Epilepsia, **42**(6):796–803, 2001
Blackwell Science, Inc.

ILAE Commission Report

A Proposed Diagnostic Scheme for People with Epileptic Seizures and with Epilepsy: Report of the ILAE Task Force on Classification and Terminology

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The International League Against Epilepsy (ILAE) made a major contribution when it established standardized classifications and terminology for epileptic seizures and syndromes. This provided a universal vocabulary that not only facilitated communication among clinicians, but also established a taxonomic foundation for performing quantitative clinical and basic research on epilepsy. Much, however, has changed since the adoption of the currently used Classification of Epileptic Seizures in 1981 (1) and the Classification of Epilepsies and Epileptic Syndromes in 1989 (2). Consequently, the Executive Committee of the ILAE, which took office in July 1997, agreed that review and revision of the current classification system would be a priority for this Executive term.

A Task Force on Classification and Terminology was appointed, which divided itself into four working groups concerned with Descriptive Terminology for Ictal Events; Seizures; Syndromes and Diseases; and Impairment. During the course of several meetings and vigorous e-mail discussions, the Task Force agreed that it would not be possible to replace the current international classifications with similar revised and updated classifications that would be universally accepted and meet all the clinical and research needs such a formal organizational system would be expected to provide. Rather, the Task Force is proposing a diagnostic scheme that makes use of standardized terminology and concepts to describe individual patients (Table 1). Within this diagnostic scheme, a variety of approaches to classification are possible, and some are presented here by way of example only. The Task Force views the development of specific classifications as a continuing work in progress. Flexible and dynamic classifications will be revised periodically based not only on rapidly emerging new information, but also on the resolution of problems that will inevitably be identified through use. At this point, the proposal does include several definitive changes in concepts and terminology (Table 2), and classifications are presented as examples of what could be devised in the future.

Accepted March 9, 2001.
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RATIONALE FOR THE PROPOSAL

Although each new ILAE classification has represented considerable effort on the part of acknowledged experts from many different countries, they have always met with a certain degree of resistance from the international epileptology community. This is because, in part, a rigid classification shapes the manner in which future generations of clinical and basic neuroscientists think about epilepsy and epileptic phenomena, thereby influencing (perhaps unduly) clinical practice and research. For instance, in the current Classification of Epileptic Seizures, the division of partial seizures into "simple" and "complex" inappropriately created the impression that impairment of consciousness had certain mechanistic implications related to limbic system involvement. Confusion, and at times vociferous objections, resulted in part from the fact that the 1970 International Classification of Epileptic Seizures had used the term "complex partial seizures" synonymously with "temporal lobe seizures" (3). Over the past two decades, detailed investigations of the anatomic substrates of ictal semiology, based largely on work carried out in epilepsy surgery centers, have strongly suggested that fundamental mechanisms of certain limbic seizures are different from those of neocortical seizures, and that both can be associated with impairment of consciousness or not. Consequently, the designation of partial seizures as "simple," or "complex," has in the process lost meaningful precision. Indeed, the 1981 Classification of Epileptic Sei-

TABLE 1. *Proposed diagnostic scheme for people with epileptic seizures and with epilepsy*

Epileptic seizures and epilepsy syndromes are to be described and categorized according to a system that uses standardized terminology, and that is sufficiently flexible to take into account the following practical and dynamic aspects of epilepsy diagnosis:

1. Some patients cannot be given a recognized syndromic diagnosis.
2. Seizure types and syndromes change as new information is obtained.
3. Complete and detailed descriptions of ictal phenomenology are not always necessary.
4. Multiple classification schemes can, and should, be designed for specific purposes (e.g., communication and teaching; therapeutic trials; epidemiologic investigations; selection of surgical candidates; basic research; genetic characterizations).

This diagnostic scheme is divided into five parts, or Axes, organized to facilitate a logical clinical approach to the development of hypotheses necessary to determine the diagnostic studies and therapeutic strategies to be undertaken in individual patients:

Axis 1: Ictal phenomenology, from the Glossary of Descriptive Ictal Terminology, can be used to describe ictal events with any degree of detail needed.

Axis 2: Seizure type, from the List of Epileptic Seizures. Localization within the brain and precipitating stimuli for reflex seizures should be specified when appropriate.

Axis 3: Syndrome, from the List of Epilepsy Syndromes, with the understanding that a syndromic diagnosis may not always be possible.

Axis 4: Etiology, from a Classification of Diseases Frequently Associated with Epileptic Seizures or Epilepsy Syndromes when possible, genetic defects, or specific pathologic substrates for symptomatic focal epilepsies.

Axis 5: Impairment, this optional, but often useful, additional diagnostic parameter can be derived from an impairment classification adapted from the WHO ICIDH-2.

TABLE 2. *Definitions of key terms*

Epileptic seizure type: An ictal event believed to represent a unique pathophysiologic mechanism and anatomic substrate. This is a diagnostic entity with etiologic, therapeutic, and prognostic implications. (new concept)

Epilepsy syndrome: A complex of signs and symptoms that define a unique epilepsy condition. This must involve more than just the seizure type: thus frontal lobe seizures *per se*, for instance, do not constitute a syndrome. (changed concept)

Epileptic disease: A pathologic condition with a single specific, well-defined etiology. Thus progressive myoclonus epilepsy is a syndrome, but Unverricht–Lundborg is a disease. (new concept)

Epileptic encephalopathy: A condition in which the epileptiform abnormalities themselves are believed to contribute to the progressive disturbance in cerebral function. (new concept)

Benign epilepsy syndrome: A syndrome characterized by epileptic seizures that are easily treated, or require no treatment, and remit without sequelae. (clarified concept)

Reflex epilepsy syndrome: A syndrome in which all epileptic seizures are precipitated by sensory stimuli. Reflex seizures that occur in focal and generalized epilepsy syndromes that also are associated with spontaneous seizures are listed as seizure types. Isolated reflex seizures also can occur in situations that do not necessarily require a diagnosis of epilepsy. Seizures precipitated by other special circumstances, such as fever or alcohol withdrawal, are not reflex seizures. (changed concept)

Focal seizures and syndromes: Replaces the terms partial seizures and localization-related syndromes. (changed terms)

Simple and complex partial epileptic seizures: These terms are no longer recommended, nor will they be replaced. Ictal impairment of consciousness will be described when appropriate for individual seizures, but will not be used to classify specific seizure types. (new concept)

Idiopathic epilepsy syndrome: A syndrome that is only epilepsy, with no underlying structural brain lesion or other neurologic signs or symptoms. These are presumed to be genetic and are usually age dependent. (unchanged term)

Symptomatic epilepsy syndrome: A syndrome in which the epileptic seizures are the result of one or more identifiable structural lesions of the brain. (unchanged term)

Probably symptomatic epilepsy syndrome: Synonymous with, but preferred to, the term cryptogenic, used to define syndromes that are believed to be symptomatic, but no etiology has been identified. (new term)

zures was purposely based purely on ictal phenomenology and associated EEG findings rather than anatomic substrates and pathophysiologic mechanisms, because insufficient information was available at the time to permit the authors to do otherwise. It is the belief of the Task Force that adequate evidence now exists to permit creation of a list of seizure types that represent diagnostic entities, as opposed to phenomenologic descriptions, based on known or presumed common anatomy and pathophysiology. Such diagnostic entities would, like syndromes, have etiologic, therapeutic, and prognostic implications, and could be used to supplement syndromic diagnoses, or stand alone when syndromic diagnoses cannot be made.

The 1981 Classification of Epileptic Seizures also has been criticized because it is not purely semiologic; post hoc etiologic information and EEG data are often required to use it properly, and the dichotomy of "partial" versus "generalized" belies a need to avoid anatomic implications. The Task Force believes that a purely descriptive phenomenologic approach to defining ictal semiology has definite clinical value, and the new diagnostic scheme proposed here includes a modification of a previously proposed classification of ictal phenomenology (4), as an option that can be used in detail where appropriate.

Similarly, the previous dichotomous classifications based on concepts of "partial" or "localization related" versus "generalized" abnormalities created the false impression that epileptic seizures, or epilepsy syndromes, were due to either localized disturbances in one hemisphere or disturbances involving the entire brain. A variety of conditions between focal and generalized epileptogenic dysfunctions include diffuse hemispheric abnormalities, multifocal abnormalities, and bilaterally symmetrical localized abnormalities. Although concepts of partial and generalized epileptogenicity have value, perhaps more with respect to ictal events than to syndromes, it is neither appropriate nor useful to attempt to contain all seizures and syndromes within one or the other of these categorizations.

The term partial itself has come under criticism be-

cause it implies part of a seizure, or part of a syndrome, rather than a seizure or syndrome that begins in part of one hemisphere. For this reason, the 1989 Classification of Epilepsies and Epileptic Syndromes replaced the term partial with "localization-related." This latter terminology has been cumbersome and is not consistently used. The Task Force is now proposing that the terms partial and localization-related be replaced with the older term "focal," which remains in common use. It must be strongly emphasized, however, that the term focal does not mean that the epileptogenic region is a small, well-delineated focus of neuronal pathology; focal seizures, as well as focal syndromes, are almost always due to diffuse, and at times widespread, areas of cerebral dysfunction.

Another change in terminology evident in this document is the omission of the words "convulsion" and "convulsive" in the list of epileptic seizure types and epilepsy syndromes. The Task Force thought that these are nonspecific lay terms, and at times improperly used. Consequently it was agreed to be consistent, not only in descriptive ictal terminology, but also in naming epileptic seizure types and syndromes, to avoid these terms. For instance, the Task Force is proposing that the term "febrile convulsions" be replaced by "febrile seizures."

There also has been dissatisfaction with the terms "idiopathic" and "cryptogenic." Problems with the former have resulted from misunderstanding of the correct definition of idiopathic, which means a disorder unto itself, sui generis, and not etiology unknown. Problems with the latter have been due to an imprecision in definition; cryptogenic is usually used to designate conditions that are not idiopathic, or are presumed to be symptomatic, when the etiology has not been determined, but it also is used by some for conditions in which it is not known whether they are idiopathic or symptomatic. The Task Force has been unable to find an acceptable alternative to the term idiopathic, which, when used correctly, confers a useful taxonomic concept. The terms "benign" and "genetic" were discarded because not all idiopathic epilepsies are necessarily benign, and not all genetic epileptic conditions (e.g., the progressive myoclonus epilepsies) are idiopathic. Although the term "essential" also is used in medicine to convey the same meaning, the Task Force believes that most epileptologists have now learned to use the term idiopathic correctly, and that there is value in maintaining continuity. Consequently, it is recommended that the terms idiopathic and symptomatic be retained, but that the term cryptogenic, although still acceptable, be replaced by the more precise term "probably symptomatic." Therefore, some epilepsy syndromes are referred to as either idiopathic or symptomatic, but a dichotomous classification system that attempts to categorize all syndromes in this manner has been avoided.

Another important criticism of previous rigid syndromic classifications has been a failure to recognize the fact that some syndromes are well accepted, whereas others are controversial, or lack sufficient data. Formally recognizing a syndrome by including it in an official international classification may give it inappropriate legitimacy, whereas failing to recognize a syndrome in the official classification can discourage studies that are necessary to lead to its acceptance. Any official ILAE-sanctioned list of epilepsy syndromes must differentiate between universally accepted syndromes and those in development, and must also be sufficiently flexible to permit additions and deletions of syndromes as new information becomes available.

The rapidly moving field of genetics has contributed greatly in recent years to our understanding of many diseases, including some epileptic disorders, but the relationship between genetic disturbances and phenotypic expression remains complicated and poorly understood. Because a single, relatively well defined, idiopathic epilepsy syndrome can be due to more than one genetic abnormality, and different members of a family sharing a common genetic abnormality can have different epilepsy syndromes, it was considered premature to attempt to classify epilepsy syndromes, to any great extent, on the basis of specific genetic etiologies. There is no doubt, however, that in the near future, genetic classifications of certain epilepsy syndromes will become possible, and that these classifications will have considerable clinical value. It will be necessary for such classifications to include syndromes of families, in addition to syndromes of individuals, and indeed the Task Force has included three such conditions in the current recommended list of epilepsy syndromes (cf. Table 4): generalized epilepsy with febrile seizures plus, familial focal epilepsy with variable foci, and idiopathic generalized epilepsies with variable phenotypes. The first two of these are considered to be syndromes in development, and diagnosis would not be possible without evidence of multiple affected family members. The third is a new concept, which remains under discussion.

DESCRIPTION OF THE PROPOSAL

The Task Force is asking the General Assembly to approve a diagnostic scheme, rather than a fixed classification, when it next meets in Buenos Aires in May 2001. This diagnostic scheme is intended to provide the basis for a standardized description of individual patients, and consists of five levels, or Axes (Table 1). The Axes are organized to facilitate a logical clinical approach to the development of hypotheses necessary to determine the diagnostic studies that should be performed, and the therapeutic strategies to be undertaken.

The diagnostic scheme described here will be made up

TABLE 3. *Epileptic seizure types and precipitating stimuli for reflex seizures*

- Self-limited seizure types
  - Generalized seizures
    - Tonic–clonic seizures (includes variations beginning with a clonic or myoclonic phase)
    - Clonic seizures
      - Without tonic features
      - With tonic features
    - Typical absence seizures
    - Atypical absence seizures
    - Myoclonic absence seizures
    - Tonic seizures
    - Spasms
    - Myoclonic seizures
    - Eyelid myoclonia
      - Without absences
      - With absences
    - Myoclonic atonic seizures
    - Negative myoclonus
    - Atonic seizures
    - Reflex seizures in generalized epilepsy syndromes
  - Focal seizures
    - Focal sensory seizures
      - With elementary sensory symptoms (e.g., occipital and parietal lobe seizures)
      - With experiential sensory symptoms (e.g., temporoparietooccipital junction seizures)
    - Focal motor seizures
      - With elementary clonic motor signs
      - With asymmetric tonic motor seizures (e.g., supplementary motor seizures)
      - With typical (temporal lobe) automatisms (e.g., mesial temporal lobe seizures)
      - With hyperkinetic automatisms
      - With focal negative myoclonus
      - With inhibitory motor seizures
    - Gelastic seizures
    - Hemiclonic seizures
    - Secondarily generalized seizures
    - Reflex seizures in focal epilepsy syndromes
- Continuous seizure types
  - Generalized status epilepticus
    - Generalized tonic–clonic status epilepticus
    - Clonic status epilepticus
    - Absence status epilepticus
    - Tonic status epilepticus
    - Myoclonic status epilepticus
  - Focal status epilepticus
    - Epilepsia partialis continua of Kojevnikov
    - Aura continua
    - Limbic status epilepticus (psychomotor status)
    - Hemiconvulsive status with hemiparesis
- Precipitating stimuli for reflex seizures
  - Visual stimuli
    - Flickering light: color to be specified when possible
    - Patterns
    - Other visual stimuli
  - Thinking
  - Music
  - Eating
  - Praxis
  - Somatosensory
  - Proprioceptive
  - Reading
  - Hot water
  - Startle

TABLE 4. *Epilepsy syndromes and related conditions*

- Benign familial neonatal seizures
- Early myoclonic encephalopathy
- Ohtahara syndrome
- [a]Migrating partial seizures of infancy
- West syndrome
- Benign myoclonic epilepsy in infancy
- Benign familial infantile seizures
- Benign infantile seizures (nonfamilial)
- Dravet's syndrome
- HH syndrome
- [a]Myoclonic status in nonprogressive encephalopathies
- Benign childhood epilepsy with centrotemporal spikes
- Early-onset benign childhood occipital epilepsy (Panayiotopoulos type)
- Late-onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Epilepsy with myoclonic–astatic seizures
- Lennox–Gastaut syndrome
- Landau–Kleffner syndrome (LKS)
- Epilepsy with continuous spike-and-waves during slow-wave sleep (other than LKS)
- Childhood absence epilepsy
- Progressive myoclonus epilepsies
- Idiopathic generalized epilepsies with variable phenotypes
  - Juvenile absence epilepsy
  - Juvenile myoclonic epilepsy
  - Epilepsy with generalized tonic–clonic seizures only
- Reflex epilepsies
  - Idiopathic photosensitive occipital lobe epilepsy
  - Other visual sensitive epilepsies
  - Primary reading epilepsy
  - Startle epilepsy
- Autosomal dominant nocturnal frontal lobe epilepsy
- Familial temporal lobe epilepsies
- [a]Generalized epilepsies with febrile seizures plus
- [a]Familial focal epilepsy with variable foci
- Symptomatic (or probably symptomatic) focal epilepsies
  - Limbic epilepsies
    - Mesial temporal lobe epilepsy with hippocampal sclerosis
    - Mesial temporal lobe epilepsy defined by specific etiologies
    - Other types defined by location and etiology
  - Neocortical epilepsies
    - Rasmussen syndrome
    - Other types defined by location and etiology
- Conditions with epileptic seizures that do not require a diagnosis of epilepsy
  - Benign neonatal seizures
  - Febrile seizures
  - Reflex seizures
  - Alcohol-withdrawal seizures
  - Drug or other chemically induced seizures
  - Immediate and early posttraumatic seizures
  - Single seizures or isolated clusters of seizures
  - Rarely repeated seizures (oligoepilepsy)

[a] Syndromes in development.

of flexible and dynamic modules within which the Task Force will make periodic changes and updates as needed, with the approval of the Executive Committee. The Task Force is proposing that this diagnostic scheme include the development of flexible, rather than rigid, classifications, eliminating the need for the General Assembly, which meets only once every 2 years, to agree on every revision. Acceptance of this diagnostic scheme, therefore, does not exclude the creation of various classification systems for seizures and syndromes, or the contin-

TABLE 5. *An example of a classification of epilepsy syndromes*

Groups of syndromes	Specific syndromes
Idiopathic focal epilepsies of infancy and childhood	Benign infantile seizures (nonfamilial)
	Benign childhood epilepsy with centrotemporal spikes
	Early-onset benign childhood occipital epilepsy (Panayiotopoulos type)
	Late-onset childhood occipital epilepsy (Gastaut type)
Familial (autosomal dominant) focal epilepsies	Benign familial neonatal seizures
	Benign familial infantile seizures
	Autosomal dominant nocturnal frontal lobe epilepsy
	Familial temporal lobe epilepsy
	Familial focal epilepsy with variable foci[a]
Symptomatic (or probably symptomatic) focal epilepsies	Limbic epilepsies
	Mesial temporal lobe epilepsy with hippocampal sclerosis
	Mesial temporal lobe epilepsy defined by specific etiologies
	Other types defined by location and etiology
	Neocortical epilepsies
	Rasmussen syndrome
	Hemiconvulsion–hemiplegia syndrome
	Other types defined by location and etiology
	Migrating partial seizures of early infancy[a]
Idiopathic generalized epilepsies	Benign myoclonic epilepsy in infancy
	Epilepsy with myoclonic astatic seizures
	Childhood absence epilepsy
	Epilepsy with myoclonic absences
	Idiopathic generalized epilepsies with variable phenotypes
	Juvenile absence epilepsy
	Juvenile myoclonic epilepsy
	Epilepsy with generalized tonic–clonic seizures only
	Generalized epilepsies with febrile seizures plus[a]
Reflex epilepsies	Idiopathic photosensitive occipital lobe epilepsy
	Other visual sensitive epilepsies
	Primary reading epilepsy
	Startle epilepsy
Epileptic encephalopathies (in which the epileptiform abnormalities may contribute to progressive dysfunction)	Early myoclonic encephalopathy
	Ohtahara syndrome
	West syndrome
	Dravet syndrome (previously known as severe myoclonic epilepsy in infancy)
	Myoclonic status in nonprogressive encephalopathies[a]
	Lennox–Gastaut syndrome
	Landau–Kleffner syndrome
	Epilepsy with continuous spike–waves during slow-wave sleep
Progressive myoclonus epilepsies	See specific diseases
Seizures not necessarily requiring a diagnosis of epilepsy	Benign neonatal seizures
	Febrile seizures
	Reflex seizures
	Alcohol-withdrawal seizures
	Drug or other chemically induced seizures
	Immediate and early posttraumatic seizures
	Single seizures or isolated clusters of seizures
	Rarely repeated seizures (oligoepilepsy)

[a] Syndromes in development.

ued use of some aspects of the current classification. The Task Force will be concerned with the construction of classification systems during the next Executive term, but it is anticipated that seizures and syndromes will not be organized into fixed dichotomous classifications, but rather categorized in various ways for various purposes.

Axis 1 consists of a description of the ictal semiology, using a standardized Glossary of Descriptive Terminology. The description of the ictal event, without reference to etiology, anatomy, or mechanisms, can be very brief or extremely detailed, as required for clinical or research purposes. Although detailed descriptions of the onset and evolution of localized ictal phenomena often are not necessary, they can be useful; for instance, in patients who are candidates for surgical treatment, or for research designed to elucidate the anatomic substrates or pathophysiologic mechanisms underlying specific clinical behaviors. Communication among clinicians, and among researchers, will be greatly enhanced by the establishment of standardized terminology for describing ictal semiology.

Axis 2 is the epileptic seizure type, or types, experienced by the patient, derived from a list of accepted seizure types that represent diagnostic entities with etio-

TABLE 6. *An example of a classification of diseases frequently associated with epileptic seizures or syndromes*

Groups of diseases	Specific diseases
Progressive myoclonic epilepsies	Ceroid lipofuscinosis
	Sialidosis
	Lafora disease
	Unverricht–Lundborg disease
	Neuroaxonal dystrophy
	MERRF
	Dentatorubropallidoluysian atrophy
	Other
Neurocutaneous disorders	Tuberous sclerosis complex
	Neurofibromatosis
	Hypomelanosis of Ito
	Epidermal nevus syndrome
	Sturge–Weber syndrome
Malformations due to abnormal cortical developments	Isolated lissencephaly sequence
	Miller–Dieker syndrome
	X-linked lissencephaly
	Subcortical band heterotopia
	Periventricular nodular heterotopia
	Focal heterotopia
	Hemimegalencephaly
	Bilateral perisylvian syndrome
	Unilateral polymicrogyria
	Schizencephalies
	Focal or multifocal cortical dysplasia
	Microdysgenesis
Other cerebral malformations	Aicardi syndrome
	PEHO syndrome
	Acrocallosal syndrome
	Other
Tumors	DNET
	Gangliocytoma
	Ganglioglioma
	Cavernous angiomas
	Astrocytomas
	Hypothalamic hamartoma (with gelastic seizures)
	Other
Chromosomal abnormalities	Partial monosomy 4P or Wolf–Hirschhorn syndrome
	Trisomy 12p
	Inversion duplication 15 syndrome
	Ring 20 chromosome
	Other
Monogenic mendelian diseases with complex pathogenetic mechanisms	Fragile X syndrome
	Angelman syndrome
	Rett syndrome
	Other
Inherited metabolic disorders	Nonketotic hyperglycinemia
	D-Glyceric acidemia
	Propionic acidemia
	Sulphite-oxidase deficiency
	Fructose 1-6 diphosphatase deficiency
	Other organic acidurias
	Pyridoxine dependency
	Aminoacidopathies (maple syrup urine disease, phenylketonuria, other)
	Urea cycle disorders
	Disorders of carbohydrate metabolism
	Disorders of biotin metabolism
	Disorders of folic acid and B_{12} metabolism
	Glucose transport protein deficiency
	Menkes' disease
	Glycogen-storage disorders
	Krabbe disease
	Fumarase deficiency
	Peroxisomal disorders
	Sanfilippo syndrome
	Mitochondrial diseases (pyruvate dehydrogenase deficiency, respiratory chain defects, MELAS)

TABLE 6. *Continued*

Groups of diseases	Specific diseases
Prenatal or perinatal ischemic or anoxic lesions or cerebral infections causing nonprogressive encephalopathies	Porencephaly
	Periventricular leukomalacia
	Microcephaly
	Cerebral calcifications and other lesions due to toxoplasmosis, CVI, HIV, etc.
Postnatal infections	Cysticercosis
	Herpes encephalitis
	Bacterial meningitis
	Other
Other postnatal factors	Head injury
	Alcohol and drug abuse
	Stroke
	Other
Miscellaneous	Celiac disease (epilepsy with occipital calcifications and celiac disease)
	Northern epilepsy syndrome
	Coffin–Lowry syndrome
	Alzheimer's disease
	Huntington disease
	Alpers' disease

MERRF, myoclonus epilepsy with ragged red fibers; DNET, dysembryoplastic neuroepithelial tumor; MELAS, mitochondrial encephalomyopathy, lactic acidosis, and stroke-like symptoms; CVI, cerebrovascular incident; HIV, human immunodeficiency virus.

logic, therapeutic, and/or prognostic implications. Localization within the brain should be specified when this is appropriate, and in the case of reflex seizures, the specific stimulus also will be specified here. The Task Force has constructed a list of accepted epileptic seizure types, including forms of status epilepticus, and precipitating factors for reflex seizures (Table 3). Seizure types have been divided into self-limited seizures and continuous seizures, and further divided into generalized seizures and focal seizures, but it is anticipated that other approaches to organization, categorization, and classification of seizure types will be devised for specific purposes.

Axis 3 is the syndromic diagnosis derived from a list of accepted epilepsy syndromes (Table 4), although it is understood that a syndromic diagnosis may not always be possible. The recommended list distinguishes between epilepsy syndromes and conditions with epileptic seizures that do not require a diagnosis of epilepsy, and also identifies which syndromes are still in development. It is important to stress that the list shown in Table 4 contains syndromes that are still under discussion, such as the new concept of Idiopathic generalized epilepsies with variable phenotypes, and the reflex epilepsies, and that the Task Force will continue to revise this list based on the results of further deliberations, input from the membership, and new information. As with epileptic seizures, it is anticipated that different approaches to organization, categorization, and classification of epilepsy syndromes will be created for specific purposes. One example of an approach to classification of epilepsy syndromes is shown in Table 5. Whereas this classification system may be easy for epileptologists to understand, a more simplified version will likely be constructed for teaching purposes, and used by primary care physicians, whereas more detailed, or completely different, classification systems might be necessary for epidemiologic studies, clinical drug trials, presurgical evaluation, basic research, and genetic characterizations.

Axis 4 will specify etiology when this is known. The etiology could consist of a specific disease derived from a classification of diseases frequently associated with epileptic seizures or syndromes (Table 6), a genetic defect, or a specific pathologic substrate, for instance for the symptomatic focal epilepsies. The classification of diseases frequently associated with epileptic seizures shown in Table 6 is preliminary and will require considerable effort over the course of the next Executive Term to be made as comprehensive as possible.

Axis 5 is an optional designation of the degree of impairment caused by the epileptic condition. Classification of impairment will be derived from the World Health Organization ICIDH-2 International Classification of Functioning and Disability (5), which is currently in preparation. Modification may be necessary for application to seizure disorders.

The most recent draft of the Glossary of Descriptive Terminology for Ictal Semiology (Axis 1), detailed descriptions of epileptic seizure types (Axis 2), and epileptic syndromes (Axis 3), and the current draft of the classification of the WHO ICIDH-2 (Axis 5) can be viewed on the ILAE classification website http://www.epilepsy.org/ctf. Although the proposal to be put to the ILAE General Assembly in May merely requests approval of the overall diagnostic scheme, with permission to continue to revise and update the details within each Axis in an ongoing, flexible manner, input from our membership now on these details, as well as on the overall scheme, will be most welcome. The Task Force would particularly like to invite comments on some of

the more important remaining problems, including terms for describing ictal impairment of consciousness, the acceptance of idiopathic generalized epilepsies with variable phenotypes as a single syndrome, the inclusion of a category of epileptic encephalopathies, and the proposed categorization of reflex seizures and syndromes (see Classification Task Force Questionnaire). Comments for the Task Force can be directed to the chair by e-mail: engel@ucla.edu, mail, or fax (1-310-206-8461).

Acknowledgment: The work of the Task Force and the Classification website were supported in part by a generous grant from GlaxoSmithKline. The Axis concept was originally suggested by Dr. Jeffrey Nicholl.

NOTE

Members of the task force on classification and terminology include Jerome Engel, Jr., Los Angeles, California, U.S.A.: Chair; Warren Blume, London, Ontario, Canada: Chair, Working Group on Glossary of Descriptive Terminology; Peter Williamson, Lebanon, New Hampshire, U.S.A.: Chair, Working Group on Seizures; Natalio Fejerman, Buenos Aires, Argentina: Chair, Working Group on Syndromes and Diseases; Harry Meinardi, The Hague, Netherlands: Chair, Working Group on Impairment; Jean Aicardi, Paris, France; Frederick Andermann, Montreal, Quebec, Canada; Alexis Arzimanoglou, Paris, France; Giuliano Avanzini, Milan, Italy; Samuel Berkovic, Melbourne, Australia; Carol Camfield, Halifax, Nova Scotia, Canada; Bernardo Dalla Bernardina, Verona, Italy; Charlotte Dravet, Marseille, France; Francois Dubeau, Montreal, Quebec, Canada; Olivier Dulac, Paris, France; Orvar Eeg-Olofsson, Uppsala, Sweden; Robert Fraser, Seattle, Washington, U.S.A.; Renzo Guerrini, London, U.K.; Allen Hauser, New York, New York, U.S.A.; Matilde Leonardi, Geneva, Switzerland; Hans Lüders, Cleveland, Ohio, U.S.A.; Eli M. Mizrahi, Houston, Texas, U.S.A.; Claudio Munari (deceased), Milan, Italy; Jeffrey Noebels, Houston, Texas, U.S.A.; C.P. Panayiotopoulos, London, U.K.; Perrine Plouin, Paris, France; Marieke Reuvekamp-Thuss, Groningen, Netherlands; Masakazu Seino, Shizuoka, Japan; Arthur Sonnen (deceased), Breda, Netherlands; Carlo Alberto Tassinari, Bologna, Italy; Rupprecht Thorbecke, Bielefeld, Germany; David Treiman, New Brunswick, New Jersey, U.S.A.; Walter van Emde Boas, Heemstede, Netherlands; Federico Vigevano, Rome, Italy; Heinz-Gregor Wieser, Zürich, Switzerland; Peter Wolf, Bielefeld, Germany; and Benjamin Zifkin, Montreal, Quebec, Canada.

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(For ILAE Commission Report by Jerome Engel, Jr., pages 796–804.)

CLASSIFICATION TASK FORCE QUESTIONNAIRE

Name (optional) _____

Chapter _____

Specialty (e.g., adult epilepsy, pediatric epilepsy _____

		Yes	No
1. Do you use the current (1981) seizure classification?		☐	☐
2. Do you use the current (1989) syndrome/epilepsy classification?		☐	☐
3. If yes, do you use them for:			
Undergraduate teaching?	☐		
Postgraduate teaching?	☐		
Clinical trials of AEDs?	☐		
Indications of new AEDs?	☐		
Epidemiological studies?	☐		
Scientific communication?	☐		
4. Do you favor a multiple Axes approach to describing individual patients?		☐	☐
5. If an Axes system is adopted, would you prefer (check one):			
Five Axes	☐		
Three axes with the present optional Axes 1 and 5 as appendices?	☐		
6. For seizures and syndromes, do you believe that there should be (check one):			
One classification only?	☐		
Several variations or classifications for different purposes?	☐		
7. Do you believe that classifications should be (check one):			
Fixed and revised by the General Assembly only?	☐		
Flexible and revised by the Executive Committee?	☐		
8. Do you agree with the concept of diagnostic seizure types, in addition to syndromes?		☐	☐
9. Do you agree with the concept of epileptic encephalopathies?		☐	☐
10. Do you agree with the distinction between reflex seizures and reflex epilepsy syndromes?		☐	☐
11. Do you prefer the term (check one):			
Probably symptomatic?	☐		
Cryptogenic?	☐		
12. Do you prefer the term (check one):			
Partial/Localization-related?	☐		
Focal?	☐		
13. Do you agree that seizures should not be primarily classified according to whether consciousness is impaired (complex vs. simple)?		☐	☐
14. Do you agree with the distinction between Epilepsy syndromes and Epilepsy diseases?		☐	☐
15. Do you like the idea of a syndrome of Idiopathic generalized epilepsies with variable phenotypes?		☐	☐
16. Are there seizure types or syndromes listed that you do not believe exist? If yes, specify.		☐	☐

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17. Are there seizure types or syndromes that should be listed but are not? If yes, specify.		☐	☐

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Return to: Jerome Engel, Jr., M.D., Ph.D., UCLA School of Medicine, Department of Neurology, 710 Westwood Plaza, Los Angeles, CA 90095-1769; Fax: (310) 206-8461