves recurring rhythmically on the occipital and posterior temporal areas of one or both hemispheres, but only when the eyes are closed. During seizures, the occipital discharge may spread to the central or temporal region. At present no definite statement on prognosis is possible.

#### *Idiopathic generalized epilepsies (age-related)*

These are forms of generalized epilepsies in which all seizures are initially generalized, and their EEG expression is a generalized, bilateral, synchronous, symmetrical discharge (such as described in the seizures classification for the corresponding type). They occur in a patient usually presenting a normal interictal state, without neurological or neuroradiological signs. In general, interictal EEGs show normal background activity and generalized discharges, such as spikes, polyspikes, spike-waves, and polyspike-waves at 3 c/s or more. The discharges are increased by slow sleep. The various syndromes of idiopathic generalized epilepsies differ mainly in their ages of onset.

#### *Idiopathic and/or symptomatic generalized epilepsies*

This category includes syndromes comprising both idiopathic and symptomatic cases (e.g., West and Lennox-Gastaut syndromes), as well as syndromes whose status as idiopathic or symptomatic epilepsies is unclear because they have some features suggesting an idiopathic (lack of known etiology, strong genetic predisposition) and others suggesting a symptomatic (intellectual delay or deficit, neurological abnormalities) origin.

### *Symptomatic generalized epilepsies and syndromes*

These generalized epilepsies, most often occurring in infancy and childhood, are characterized by generalized seizures with clinical and EEG features different from those of idiopathic generalized epilepsies. They may be of one type only but, more often, are of several types, including myoclonic jerks, tonic seizures, atonic seizures, and atypical absences. Their EEG expression is bilateral but less rhythmical than in idiopathic generalized epilepsies, and more or less asymmetrical. Interictal EEG abnormalities differ from those of idiopathic generalized epilepsies, appearing as suppression bursts, hypersarrhythmia, slow spike-waves, or generalized fast rhythms. Focal abnormalities may be associated with any of the above. There are clinical, neuropsychological, and neuroradiological signs of a usually diffuse, specific, or nonspecific encephalopathy.

### *Generalized idiopathic epilepsies (age-related)*

**Benign neonatal familial convulsions.** These are rare, dominantly inherited disorders manifesting mostly on the second and third days of life, with clonic or apneic seizures and no specific EEG criteria. History and investigations reveal no etiological factors. About 14% of these patients later develop epilepsy.

**Benign neonatal convulsions.** These are very frequently repeated clonic or apneic seizures occurring around the fifth day of life, without known etiology or concomitant metabolic disturbance. Interictal EEG often shows alternating sharp theta waves. There is no recurrence of seizures, and the psychomotor development is not affected.

**Benign myoclonic epilepsy in infancy.** This form is characterized by brief bursts of generalized myoclonus that occur during the first or second year of life in otherwise normal children who often have a family history of convulsions or epilepsy. EEG recordings show generalized spike-waves occurring in brief bursts during the early stages of sleep. These attacks are easily controlled by appropriate treatment. They are not accompanied by any other types of seizure, although GTCS may occur during adolescence. The epilepsy may be accompanied by a relative delay of intellectual development and minor personality disorders.

**Childhood absence epilepsy (pyknolepsy).** This syndrome occurs in children of school age (peak manifestation, ages 6–7) with a strong genetic predisposition in otherwise normal children. It appears more frequently in girls than in boys. It is characterized by very frequent (several to many per day) absences. The EEG reveals bilateral, synchronous symmetrical spike-waves, usually 3/s, on a normal background activity. During adolescence, GTCS often develop. Otherwise, absences may remit or, more rarely, persist as the only seizure type.

**Juvenile absence epilepsy.** The absences of this syndrome are the same as in pyknolepsy, but absences with retropulsive movements are less common. Age of manifestation is around puberty. Seizure frequency is lower than in pyknolepsy, with absences occurring less frequently than every day, mostly sporadically. Association with GTCS is frequent, and they precede the absence manifestations more often than in childhood absence epilepsy, often occurring on awakening. Not infrequently, the patients also have myoclonic seizures. Sex distribution is equal. The spike-waves are often faster than 3/s. Response to therapy is excellent.

**Juvenile myoclonic epilepsy (impulsive petit mal).** This syndrome appears around puberty and is characterized by seizures with bilateral, single or repetitive, arrhythmic, irregular myoclonic jerks, predominantly in the arms. Some patients may suddenly fall from a jerk. No disturbance of consciousness is noticeable. The disorder may be inherited and sex distribution is equal. Often, there are GTCS and, less often, infrequent absences. The seizures usually occur shortly after awakening, and are often precipitated by sleep deprivation. Interictal and ictal EEG have rapid, generalized, often irregular spike-waves and polyspike-waves; there is no close phase correlation between EEG spikes and jerks. Frequently, the patients are photosensitive. Response to appropriate drugs is good.

**Epilepsy with GTCS on awakening.** This is a syndrome with onset mostly in the second decade of life. The grand mal seizures are presumably mainly GTCS and occur exclusively or predominantly (over 90% of the time) shortly after awakening regardless of the time of day, or in a second seizure peak in the evening period of relaxation. If there are other seizures, these are mostly absences or myoclonic, as in juvenile myoclonic epilepsy. Seizures may be precipitated by sleep deprivation and other external factors. Genetic predisposition is relatively frequent. The EEG shows one of the patterns of idiopathic generalized epilepsy. There is a significant correlation with photosensitivity.

**West syndrome (infantile spasms, Blitz-Nick-Salaam Krämpfe).** Usually, West syndrome consists of a characteristic triad: infantile spasms, arrest of psychomotor development, and hypersarrhythmia, although one element may be missing. Spasms may be flexor, extensor, lightning, or nods but most commonly are mixed. Onset peaks between 4 and 7 months and is always before 1 year. Boys are more commonly affected, and the prognosis is generally poor. West syndrome may be separated into two groups. The symptomatic group is characterized by the previous existence of brain damage signs (psychomotor retardation, neurological signs, radiological signs, or other types of seizures) or by a known etiology. The smaller, idiopathic group is characterized by the absence of previous signs of brain damage and of

known etiology. The prognosis is partly based on early therapy with adrenocorticotrophic hormone (ACTH) or oral steroids. However, it principally depends on the symptomatic or idiopathic character of the syndrome. A number of idiopathic cases have had favorable prognoses without psychic impairment and later epilepsy when treated early.

**Lennox-Gastaut syndrome.** This syndrome manifests itself in children from 1 to 8 years of age, but appears mainly in preschool-age children. The most common seizure types are tonic-axial, atonic, and absence seizures, but other types such as myoclonic, GTCS, or partial are frequently associated with this syndrome. Seizure frequency is high, and status epilepticus frequent (stuporous states with myoclonias, tonic, and atonic seizures). The EEG usually has abnormal background activity, slow spike-waves of less than 3/s and, often, multifocal abnormalities. During sleep, bursts of fast rhythms (around 10/s) appear. In general, there is mental retardation. Seizures are difficult to control, and the development is mostly unfavorable. In 60% of cases, the syndrome occurs in children suffering from a previous encephalopathy, but it is primary in other cases.

**Epilepsy with myoclonic-astatic seizures.** Manifestation begins between 7 months and 6 years, mostly from 2 to 5 years, with (except if beginning in the first year) twice as many boys affected. There is frequently hereditary predisposition and usually a normal developmental background. The seizures are myoclonic, astatic, myoclonic-astatic, absences with clonic and tonic components, and tonic-clonic. Status frequently occurs. Tonic seizures develop late in the course of unfavorable cases. The EEG, initially often normal except for 4-7/s rhythms, may have irregular fast spike-wave or polyspike-wave and, during status, irregular 2-3/s spike-wave. Course and outcome are variable.

**Epilepsy with myoclonic absences.** This syndrome is clinically characterized by absences accompanied by severe bilateral rhythmical clonic jerks, often associated with a tonic contraction. On the EEG they are always accompanied by bilateral, synchronous, and symmetrical discharge of rhythmical spike-wave at 3/s, similar to childhood absence. These seizures occur many times a day. Awareness for the jerks may be maintained. Associated seizures are rare. Age of onset is about 7 years and there is a male preponderance. Prognosis is less favorable than in pyknolepsy due to resistance to therapy of the seizures, mental deterioration, and possible evolution to other types of epilepsy such as Lennox-Gastaut syndrome.

*Generalized symptomatic epilepsies of nonspecific etiology (age-related)*

**Early myoclonic encephalopathy.** The principal features of this syndrome are onset before 3 months of

age, initially fragmentary myoclonus, then erratic partial seizures, massive myoclonias, or tonic spasms. The EEG is characterized by suppression-burst activity, which may evolve into hypsarrhythmia. The course is severe, psychomotor development is arrested, and death may occur in the first year. Familial cases are frequent and suggest the influence of one or several congenital metabolic errors, but there is no constant genetic pattern.

The status of early infantile epileptic encephalopathy with suppression bursts, described by Ohtahara in relation to early myoclonic encephalopathy, is at present unclear, especially in view of its ictal features and its frequent evolution into a syndrome indistinguishable from West syndrome.

### **Epilepsies and syndromes undetermined as to whether they are focal or generalized**

#### *Neonatal seizures*

Neonatal seizures differ from those of older children and adults. The most frequent neonatal seizures are described as subtle because the clinical manifestations are frequently overlooked. These include tonic, horizontal deviation of the eyes with or without jerking, eyelid blinking or fluttering, sucking, smacking, or other buccal-lingual oral movements, swimming or pedaling movements, and occasionally, apneic spells. Other neonatal seizures occur as tonic extension of the limbs, mimicking decerebrate or decorticate posturing. These are particularly seen in premature infants. Multifocal clonic seizures characterized by clonic movements of a limb, which may migrate to other body parts or other limbs, or focal clonic seizures, which are much more localized, may occur. In the latter, the infant is usually not unconscious. Rarely, myoclonic seizures may occur, and the EEG pattern is frequently that of suppression-burst activity. The tonic seizures have a poor prognosis because they frequently accompany intraventricular hemorrhage. The myoclonic seizures also carry a poor prognosis because they are frequently a part of the early myoclonic encephalopathy syndrome.

#### *Severe myoclonic epilepsy in infancy*

Severe myoclonic epilepsy in infants is a recently defined syndrome. The characteristics include a family history of epilepsy or febrile convulsions, normal development before onset, seizures beginning during the first year of life in the form of generalized or unilateral febrile clonic seizures, secondary appearance of myoclonic jerks, and often partial seizures. EEGs show generalized spike-waves and polyspike-waves, early photosensitivity, and focal abnormalities. Psychomotor development is retarded from the second year of life on, and ataxia, pyramidal signs, and interictal myoclonus appear. This type of epilepsy is very resistant to all forms of treatment.

*Epilepsy with continuous spike-waves during slow sleep*

This syndrome results from the association of various seizure types, partial or generalized, occurring during sleep, and atypical absences when awake. Tonic seizures do not occur. The characteristic EEG pattern consists of continuous diffuse spike-waves during slow wave sleep, which is seen after the onset of seizures. Duration varies from months to years. The prognosis is guarded because of the appearance of neuropsychologic disorders, despite the usually benign evolution of seizures.

*Acquired epileptic aphasia (Landau-Kleffner syndrome)*

The Landau-Kleffner syndrome is a childhood disorder associating an acquired aphasia, multifocal spikes, and spike and wave discharges. Epileptic seizures and behavioral and psychomotor disturbances occur in two-thirds of the patients. There is verbal auditory agnosia and rapid reduction of spontaneous speech. The seizures are generally generalized convulsive or partial motor. They are rare, and remit before the age of 15 years, as do the EEG abnormalities.

**Special syndromes***Febrile convulsions*

Febrile convulsions are an age-related disorder almost always characterized by generalized seizures occurring during an acute febrile illness. The majority of febrile convulsions are brief and uncomplicated, but a minority may be more prolonged and followed by transient or permanent neurological sequelae, such as the Hemiplegia hemiatrophy epilepsy (HHE) syndrome. There is a tendency for recurrence of febrile convulsions in about one-third of those affected. Controversy about the risks of developing epilepsy afterwards have largely been resolved by some recent large studies, and it seems that the overall risk is not >4%. The indications for prolonged drug prophylaxis against recurrence of febrile convulsions are more clearly defined now and the majority do not require it. Essentially, this condition is a relatively benign disorder of early childhood.

*Epilepsies characterized by specific modes of seizure precipitation (reflex epilepsies)*

In simple forms, seizures are precipitated by simple sensory stimuli (e.g., light flashes). The intensity of the stimuli is decisive, the latency of the response short (seconds or less), and mental anticipation of stimulus without effect. In complex forms, the triggering mechanisms are elaborate (e.g., sight of one's own hand, listening to a certain piece of music). The specific pattern of the stimulus, not the intensity, is the decisive factor. Latency of response is longer (in the range of minutes), and mental anticipation of stimulus, even in dreams, may be effective.

*Kojewnikow's syndrome*

Two types of Kojewnikow's syndrome are now recognized, but only one of these two types is included among the epileptic syndromes of childhood, because the other one is not specifically related to this age. The first type represents a particular form of rolandic partial epilepsy, in both adults and children, and is related to a variable lesion of the motor cortex. Its principal features are motor partial seizures, always well localized; often late appearance of myoclonias in the same site where there are somato-motor seizures; an EEG with normal background activity and focal paroxysmal abnormalities (spikes and slow waves); occurrence at any age in childhood and adulthood; frequently demonstrable etiology (tumoral, vascular); and no progressive evolution of the syndrome (clinical, electroencephalographic, or psychological), except the evolutive character of the causal lesion. The childhood disorder, suspected to be of viral etiology, has onset between 2 and 10 years (peak, 6 years) with seizures that are motor partial seizures, but are often associated with other types. Fragmentary motor seizures appear early in the course of the illness and are initially localized, but later become erratic and diffuse, and persist during sleep. A progressive motor deficit follows, and mental deterioration occurs. The EEG background activity shows asymmetric and slow diffuse delta waves, with numerous ictal and interictal discharges that are not strictly limited to the rolandic area.

**Appendix I: Varieties of symptomatic localization-related epilepsies according to the anatomical localization**

Preliminary descriptions of syndromes related to sometimes unusually precise localizations are offered. The basis for these descriptions includes data collected by contributors working with depth electrodes. It is fully recognized that patients thus studied represent a highly selected minority and that these descriptions are, to a certain extent, abstractions, because frequently-discharging lesions are not confined to such closely circumscribed loci. However, the commission felt sufficiently confident in the integrity of these studies to incorporate them in the classification.

*Frontal lobe epilepsies*

A feature of epilepsies with a frontal focus is very frequent, short attacks with minimal or no postictal confusion, which are often mistaken for psychogenic seizures. Status epilepticus is particularly frequent in frontal lobe epilepsies.

*Supplementary motor*

Seizure patterns are postural, simple focal tonic, with vocalization, speech arrest, fencing, and complex focal with urinary incontinence. The ictal EEG shows flatten-

ing rhythmic polyspikes (16–24 Hz), and secondary generalization. Depth electrode exploration is often required for detection. Psychological tests may show impaired verbal fluency (dominant hemisphere), or impaired design fluency (nondominant hemisphere). Common etiologies are focal atrophy, tumors, and arteriovenous malformations.

#### *Cingulate*

Seizure patterns are complex focal with initial automatisms with sexual features, vegetative signs, changes in mood and affect, and urinary incontinence. For the detection of EEG focus, depth electrode exploration is mandatory. Psychological findings and etiology are similar to the previous syndrome.

#### *Anterior (polar) frontal region*

The seizure patterns include initial loss of contact, adersive and subsequent contraversive movements of head and eyes, axial clonic jerks and falls, and autonomic signs. There is frequently evolution to GTCS.

#### *Orbito-frontal*

Seizure patterns are complex focal with initial automatisms or olfactory hallucinations, autonomic signs, and urination. The ictal EEG shows flattening, rhythmic polyspikes (16–24 Hz), and secondary generalization. Nasoethmoidal and orbital electrodes are often required for detection. Common etiologies are trauma, astrocytomas, and oligodendrogliomas.

#### *Dorsolateral*

Seizure patterns are simple focal tonic with versive moments and aphasia, and complex focal with initial automatisms. The EEG focus is, in general, satisfactorily detected by interictal and ictal surface EEG. As psychological findings, perseveration, poor judgment, and disinhibition have been reported. Common etiologies are traumas, astrocytomas, and oligodendrogliomas.

#### *Epilepsies of the motor cortex*

These are mainly characterized by simple partial seizures, and their localization depends on the side and topography of the area involved. In cases of the lower prerolandic area, there may be speech arrest, vocalization or dysphasia, tonic-clonic movements of the face on the contralateral side, or swallowing. Generalization of the seizure frequently occurs. In the rolandic area, partial motor seizures without march or jacksonian seizures, particularly beginning in the contralateral upper extremity, occur. The nature of the attack is imparted by the characteristics of the seizure propagation, which impart the motor patterns, speech disturbances, and somatosen-

sory symptoms. In the case of seizures involving the paracentral lobule, tonic movements of the ipsilateral foot may occur as well as the expected contralateral leg movements.

#### *Temporal lobe epilepsies*

**Hippocampal (mesiobasal limbic or primary rhinencephalic psychomotor) epilepsy.** This comprises 70–80% of temporal lobe epilepsies and commonly combines with amygdalar epilepsy. Seizures occur in clusters at intervals or randomly; they are complex focal, starting with strange, indescribable feelings, experiential hallucinations or interpretative illusions, followed by arrest (motionless stare), oral, and alimentary automatisms. They last an average of 2 min. GTCS may occur as a consequence of progressing propagation of seizure discharges. Interictal EEG typically shows anterior temporal sharp waves, especially during sleep, while ictal EEG typically shows initial unilateral flattening, especially in temporal lobe. Nonspecific changes of background activity or nonfocal, even nonlateralizing, surface changes may occur, as well as focal, lateralized, or bilateral 4–6 Hz sharp waves. Stereo EEG reveals high-frequency (16–28 Hz) low-voltage spikes building up in one hippocampus propagating to ipsilateral amygdala and cingulate gyrus, but also to contralateral mesiobasal structures. Psychological investigations reveal impaired learning and memory (verbal or nonverbal, depending on dominant or nondominant hemisphere foci). The most common finding is incisural or hippocampal sclerosis. Less frequent causes are gangliogliomas, hamartomas, arteriovenous malformations, astrocytomas, and oligodendrogliomas.

**Amygdalar (anterior polar-amygdalar).** Seizures are characterized by rising epigastric discomfort, nausea, marked autonomic signs, and other symptoms including borborygmi, belching, pallor, fullness of face, flushing of face, arrest of respiration, pupillary dilatation, fear, panic, and olfactory-gustatory hallucinations. Onset of unconsciousness is gradual, followed by staring, oral and alimentary automatisms, and confusion. Association with GTCS of focal onset is less common (about 30%). Rapid eye movement (REM) sleep facilitates amygdalar spiking. Surface EEG findings are similar in hippocampal epilepsy, and stereo EEG reveals high-frequency, low-amplitude 16–28 Hz rhythms in amygdala or amygdala and anterior temporal pole, with spread to hypothalamus, homolateral fronto-orbital regions and hippocampal formation, and to contralateral homologous areas. Common etiologies are gangliogliomas, small gliomas, atypical cell layers in amygdala (and hippocampal formation), anterior temporal pole gliosis, arteriovenous malformations, hamartomas, and traumas with focal gliosis.

**Lateral posterior temporal.** Seizures are characterized by auras of auditory hallucinations, visual percep-

tual hallucinations, or language disorder in case of language-dominant hemisphere focus. These are followed by dysphasia, disturbed orientation or prolonged auditory hallucinations, head movement to one side, and sometimes staring automatisms. Surface EEG shows bilateral midtemporal or posterior temporal spikes, and stereo EEG shows low-frequency rapid spikes (16–28 Hz) building up in the supramarginal angular gyrus and posterior temporal regions. Stepwise involvement of anterior temporal and mesiobasal limbic structures is frequent. The most common etiologies are trauma with focal gliosis, gliomas, arteriovenous malformations, or as sequelae of inflammations or cerebral infarctions.

**Opercular (insular).** Seizures may be characterized either by vestibular or acoustic hallucinations, borborygmi, belching, and autonomic signs, or unilateral face twitching and paresthesia. Olfactory-gustatory hallucinations may occur. EEG and stereo EEG reveal opercular rapid spikes (16–28 Hz), often with minimal spread. Common etiologies are arteriovenous malformations, gliomas, astrocytomas, and venous angioma and scars from cerebral infarction.

#### *Parietal lobe epilepsies*

Seizures are simple partial sensory attacks with many characteristics. Positive phenomena consist of tingling and a feeling of electricity, which may be confined or may spread in a jacksonian manner. There may be a desire to move a body part or a sensation as if a part was being moved. The parts most frequently involved are those with the largest cortical representation, e.g., the hand, arm, and face. There may be tongue sensations of crawling, stiffness, or coldness, and facial sensory phenomena may occur bilaterally. Occasionally an intraabdominal sensation of sinking, choking, or nausea may occur, particularly in cases of inferior and lateral parietal lobe involvement. Rarely, there may be pain, and this may take the form of a superficial burning dysesthesia or a vague, very severe, episodic painful sensation. Parietal lobe visual phenomena may occur as photophasias or as hallucinations of a formed variety, including colors and animal shapes. Metamorphopsia with distortions, foreshortenings, and elongations may occur, and are more frequently seen with nondominant hemisphere discharges. Negative phenomena include numbness, feeling as if a body part were absent, and a loss of awareness of a part or a half of the body, known as asomatognosia. This is particularly the case in right-sided attacks. Severe vertigo may be indicative of suprasylvian parietal lobe seizures. Posterior left parietal seizures result in a variety of receptive or conductive speech disturbances. A rare sensory disturbance with involvement of the paracentral lobule involves both lower extremities. The EEG of parietal lobe epilepsy shows appropriately localized sharp wave discharges. Seizures

of the paracentral lobule area have a tendency to become secondarily generalized.

#### *Occipital lobe epilepsy*

The clinical seizure manifestations usually, but not necessarily, include visual manifestations with no corresponding external stimuli, but not to the exclusion of other manifestations. Elementary visual seizures are characterized by fleeting visual manifestations (paropsia), which may be either negative (scotoma, hemianopsia, amaurosis) or, more commonly, positive (sparks or flashes, phosphenes). Such sensation appears in the visual field contralateral to the discharging lesion in the specific visual cortex, but can spread to the whole visual field. Perceptive illusions, in which the objects appear to be distorted, may occur. The following varieties can be distinguished: polyoptic illusion (monocular diplopia), dysmetropsic illusions (a change in size—macropsia or micropsia, or a change in distance—macroproxiopia or microtelepsia), plagiopsic illusions (inclination of objects in a given plane of space), and dysmorphopsic illusion (distortion of objects) or a sudden change of shape (metamorphopsia). Visual hallucinatory seizures are occasionally characterized by complex visual perceptions, e.g., colorful scenes of varying complexity. In some cases, the scene is distorted or made smaller, and in rare instances, the subject sees his own image (autoscopy). Such illusionary and hallucinatory visual seizures result from epileptic discharge in the temporal-occipital cortex. The initial signs may also include clonic and/or tonic contraversion of eyes and head or eyes only (oculoclonic or oculozytic deviation), palpebral jerks, and forced closure of eyelids. Sensation of ocular oscillation or of the whole body, vertiginous sensation (environment tipping), and tinnitus along with headache or migraine may occur at the onset of seizures. The discharge may spread to the temporal lobe, producing seizure manifestations of either lateral posterior temporal, hippocampal, or amygdalar epilepsies. When the primary focus is located in the supracalcarine area, the discharge can spread forward on the suprasylvian convexity or the mesial surface, mimicking those of parietal lobe or supplementary motor epilepsy. There is an occasional tendency to become secondarily generalized.

#### **Appendix II: Symptomatic generalized epilepsies of specific etiologies**

Only disease in which epileptic seizures are the presenting or a prominent feature are listed here. These diseases often have epileptic pictures that resemble symptomatic generalized epilepsies without specific etiology, appearing at similar ages.

### Malformations

Aicardi syndrome occurs in females, and is noted for retinal lacunae absence of the corpus callosum; infantile spasms with early onset; and often asymmetric, diffuse EEG abnormalities generally asynchronous with suppression bursts and/or atypical hypsarrhythmia.

Lissencephaly-Pachygyria is characterized by facial abnormalities and specific CT scan features, axial hypotonia, and infantile spasms. The EEG shows fast activity of high voltage "alpha-like" patterns without change during wakefulness and sleep.

There is no typical electroclinical pattern for the individual phacomatoses. It must be emphasized that West syndrome is frequent in tuberous sclerosis, and that generalized and partial seizures may follow the otherwise typical course of infantile spasms. Sturge-Weber syndrome is a frequent cause of simple partial seizures followed by hemiparesis.

### Proven or suspected inborn errors of metabolism

**Neonate.** Metabolism errors in the neonate include nonketotic hyperglycinemia and D-glycericacidemia, showing early myoclonic encephalopathy with erratic myoclonus, partial seizures, and suppression-burst EEG patterns.

**Infant.** The classical phenylketonuria can express itself as a West syndrome.

A variant of phenylketonuria with bipterins deficiency causes seizures starting in the second semester of life in infants who have been hypotonic since birth. The seizures are generalized motor seizures associated with erratic myoclonic jerks and oculogyric fits.

Tay Sachs and Sandhoff disease present with acoustic startle or myoclonus in the first months of life, without EEG manifestation. In the second year, myoclonic jerks and erratic partial seizures occur, along with marked slowing of the background rhythms.

Another type of metabolism error is early infantile type of ceroid-lipofuscinosis (Santuavori Haltia Hagberg disease). Massive myoclonus starts between 5 and 18 months, with a highly suggestive EEG pattern of vanishing EEG.

Pyridoxine dependency is manifested by seizures that have no suggestive characteristics, but this condition must always be suspected for therapeutic reasons.

**Child.** Late infantile ceroid-lipofuscinosis (Jansky Bielschowski disease) is characterized by onset between 2 and 4 years of massive myoclonic jerks, atonic, or atstatic seizures. The EEG shows slow background rhythms, multifocal spikes, and characteristic response to intermittent light stimulation (ILS) at a slow rhythm.

An infantile type of Huntington's disease appears after 3 years by a slowing of mental development, followed by dystonia, generalized tonic-clonic seizures, atypical absences, and myoclonias. The EEG shows discharges

of generalized spike-waves and polyspike-waves, with the usual photosensitivity.

**Child and adolescent.** A juvenile form of Gaucher disease is marked by onset around 6–8 years of age, with epileptic seizures of various types, most commonly tonic-clonic or partial motor. The EEG shows progressive deterioration of background activity, abnormal ILS response, diffuse paroxysmal abnormalities, and multifocal abnormalities with a clear posterior predominance.

The juvenile form of ceroid-lipofuscinosis (Spielmeyer-Vogt-Sjogren disease) is characterized by onset between 6 and 8 years of age, and a decrease in visual acuity, slowing of psychomotor development, and the appearance of cerebellar and extrapyramidal signs. After 1–4 years, generalized convulsive seizures and fragmental, segmental and massive myoclonus occur. The EEG shows bursts of slow waves, and slow spikes and waves.

Onset of Lafora disease occurs between 6 and 19 years of age (mean, 11.5 years) and is characterized by generalized clonic, tonic-clonic seizures, with a frequent association of partial seizure of visual type, constant myoclonic syndrome (fragmentary, segmental, and massive myoclonus), and rapidly progressive mental deterioration. The EEG shows discharges of fast spike-waves and polyspike-waves, photosensitivity, deterioration of background activity, and the appearance of multifocal abnormalities, particularly posteriorly. Death occurs an average of 5.5 years after onset.

The so-called degenerative progressive myoclonic epilepsy (Lundborg type) also falls into this category. The only significant well-individualized group is the Finnish type, described by Koskiniema et al. Onset occurs between 8 and 13 years of age, with myoclonus (segmental, fragmentary, and massive) and tonic-clonic generalized seizures, associated cerebellar ataxia, and slowly progressive, but generally mild, mental deterioration. The EEG shows slow abnormalities (theta rhythms and delta rhythms, later), with generalized spike-waves predominantly in frontal area and photosensitivity. Patients survive for 15 years and more.

Dyssynergia cerebellaris myoclonica (DCM) with epilepsy (Ramsay-Hunt syndrome) appears between 6 and 20 years of age (mean, 11 years) with myoclonias or GTCS. The myoclonic syndrome is above all characterized by action and intention myoclonus. The generalized seizures are rare and sensitive to therapy. The mental deterioration, when present, is slow. Most of the neurological manifestations are limited to cerebellar signs. In the EEG, the background activity remains normal, with generalized paroxysmal abnormalities (spikes, spike-waves, and polyspike-waves), and photosensitivity. During REM sleep, rapid polyspikes localized in the central and vertex regions appear.

The clinical picture for the cherry red spot myoclonus syndrome (sialidosis with isolated deficit in neuramini-

dase) is very similar to that of the Ramsay-Hunt syndrome, with myoclonus, photosensitivity, and cerebellar syndrome. Other characteristics include the nearly constant existence of amblyopia and discovery of a cherry red spot on fundoscopic examination. The EEG is similar to that of DCM with the following specific features: the polyspike-wave discharges always correspond to a massive myoclonus and there is no photosensitivity.

A Ramsay-Hunt-like syndrome can also be associated with a mitochondrial myopathy, with abnormalities of lactate and pyruvate metabolism (Fukuhara et al.).

**Adult.** Kuf's Disease is a relatively slow, progressive storage disease with frequent generalized seizures that may be quite intractable. Unlike juvenile storage diseases, the optic fundi may be normal. The main characteristic is an extreme photic sensitivity on slow photic stimulation.

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