CHAPTER 1

Definition (Terminology) and Classification in Epilepsy: A Historical Survey and Current Formulation, with Special Reference to the ILAE

Simon Shorvon
UCL Institute of Neurology, London, UK

Definition (terminology) and its twin, classification, are minefields in the epilepsy theatre of war. Both are topics that only the battle hardy should venture upon. Strong and divergent views are often held, and passions excited, but not much clarity. Nevertheless, definition and classification are the topics of the first chapter of this textbook, for both are essential for communication and precision in clinical practice, and thus both necessarily underpin successful treatment.

In this chapter, various aspects are considered, especially from the perspective of the International League Against Epilepsy (ILAE). A brief overview of the historical evolution of the topic is given, as it is only by understanding this that the current schema becomes comprehensible. Furthermore, as will become obvious, the same issues have periodically bubbled to the surface, a reiteration that could have been avoided with more historical knowledge. The current definitions and classifications are outlined, and I focus here critically also on their more controversial aspects, as these are of the greatest interest, although I recognize that some of my views are not shared.

Definition

Epilepsy and epileptic seizures

It could be argued that the modern definitions of epilepsy and epileptic seizures were developed in the 1870s with the writing of John Hughlings Jackson. With astounding prescience, in 1870 he defined an epileptic seizure as the clinical correlate of 'an occasional, sudden and excessive discharge of grey matter' [1], and in 1873 extended this definition to include reference to cerebral 'grey matter' reflecting the advances made in the new science of cerebral localization: 'Epilepsy is the name for occasional, sudden, excessive, rapid and local discharges of grey matter' [2]. He recognized that the beginning of the seizure (the aura) gives a clue to the seizure focus and thus that seizure semiology should be meticulously analysed. He also devoted pages to a discussion of the nature and pathophysiology of epilepsy, and based his definitions and classification schemes on this. It is, from the perspective of today, quite remarkable to see how his definition of epileptic seizures remains at the core of all schemes and how little it has been changed. His views on classification also remain valid and unchallenged. Indeed, his has been the greatest single individual contribution to the subject.

After Jackson, less influential authors suggested various alternative definitions and terminologies. The advent of electroencephalography (EEG), in the early 1940s, also had a profound effect on seizure definition and classification and the 'electroclinical' approach was born.

By the mid-twentieth century there was a need for standardization. Henri Gastaut, the second great commander of this battle, realized this and, under the auspices of the ILAE, he assumed leadership in this field and crystallized both definition and particularly classification. Over a period of several decades, with the work particularly of Fritz Dreifuss, a definition and classification system evolved which has become universally used, and which had the effect also of catapulting the ILAE into international prominence.

In the 1960s, the World Health Organization (WHO) became involved, concerned that definitions and terminologies in many fields of neurology and psychiatry were confusing and contradictory. WHO set up expert panels in different fields, and Gastaut was asked to chair the panel on epilepsy and to produce a *Dictionary of Epilepsy* (in effect a glossary of terms) which was published eventually in 1973 [3]. The urgency for standardization in the field was recognized and in the introduction to the published dictionary it is stated that: 'The situation has deteriorated with the growth of published information; terms are frequently not defined and may have different meanings for author and reader . . . the need for accuracy and comparability in reporting the primary data is therefore becoming increasingly urgent.' The first draft of the *Dictionary* was prepared by Gastaut and then reviewed several times by small groups of experts (Gastaut in the Chair, with Masland, Pond, Collomb, Saradzisvilli, Broughton, Valasco Suarez, and Wada) and then by a wider group of experts from 16 countries. The final version was produced as a consensus document [3], and therein, for the first time, many of the types of epilepsy and epileptic seizures and other terms related to epilepsy were formally defined.

The urge to tinker with the definitions has proved irresistible to the ILAE and in the last 10 years a series of revised 'official' definitions has been published (Table 1.1) [3,4,5,6]. The first was in 2001,

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Table 1.1 The 'official' definitions of epilepsy and epileptic seizures 1973–2014.

Study	Definition of epilepsy	Definition of epileptic seizure
1973 [3]	A chronic brain disorder of various aetiologies characterized by recurrent seizures due to excessive discharges of cerebral neurons, associated with a variety of clinical and laboratory manifestations	A cerebral attack resulting from an excessive neuronal discharge
2001 [4]	A chronic neurologic condition characterized by recurrent epileptic seizures	A manifestation of epileptic (excessive and/or hypersynchronous), usually self-limited activity of neurons in the brain
2006 [5]	A disorder of the brain characterized by an enduring predisposition to generate epileptic seizures and by the neurobiologic, cognitive, psychological and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure	A transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain
2014 [6]	A disease of the brain defined by any of the following conditions: (1) At least two unprovoked (or reflex) seizures occurring >24 h apart; (2) one unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years; (3) diagnosis of an epilepsy syndrome[a]	

[a]The 2014 taskforce report also added that epilepsy should be considered to have resolved when an individual: (a) exceeds the age of an 'age-dependent epilepsy syndrome' or (b) who have remained seizure-free for the last 10 years and off antiseizure medicines for at least the last 5 years.

a revised glossary of terms was published by the ILAE Taskforce on Classification and Terminology [4], with the stated intention of being 'descriptive and phenomenologic, and providing a standard terminology for health workers to communicate what is observed and what a patient reports during a seizure'. The definitions of epilepsy and epileptic seizures were reformulated. Unfortunately, in 2005, the ILAE again felt it necessary to produce updated definitions, claiming that there was 'little common agreement' on the definitions of epilepsy and seizures, although it is not clear why this was thought. It was also stated that the 2001 glossary definition was 'preliminary', although there was nothing to suggest this in the 2001 publication. Then, in 2014, the same taskforce [6], some might feel rather ingenuously claiming that the 2005 definition of epilepsy was a 'conceptual' definition, decided that this could be 'translated' into a new 'operational definition' (a practical clinical definition) which was to supersede previous efforts. These definitions are shown in Table 1.1 and it is interesting to see how they have evolved. The definition of a seizure has changed little, and indeed is still largely as conceived by Jackson. The definition of epilepsy has evolved, and possibly not for the better, and one wonders really what is the point of these incessant changes.

The current ILAE definition is clear but somewhat unwieldy (and with a verbosity that surprisingly Jackson's definitions did not suffer from!) and, in most practical clinical settings, Gastaut's 1973 formulation is still used and still suffices.

Classification

ILAE classifications of epileptic seizures and epilepsies

As with definition, classification is important for communication and precision in clinical and research practice. A classification scheme is also important for another reason – it provides a framework on which to conceptualize knowledge and research; a poor scheme will potentially lead to unfocused or futile future research. It is therefore a topic of great significance.

Jackson made another extremely important point when he distinguished between what he called a scientific classification and one that he considered 'purely utilitarian' [7]. He used the analogy of a classification of plants. The scientific classification was based on taxonomy and is what a botanist might use, by providing a listing of natural classes for instance of species, genera, phila, and so on. The practical classification, on the other hand, is what a gardener might use and is 'such an arrangement [that] goes by what is most superficial or striking'. The practical classification facilitates the identification and the application of knowledge for utilitarian purposes but, as Jackson pointed out: '[it can] not be trusted as a natural classification. However much of it may be further elaborated, it makes not even an approach to a scientific classification.'

This analogy of the gardener and botanist, and of the utilitarian versus scientific appears in various places in Jackson's oeuvre, and the importance of this distinction was clear to him and to his readers. Classification appeared often in his writings, with detailed and discursive accounts, and Jackson as always attempted to align this with his theories of neurological structure and function, in a manner which the modern reader might profit much from.

ILAE clinical and electroencephalographic classification of epileptic seizures (1964 and 1969/1970)

After Jackson and until the 1940s not much notable development occurred in the field of classification, but this changed with the introduction of the EEG which seemingly offered the potential for a more 'scientific' approach. This stimulated much activity

Table 1.2 The classification scheme of Sir Charles Symonds (1955). A pre-ILAE scheme showing opinion about classification in the 1950s soon after the introduction of EEG.

	Clinical	Anatomical	Physiological	Pathological	Therapeutic
Central epilepsy	Major – generalized Minimal (a) lapses (b) jerks	Central	Bilateral synchronous, symmetrical EEG discharge	Idiopathic (genetic)	— Dione-responsive
Partial epilepsy	Variable, focal onset depending on location	Variable focal	Focal EEG abnormality	Anatomical lesion present	Phenobarbital, diphenyhydantoin

Note the similarity to the initial ILAE classifications. Source: Derived from references Symonds 1955 [8] and Masland 1959 [10].

in the field of classification, with important contributions made by Sir Charles Symonds (a leading Queen Square neurologist) (Table 1.2) [8], Francis McNaughton (ILAE President 1961–1965) [9] and Richard Masland (Director NINDS 1958–1968 and President WFN 1981–1989) [10], for instance, but it was Henri Gastaut (ILAE Secretary General 1965–1969 and President 1969–1973) whose work had the greatest influence. He, with colleagues, proposed to create 'an international classification' because, as he wrote: 'current classifications of epileptic seizures vary considerably, and the need for a standardized and uniform system of grouping is very apparent' [11]. He embarked on a series of classification schemes in the 1960s and 1970s which were to become universally adopted.

In passing, it is interesting to observe the process Gastaut instituted to formulate his classification. He drafted a proposal himself and then gathered together 120 leading figures in a meeting in Marseilles on 1–2 April 1964. They debated for two days, until it seems exhaustion set in. After two days, a new draft was created which was then submitted to a newly formed Commission on Terminology consisting of representatives of the American and European Branches of the ILAE and of representatives of the World Federation of Neurology (WFN) and of the International Federation of Societies for Electroencephalography and Clinical Neurophysiology (IFSECN). This commission met in May 1964 in Heemstede, and Gastaut then published their proposal for an International Classification of Epileptic Seizures in the same year [11]. A copy was sent to all neurologists who were members of a national neurological society and it seems Gastaut received 170 comments. The amended draft was then debated at the Quadrennial ILAE Congress in Vienna in 1965 (held in conjunction with the 8th Congress of Neurology and the 6th International Congress of EEG and Clinical Neurophysiology) and then sent back to the Commission, and with various comments was debated at the ILAE Executive meeting in 1967. A shortened summary form of the classification was also published at the end of the 1964 version, but it was clear that Gastaut disliked this and he suppressed it from later versions (although it was simple to use and rapidly adopted outside specialist epilepsy practice; Table 1.3).

The classification scheme was published in 1969 in a supplement to *Epilepsia* as part of the programme of the 1969 New York conference and then republished in an identical form in *Epilepsia* in 1970 (Table 1.4) [12,13]. It is clear that, despite his protestations of wide consultation, Gastaut did not accept many amendments to his 1964 draft. Intransigence was one of his hallmarks, and he bulldozed the classification through in inimitable fashion. Nevertheless, there were a few significant differences between the 1964 and the final 1969/1970 versions, notably changes to the terminology of absence

Table 1.3 Summary form of ILAE 1964 classification of epileptic seizures.

1. **Partial seizures or seizures beginning locally**
   - A. With elementary symptomatology (motor, sensory or autonomic symptoms)
   - B. With complex symptomatology (automatism, ideational, psychosensory, psychomotor symptoms)
   - C. Generalized seizures with local onset (NB All partial seizures can develop into generalized seizures, sometimes so rapidly that the local features may not be observable)
2. **Generalized seizures or seizures without local onset**
   - A. Absences of differing form and duration, including 'absence status'. Absences may occur alone, or in combination with myoclonic jerks, or with increase or loss of postural tone, or with automatisms
   - B. Generalized convulsive seizures, in the form of tonic, clonic, tonic-clonic and/or myoclonic attacks
3. **Unilateral or predominantly unilateral seizures (tonic and/or clonic) in children**
4. **Erratic seizures in newborn infants**
5. **Unclassified seizures**

Source: Gastaut *et al.* 1964 [11]. Reproduced with permission from John Wiley & Sons.
This summary form of the 1964 classification was disliked by Gastaut and no summary form was included in the 1969/1970 schemes. However, it became popular in non-specialist practice.

Table 1.4 1969/1970 proposal for the classification of epileptic seizures.

Clinical seizure type	Electro-encephalographic seizure type	Electro-encephalographic interictal expression[a]	Anatomical substrate	Etiology	Age
1. Partial seizures or seizures beginning locally					
A. Partial seizures with elementary symptomatology (generally without impairment of consciousness)					
1. With motor symptoms focal motor (without march), including localized epileptic myoclonus Jacksonian versive (generally contraversive) postural somatic inhibitory(?) aphasic (vii) phonatory (vocalization and arrest of speech)	Local contralateral discharge starting over the corresponding area of cortical representation (not always recorded on the scalp	Local contralateral discharges	Various cortical and/or subcortical regions corresponding with functional representation in one hemisphere	Usually related to a wide variety of local brain lesions (cause known, suspected or unknown). Constitutional factors may be important	Possible at all ages but more frequent with increasing age
2. With special sensory or somatosensory symptoms (i) somatosensory (ii) visual (iii) auditory (iv) olfactory (v) gustatory (vi) vertiginous					
3. With autonomic symptoms					
4. Compound forms[b]					
B. Partial seizures with complex symptomatology (generally with impairment of consciousness; may sometimes begin with elementary symptomatology)					
1. With impaired consciousness only	Unilateral or bilateral discharge, diffuse, or focal in temporal or fronto-temporal regions	Unilateral or bilateral, generally asynchronous focus; usually in the temporal region(s)	Usually cortical and/or subcortical temporal or fronto-temporal regions (including rhinencephalic structures), unilateral or bilateral	As above	As above
2. With cognitive symptomatology (i) with dysmnesic disturbances (conscious amnesia, 'déjà vu', 'deja vecu') (ii) with ideational disturbances (including 'forced thinking', dreamy state)					
3. With affective symptomatology					
4. With 'psychosensory' symptomatology (i) illusions (e.g. macropsia), (ii) hallucinations, metamorphopsia)					
5. With 'psychomotor' symptomatology (automatisms)					
6. Compound forms					

Clinical seizure type	Electro-encephalographic seizure type	Electro-encephalographic interictal expression[a]	Anatomical substrate	Etiology	Age
C. Partial seizures secondarily generalized (all forms of partial seizures, with elementary or complex symptomatology, can develop into generalized seizures, sometimes so rapidly that the focal features may be unobservable. These generalized seizures may be symmetrical or asymmetrical, tonic or clonic, but most often tonic–clonic in type)					
	Above discharge becomes secondarily and rapidly generalized			As above	As above
2. Generalized seizures, bilateral symmetrical seizures or seizures without local onset					
Convulsive or non-convulsive symptomatology, without signs referable to a unilateral system localized in one hemisphere	Bilateral, essentially synchronous and symmetrical discharge from the start	Bilateral, essentially synchronous and symmetrical discharge	Unlocalized (? meso-diencephalon)	No cause found or: (i) diffuse or multiple bilateral lesions, and/or: (ii) toxic and/or metabolic disturbances, and/or: (iii) constitutional, often genetic factors (epileptic predisposition)	All ages
1. Absences (a) Simple absences, with impairment of consciousness only (b) Complex absences with other phenomeno associated with impairment of consciousness: (i) with mild clonic components (myoclonic absences) (ii) with increase of postural tone (retropulsive absences) (iii) with diminution or abolition of postural tone (atonic absences) (iv) with automatisms (automatic absences) (v) with autonomic phenomena (e.g. enuretic absences) (vi) as mixed forms	1. With rhythmical 3 c/s spike and wave discharge ('petit mal' or atypical absence)	Spike and waves and/or dpolyspikes and wave discharges	As above	As above (organic etiology is usual	Especially in children
	2. Without 3 c/s spike and wave (variant of 'petit mal' or atypical absence): (i) low-voltage fast activity or rhythmic discharge at 10 or more c/s, or (ii) more or less rhythmic discharge of sharp and slow waves, sometimes asymmetrical	More or less rhythmic discharges of sharp and slow waves, some times asymmetrical	As above	As above (organic etiology is usual; cerebral metabolic disturbances superimposed on previous brain lesion may be important)	Especially in children
			As above	As above	As above
2. Bilateral massive epileptic myoclonus (myoclonic jerks)	Polyspike and waves or, sometimes, spike and waves or sharp and slow waves	Polyspike and waves, or spike and waves sometimes sharp and slow waves	As above	As above	All ages

(continued)

Table 1.4 *(Continued)*

Clinical seizure type	Electro-encephalographic seizure type	Electro-encephalographic interictal expression[a]	Anatomical substrate	Etiology	Age
3. Infantile spasms	Flattening of the hypsarhythmia during the spasm, or exceptionally, more prominent spikes and slow waves	Hypsarhythmia	As above	As above (cerebral metabolic disturbances superimposed on previous brain lesion may be important)	Infants only
4. Clonic seizures	Mixture of fast (10 c/s or more) and slow waves with occasional spike and wave patterns	Spike and waves and/or polyspike and wave discharges	As above	As above	Especially in children
5. Tonic seizures	Low voltage fast activity or a fast rhythm (l0 c/s or more) decreasing in frequency and increasing in amplitude	More or less rhythmic discharges of sharp and slow waves, sometimes asymmetrical	As above	As above (organic etiology is usual)	Especially in children
6. Tonic–clonic seizures ('grand mal' seizures)	Rhythm at 10 or more c/s, decreasing in frequency and increasing in amplitude during the tonic phase, interrupted by slow waves during the clonic phase	Polyspike and waves and/or spike and waves or, sometimes, sharp and slow wave discharges	As above	As above	Less frequent in young children than other forms of generalized seizures. All ages except infancy
7. Atonic seizures sometimes associated with myoclonic jerks					
(a) of very brief duration (epileptic drop attacks)	Polyspike and waves (more waves than in the myoclonic polyspike and wave)	Polyspike and wave	As above	As above (organic etiology is usual)	Especially in children
(b) of longer duration (including atonic absences)	Rhythmic spike and wave (3–1 c/s) or mixture of fast and slow waves with occasional spike and wave patterns	Polyspike and waves and/or spike and waves or, sometimes, sharp and slow wave discharges			
8. Akinetic seizures (loss of movement without atonia)	Rhythmic spike and wave (3–1 c/s) or mixture of fast and slow waves with occasional spike and wave patterns	Polyspike and waves and/or spike and waves or, sometimes, sharp and slow wave discharges	As above	As above	Especially in children

Source: Gastaut 1969 [12]. Reproduced with permission from John Wiley & Sons.

seizures (typical/atypical changed to simple/complex), the inclusion in 1969/1970 of infantile spasms as a generalized seizure type, the absence/presence of alteration of consciousness mentioned with simple/complex partial seizures, and the exclusion of the 1964 categories of erratic neonatal seizures.

The revised classification was presented to the General Assembly of the Quadrennial ILAE Congress in New York in 1969, where it was further discussed. Despite the fact that it seems not to have been formally approved at the New York assembly, the 'seizure type classification', as it became known, was widely adopted, no doubt in large part due to Gastaut's untiring promotion. Moreover, as noted elsewhere [14], the tagging of the classification with the ILAE name was a publicity coup. By virtue of this single act, ILAE became synonymous with professional authority in epilepsy, and this more than any other activity moved ILAE on to the top table in the world of epilepsy.

In the 1969/1970 classification scheme (Table 1.3), seizures were defined in six axes (Gastaut called these criteria): clinical signs, ictal EEG, interictal EEG, anatomy, aetiology and age. In this regard, the ILAE classification was similar to Symonds' classification structure with his five axes: clinical, pathological, anatomical, physiological and therapeutic (Table 1.2). The seizures were subdivided into two fundamental groups in both schemes: partial and generalized seizures in Gastaut's, and partial and central in Symonds'.

Partial seizures were defined by Gastaut as:

> Seizures in which the first clinical changes indicate activation of an anatomical and/or functional system of neurons limited to a part of a single hemisphere; in which the inconsistently present electrographic seizure patterns are restricted, at least at their onset, to one region of the scalp (the area corresponding to the cortical representation of the system involved); and in which the initial neuronal discharge usually originates in a narrowly limited or even quite diffuse cortical (the most accessible and vulnerable) part of such a system.

The partial seizure category also included secondarily generalized seizures which could evolve from either elementary (simple) or complex symptomatology, and the generalized seizures could be symmetrical or asymmetrical, tonic or clonic, but most often tonic–clonic in type.

Generalized seizures were defined in Gastaut's scheme as:

> Seizures in which the clinical features do not include any sign or symptom referable to an anatomical and/or functional system localized in one hemisphere, and usually consist of initial impairment of consciousness, motor changes which are generalized or at least bilateral and more or less symmetrical and may be accompanied by an 'en masse' autonomic discharge; in which the electroencephalographic patterns from the start are bilateral, grossly synchronous and symmetrical over the two hemispheres; and in which the responsible neuronal discharge takes place, if not throughout the entire grey matter, then at least in the greater part of it and simultaneously on both sides.

Here of course, ILAE was simply following the basic concepts, which originated with Jackson [2], that generalized seizures were widely generated (centroencephalic) and partial seizures were 'focal', and followed the same pattern of Symonds [8], McNaughton [9], Masland [10] and Penfield and Jasper [15]. However, the detailed description and the careful definitions in the ILAE scheme distinguished it from the others and resulted in a structure that still stands today.

It should be noted too that Gastaut was careful not to use the term 'focal' to refer to 'partial' seizures as had been the practice up until then, as he realized that a system (a network) involving wide areas of cortex and deep grey matter and other connections underpinned many partial seizures (for a contemporary discussion of this topic see [10]). This distinction has now regrettably been lost again in the recent proposed ILAE revisions.

ILAE clinical and electroencephalographic classification of epileptic seizures (1981)

Interestingly, at the 1969 New York Congress, at which the ILAE classification was debated, there was also a paper on EEG telemetry. Over the next decade, EEG telemetry became very widely available and the findings from telemetry were thought necessary to include in the seizure-type classification. The ILAE Commission on Terminology continued to meet, now chaired Fritz Dreifuss (ILAE Secretary General 1981–1985 and President 1985–1989), and considered its role to be to 'update, amend and improve the classification in the light of the capability afforded by the newer techniques [i.e. telemetry] to study seizures'. In 1981 a revised seizure type classification was published as 'a compromise which represents a synthesis of the efforts of many persons examining hundreds of seizures over many years. This compilation of knowledge has been brought in line with the state-of-the-art technology without extrapolating to what cannot be observed, but cognizant of the evanescence of any living semantic endeavor which must remain subject to continual revision' [16].

This revised 1981 clinical and electroencephalographic classification of epileptic seizures was approved by the ILAE General Assembly of the ILAE in Kyoto in 1981, and remains the classification that is still the most widely accepted today (Table 1.5).

The 1981 classification differed in two major ways from the scheme of 1969/1970. First, the parameters of anatomy, age and aetiology were removed. This was stated to be because 'they were largely based on historical or speculative information rather than information based on direct observation'. The 1981 classification thus became an 'electroclinical classification' restricted entirely to clinical and EEG data (much of it derived from EEG and video telemetry), in other words was a true classification of seizure type (a 'gardener's classification'). It was also termed a semiological classification; misusing the technical linguistic term referring to the meaning of signs and symbols. Second, partial seizures were separated into simple and complex categories depending on whether consciousness was disturbed, and this differed from the usage of simple and complex in the 1964 and 1969/1970 classifications. The categories of generalized seizures also changed.

The 1981 classification has stood the test of time, and has become the lingua franca of epilepsy specialists around the world. It remains the officially recognized classification of epileptic seizures to this day.

ILAE classification of the epilepsies (1969/1970)

Soon after drafting his seizure type classification, Gastaut turned his attention to an altogether more original work, the classification of 'the epilepsies'.

In July 1968, no doubt at Gastaut's instigation, the WHO formally asked its experts working on the dictionary of epilepsy to also produce a classification of epilepsy to accompany the classification of seizures. Gastaut asked the ILAE Commission on Terminology to take on this task, as he had done for the classification of seizures, in time for the New York Congress in September 1969. This was

Table 1.5 The 1981 ILAE classification of seizure type (the currently accepted official seizure type classification).

Clinical seizure type	EEG seizure type	EEG interictal expression
1. Partial (focal, local) seizures		
A. Simple partial seizures (consciousness not impaired)		
1. With motor signs: (a) focal motor without march (b) focal motor with march (c) versive (d) postural (e) phonatory (vocalization or arrest of speech)	Local contralateral discharge starting over the corresponding area of cortical representation (not always recorded on the scalp)	Local contrarateral discharge
2. With somatosensory or special sensory symptoms (simple hallucinations): (a) somatosensory (b) visual (c) auditory (d) olfactory (e) gustatory (f) vertiginous		
3. With autonomic symptoms or signs (including epigastric sensation, pallor, sweating, flushing, piloerection and pupillary dilatation)		
4. With psychic symptoms: (a) dysphasic (b) dysmnesic (e.g. deja vu) (c) cognitive (e.g. dreamy state, distortions of time sense) (d) affective (e.g. fear, anger) (e) Illusions (e.g. macrospsia) (f) structured hallucinations (e.g. music scenes)		
B. Complex partial seizures		
1. Simple partial onset followed by impairment of consciousness (a) simple partial onset followed by impairment of consciousness (b) with automatism	Unilateral or, frequently bilateral discharge, diffuse or focal in temporal or frontotemporal regions	Unilateral or bilateral generally asynchronous focus; usually in the temporal or frontal regions
2. With impairment of consciousness at onset (a) with impairment of consciousness only (b) with automatism		
C. Partial seizures evolving to secondarily generalized seizures (tonic–clonic, tonic or clonic)		
1. Simple partial seizures evolving to generalized seizures 2. Complex partial seizures evolving to generalized seizures 3. Simple partial seizures evolving to complex partial seizures evolving to generalized seizures	Above discharges become secondarily and rapidly generalized	

Clinical seizure type	EEG seizure type	EEG interictal expression
2. Generalized seizures (convulsive and non-convulsive)		
A. Absence seizures 1. Absence seizures (a) impairment of consciousness only (b) with mild clonic compoents (c) with atonic components (d) with tonic components (e) with automatisms (f) with autonomic components) (b through f may be used alone or in combination)	Usually regular and symmetrical 3 Hz but may be 2–4 Hz spike and slow wave complexes and may have multiple spike and slow wave complexes. Abnormalities are bilateral	Background activity usually normal although paroxysmal activity (such as spikes or spike and slow wave complexes) may occur. This activity is usually regular and symmetrical
2. Atypical absence seizures May have (a) changes in tone that are more pronounced than in absence (b) onset and/or offset that is not abrupt)	EEG more heterogeneous; may include irregular spike and slow wave complexes, fast activity or other paroxysmal activity. Abnormalities are bilateral but often irregular and asymmetrical	Background activity abnormal; paroxysmal activity (such as spikes or spike and slow wave complexes) frequently irregular and asymmetrical
B. Myoclonic seizures (single or multiple)	Polyspike and wave, or sometimes spike and wave or sharp and slow waves	Same as ictal
C. Clonic seizures	Fast activity (10 c/s or more) and slow waves; occasional spike and wave patterns	Spike and wave or polyspike and wave discharges
D. Tonic seizures	Low voltage, fast activity or a fast rhythm of 9–10 c/s or more decreasing in frequency and increasing in amplitude	More or less rhythmic discharges of sharp and slow waves, sometimes asymmetrical. Background is often abnormal for age
E. Tonic–clonic seizures	Rhythm at 10 c/s or more decreasing in frequency and increasing in amplitude during tonic phase, interrupted by slow waves during clonic phase	Polyspike and waves or spike and wave, or, sometimes, sharp and slow wave discharges
F. Atonic seizures (astatic seizures)	Polyspikes and wave or flattening or low-voltage fast activity	Polyspikes and slow wave
3. Unclassified epileptic seizures		

Source: International League Against Epilepsy 1981 [16]. Reproduced with permission from John Wiley & Sons.

a nominal request only, for Gastaut had in fact produced a first draft and he circulated this in August and November 1968 to the ILAE Commission members and WHO/IFECN expert panel. The timescale was also ridiculously short, and he again tried to steam-roller his classification through, but on this occasion there was significant opposition which he failed to overcome. One-third of the members he consulted approved the classification (albeit with reservations), one-third objected to the draft and one-third did not respond. Time was running out, and so Gastaut decided to submit his own draft to the New York meeting [17]. It is fairly clear that there was much contention behind the scenes and, in an unprecedented manner, Merlis, the then President of the ILAE, hurriedly convened and chaired his own International Commission for Classification of the Epilepsies, with members from WFN, the World Federation of Neurological Societies (WFNS) and ILAE (including Masland and Gastaut who were both present) a week before the New York meeting. A draft report [18] was produced and presented alongside Gastaut's draft [17] to the New York General Assembly. Bizarrely, a third classification scheme [19] was also produced, by Richard Masland, despite his being a member of the WHO panel and also Merlis' Commission. This too was presented to the New York meeting.

Gastaut absented himself from the New York Congress – extraordinarily, as he was Secretary General at the time. This was said by Merlis to be due to his urgent duties as Rector of the University of Marseilles. What actually transpired between Merlis and Gastaut is not recorded, but at the ILAE General Assembly, described as 'lively', members of the ILAE were invited to send comments on the various drafts. In fact, perhaps not surprisingly, no further progress seems to have been made. Merlis and Gastaut then left the scene

and one suspects there must have been a sense of fatigue with the topic, for not much else happened in relation to the classification of the epilepsies for the next 10 years.

It is interesting to compare all three schemes. In Gastaut's version, the epilepsies were divided into three major categories: generalized, partial and unclassifiable epilepsies. The generalized category was subdivided into primary generalized epilepsies and secondary generalized. Seven criteria (axes) were used to assign to each category: clinical and EEG manifestations; interictal EEG; age of onset; neuropsychiatric change; response to therapy; aetiology; and pathophysiology.

In Merlis' version, which was clearly based on Gastaut's, the epilepsies were categorized into the same three major categories. The generalized category was, however, subdivided into three groups, not two as by Gastaut: primary generalized epilepsies, secondary generalized epilepsies and undetermined generalized epilepsies. The main difference from the two classification schemes were the criteria (axes) used to categorize. In Merlis' scheme there were six, compared with Gastaut's seven: (a) clinical criteria: seizure form; presence of neurological or psychological evidence of brain pathology; age of onset; aetiology; and (b) EEG criteria: interictal; and ictal.

Both schemes of course were very similar in structure and conception to that of the classification of epileptic seizures, with major divisions based on the generalized/partial dichotomy. Terminology was also shared across the two schemes but with different meanings. Thus, the word **primary** used both to refer to aetiology and also to the absence of a focal onset in generalized seizures and the term **secondarily** generalized applied to seizures, and **secondary** generalized to epilepsies (this caused confusion then and still does). The criteria for the two classifications systems were also rather similar. In fact, Merlis' criteria for the classification of the epilepsies were almost identical to the six criteria used in Gastaut's 1964 seizure type classification. This whole episode was chaotic, and with the benefit of hindsight represented a failure to grasp the real differences between a seizure and an epilepsy; this was surely a lost opportunity.

Masland's formulation was somewhat different, and, in my opinion, was superior in some ways to both Merlis' and Gastaut's formulations. He collected together all the terms used for 'epilepsy' that were mentioned in the WHO glossary, and attempted to categorized them under four main headings: aetiology, physiology (his term for seizure type/EEG), anatomy, and age/precipitant/modifying conditions. Aetiology was subdivided into combined generalized epilepsy (in effect primary generalized epilepsy), unknown, metabolic and organic (in effect lesional). Seizure type/EEG was divided into generalized from onset, partial from the start, erratic and unilateral. Anatomy was divided into centroencephalic, multiple or diffuse, and partial. Age/precipitating factors were divided into age, circadian, relation to female hormonal and reflex epilepsy. Sadly, this classification seems never to have been seriously adopted.

ILAE classification of the epilepsies and epileptic syndromes (1985–1989)

After the General Assembly in 1981, Mogens Dam, the new ILAE President, appointed Peter Wolf as Chair of the Commission on Terminology and Classification of the ILAE. Wolf took up the challenge of producing a consensus draft of the classification of the epilepsies. A general framework of a new classification was rapidly agreed upon, with the epilepsies divided into four categories on the basis of two axes: idiopathic/symptomatic and generalized/localizational related. The latter term was introduced to cover all epilepsies with focal seizures including rolandic and other idiopathic childhood epilepsies where no morphological focus exists and where seizures could originate in either hemisphere. This coincided with what has turned out to be the major development in the field, the introduction of the concept of **epilepsy syndromes**. This too was an initiative of the Marseilles school in conjunction with the ILAE Commission.

In 1983, Joseph Roger organized a landmark workshop in Marseilles, at which various epilepsy syndromes were defined and listed, and at which the members of the ILAE Committee on Classification and Terminology were present. The proceedings of the workshop were published in 1985 [20] and over the following years further workshops were held and proceedings published (the publications becoming known as the 'guide bleu' of epilepsy). Syndromes that were found to be supported by sufficiently solid data were fitted into the four-category framework, and the first draft of a new International Classification of the Epilepsies and Epileptic Syndromes was presented to the ILAE General Assembly in Hamburg in 1985 [21]. After further amendments, a final draft was approved at the next meeting in New Delhi in 1989. By then the classification had a third category of 'cryptogenic' cases where evidence for their being either idiopathic or symptomatic was not present. The approved version was published in 1989 (Table 1.6) [22].

In this version, an epileptic syndrome was defined as 'an epileptic disorder characterized by a cluster of signs and symptoms customarily occurring together; these include such items as type of seizure, aetiology, anatomy, precipitating factors, age of onset, severity, chronicity, diurnal and circadian cycling, and sometimes prognosis'. It was pointed out that a syndrome is not the same as a 'disease' in the sense it does not necessarily have a common aetiology and prognosis. It was recognized then, and has become even more apparent now, that many syndromes have multiple potential underlying aetiologies, that some patients evolve from one syndrome to the next, and that many syndromes have poorly defined boundaries. Nevertheless, from a clinical perspective, the concept of the epilepsy syndrome, albeit imperfect, has proved to be a useful way of providing clinical relevant categories.

The 1989 scheme is an improvement on those that Merlis and Gastaut produced in 1970, but still retains the generalized/partial divide and still uses the terms secondary/secondarily generalized. Primary was dropped in favour of idiopathic, which became the accepted and widely used term from then on.

As Fritz Dreifuss was quoted as being fond of saying, 'seizures are to epilepsy as a cough is to pneumonia' [23], and certainly the form of a seizure tells one nothing of aetiology or pathology. However, the fact that epilepsy itself is essentially a 'symptom' of underlying brain dysfunction and not a disease (in the same sense that headache is a symptom and not a disease) is also important to recognize. There is a grey area between what is best considered a seizure and what is best considered as an epilepsy. The relationships between aetiologies and epileptic phenotypes are also highly variable and because of this a highly detailed, intricate classification of epilepsy will almost certainly be futile. Despite this, in recent years there has developed a regrettable tendency in modern epileptology to designate more and more syndromes and to subdivide categories into smaller and smaller units, which has confused the field and has become another flashpoint in the classification wars.

Finally, it must be remembered that the classification of the epilepsies and syndromes – and the classification of seizure type – are both empirical and utilitarian schema of Jackson's 'gardener's' type. They do not aspire to being 'scientific' in the botanical sense, and their value lies largely in their use as a standard lexicon. As such, expectations should not be raised, and their limitations clearly appreciated.

Table 1.6 The 1989 ILAE international classification of epilepsies and epileptic syndromes (this is still the currently accepted classification of the epilepsies and epileptic syndromes).

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

1.1 Idiopathic (with age-related onset)
- Benign childhood epilepsy with centrotemporal spike
- Childhood epilepsy with occipital paroxysms
- Primary reading epilepsy

1.2 Symptomatic epilepsy
- Chronic epilepsia partialis continua of childhood (Kojewnikow syndrome)
- Syndromes characterized by seizures with specific modes of precipitation
- Syndromes based on anatomic localization
  - Temporal lobe
    - Mesiobasal limbic
    - Lateral temporal
  - Frontal lobe
    - Supplementary motor
    - Cingulate
    - Anterior frontopolar
    - Orbitofrontal
    - Dorsolatreral
    - Opercular
  - Parietal lobe
  - Occipital lobe

1.3 Cryptogenic

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 (GTCS) seizures on awakening
- Other generalized idiopathic epilepsies not defined above
- Epilepsies with seizures precipitated by specific modes of activation

2.2 Cryptogenic or symptomatic (in order of age)
- 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 Non-specific aetiology
- Early myoclonic encephalopathy
- Early infantile epileptic encephalopathy with suppression-burst
- Other symptomatic generalized epilepsies not defined above

2.3.2 Specific syndromes
- Epileptic seizure may complicate many disease states. Under this heading are diseases in which seizures are a presenting or predominant feature

3. Epilepsies and syndromes undetermined whether 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)
- Other undetermined epilepsies not defined above

3.2 Without unequivocal generalized or focal features

All cases with generalized tonic–clonic seizures in which clinical and EEG findings do not permit classification as clearly generalized or localization-related such as in many cases of sleep grand mal (GTCS) are considered not to have unequivocal generalized or focal features

(continued)

Table 1.6 *(Continued).*

4. Special syndromes

4.1 Situation-related seizures (Gelegenheitsanfälle)

- Febrile convulsions
- Isolated seizures or isolated status epilepticus
- Seizures occurring only when there is an acute metabolic or toxic event due to factors such as alcohol, drugs, eclampsia, non-ketotic hyperglycaemia

Source: International League Against Epilepsy 1989 [22]. Reproduced with permission from John Wiley & Sons.

ILAE proposals for the classification of epilepsy since 2000

The Commission on Terminology, set up in 1963, was transmuted into the ILAE Commission on Classification and Terminology in 1973 and continued to function as such until 1997. It then became a Taskforce on Classification and Terminology and then was reconstituted as a Commission in 2005 (in ILAE-speak, a taskforce is appointed for a specific purpose and is disbanded when the purpose is achieved; a commission is a standing committee).

The 1997 taskforce was given the goals of (a) re-evaluating the classification schemes, (b) providing a diagnostic scheme for describing individual cases, and (c) producing a glossary of terms.

One of their first actions was the publication of a *Glossary of Terms* in 2001 [4]. This was shorter and more succinct than Gastaut's 1973 *Dictionary* [3], but not as comprehensive. It included a new definition of epilepsy and epileptic seizures (Table 1.1), and these definitions were revised in 2005 [5] and again in 2014 [6].

An important paper was published in 2001 by Engel, on behalf of the ILAE taskforce, on the topic of classification and terminology [24]. It was stated that it was not possible to replace the current international classifications with another that would be universally accepted, and that would meet all the clinical and research needs such a formal organizational system would be expected to provide. Rather, the taskforce proposed that the ILAE should focus upon 'a diagnostic scheme'. This scheme should provide descriptors, which could be used clinically, to define a patient's epilepsy, under five 'axes': ictal phenomenology, seizure type, syndrome, aetiology and impairment. The conception of 'axes' was based on similar work in the field of psychiatry. The paper included then useful updated lists of syndromes and seizure types. It has to be said that the idea of 'axes' was also found in the clinical criteria used in the 1964, 1969/1970, 1981 and 1989 schemes but the difference was that no attempt was being made to merge these into a single framework. This was a significant step, and it prevented the production of what would have been totally unwieldy classification schemes which would have been of no practical value (in fact, the criticism sometimes made about the 1989 classification of the epilepsies).

The next major incursion into classification was in 2006, when the Core Group of the ILAE taskforce published a report into its activities [25]. This Core Group was a working group of senior epileptologists, led by Engel, which included Hans Lüders who disagreed with the report and wished to be dissociated from it. The report described the discussions regarding: 'the feasibility of creating a paradigm shift in our concept of classifications in the field of epilepsy, based on the establishment of measurable objective criteria for recognizing epileptic seizure types and epilepsy syndromes as unique diagnostic entities or natural classes that can be reproducibly distinguished from all other diagnostic entities or natural classes'. The taskforce produced listings of seizure type (Table 1.7) and

Table 1.7 ILAE Core Group 2006 listing of seizure types.

SELF-LIMITED EPILEPTIC SEIZURES

I. Generalized onset

A. Seizures with tonic and/or clonic manifestations
1. Tonic–clonic seizures
2. Clonic seizures
3. Tonic seizures

B. Absences
1. Typical absences
2. Atypical absences
3. Myoclonic absences

C. Myoclonic seizure types
1. Myoclonic seizures
2. Myoclonic astatic seizures
3. Eyelid myoclonia

D. Epileptic spasms

E. Atonic seizures

II. Focal onset (partial)

A. Local
1. Neocortical
   (a) Without local spread
      (i) Focal clonic seizures
      (ii) Focal myoclonic seizures
      (iii) Inhibitory motor seizures
      (iv) Focal sensory seizures with elementary symptoms
      (v) Aphasic seizures
   (b) With local spread
      (i) Jacksonian march seizures
      (ii) Focal (asymmetrical) tonic seizures
      (iii) Focal sensory seizures with experiential symptoms
2. Hippocampal and parahippocampal

B. With ipsilateral propagation to:
1. Neocortical areas (includes hemiclonic seizures)
2. Limbic areas (includes gelastic seizures)

C. With contralateral spread to:
1. Neocortical areas (hyperkinetic seizures)
2. Limbic areas (dyscognitive seizures with or without automatisms [psychomotor])

D. Secondarily generalized
1. Tonic–clonic seizures
2. Absence seizures
3. Epileptic spasms (unverified)

III. Neonatal seizures

STATUS EPILEPTICUS

- **I. Epilepsia partialis continua (EPC)**
  - A. As occurs with Rasmussen syndrome
  - B. As occurs with focal lesions
  - C. As a component of inborn errors of metabolism
- **II. Supplementary motor area (SMA) status epilepticus**
- **III. Aura continua**
- **IV. Dyscognitive focal (psychomotor, complex partial) status epilepticus**
  - A. Mesial temporal
  - B. Neocortical
- **V. Tonic–clonic status epilepticus**
- **VI. Absence status epilepticus**
  - A. Typical and atypical absence status epilepticus
  - B. Myoclonic absence status epilepticus
- **VII. Myoclonic status epilepticus**
- **VIII. Tonic status epilepticus**
- **IX. Subtle status epilepticus**

Source: Engel 2006 [25]. Reproduced with permission from John Wiley & Sons.

of epilepsy syndromes (Table 1.8) which had interesting differences from, and which were generally less comprehensive than the listings in the 2001 taskforce paper. These lists are in wide usage today.

The Core Group also considered that the 1981 classification of epileptic seizure types, and the 1989 classification of epilepsy syndromes and epilepsies were generally accepted and workable and need not be discarded. The Core Group also cautioned that their listings should not be interpreted as a new classification.

In the view of the author, this was an excellent contribution and approach. However, in 2005, a new Commission on Classification and Terminology was constituted in place of the taskforce. This commission lacked the reticence of the earlier taskforce and proposed another approach in a new report published in 2010. This report has not been a success, and has introduced into the field an increasing sense of contention and confusion [26,27].

The 2010 report reiterated the opinion of the 2001 taskforce and 2006 Core Group that a new classification is not possible, but in its place saw it necessary to provide new 'terminology and concepts that better reflect the current understanding of these issues' [26]. In relation to seizure type, the 2010 Commission suggested the following changes to the 1981 scheme and also to the Core Group's 2006 scheme (Table 1.9):

1. Neonatal seizures are no longer regarded as a separate entity. Seizures in neonates can be classified within the proposed scheme.
2. The previous subclassification of absence seizures has been simplified and altered. Myoclonic absence seizures and eyelid myoclonia are now recognized as seizure types within the category of absence seizures.
3. Epileptic spasms were included in the list of seizure types.
4. In relation to focal seizures, the distinction between the different types (e.g. complex partial and simple partial) was eliminated.
5. Myoclonic atonic (previously called 'myoclonic astatic') seizures are now recognized.

Table 1.8 ILAE Core Group 2005 listing of epilepsy syndromes by age of onset and related conditions.

Neonatal period
- Benign familial neonatal seizures (BFNS)
- Early myoclonic encephalopathy (EME)
- Ohtahara syndrome

Infancy
- Migrating partial seizures of infancy
- West syndrome
- Myoclonic epilepsy in infancy (MEI)
- Benign infantile seizures
- Dravet syndrome
- Myoclonic encephalopathy in non-progressive disorders

Childhood
- Early-onset benign childhood occipital epilepsy (Panayiotopoulos type)
- Epilepsy with myoclonic astatic seizures
- Benign childhood epilepsy with centrotemporal spikes (BCECTS)
- Late-onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Lennox–Gastaut syndrome (LGS)
- Epileptic encephalopathy with continuous spike and wave during sleep (CSWS) including Landau–Kleffner syndrome (LKS)
- Childhood absence epilepsy (CAE)

Adolescence
- Juvenile absence epilepsy (JAE)
- Juvenile myoclonic epilepsy (JME)
- Progressive myoclonus epilepsies (PME)

Less specific age relationship
- Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)
- Familial temporal lobe epilepsies
- Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
- Rasmussen syndrome
- Gelastic seizures with hypothalamic hamartoma

Special epilepsy conditions
- Symptomatic focal epilepsies not otherwise specified
- Epilepsy with generalized tonic–clonic seizures only
- Reflex epilepsies
- Febrile seizures plus (FS+)
- Familial focal epilepsy with variable foci

Conditions with epileptic seizures that do not require a diagnosis of epilepsy
- Benign neonatal seizures (BNS)
- Febrile seizures (FS)

Source: Engel 2006 [25]. Reproduced with permission from John Wiley & Sons.

Some of these changes were widely agreed upon, although the thorny question of the subdivision of focal seizures (which has remained unresolved for years) caused an outburst of protest and was subsequently retracted.

The list of electroclinical syndromes ('and other epilepsies') (Table 1.10) was not altered in any substantial way from the 2001 and 2006 (or indeed the 1989) listings, although name changes were

Table 1.9 The 2010 ILAE Commission proposal for the listing of seizure type.

Generalized seizures
Tonic–clonic (in any combination)
Absence
  Typical
  Atypical
  Absence with special features
  Myoclonic absence
  Eyelid myoclonia
Myoclonic
  Myoclonic atonic
  Myoclonic tonic
Clonic
Tonic
Atonic

Focal seizures

Unknown

Epileptic spasms

Source: Berg *et al.* 2010 [26]. Reproduced with permission from John Wiley & Sons.

made. However, the concept of 'constellations' was added and also a (totally inadequate) listing of 'structural/metabolic' conditions which blurs the distinction between aetiology and syndrome and also caused confusion.

The least satisfactory aspect of this report (at least in this author's opinion) was the change in terminologies [28]. New terms are needed only when there are significant changes in meaning or concept, or when the old terminology is deficient in some way, but change should not be made for its own sake. However, this is exactly what the report embarked upon. The substitution of the aetiological terms **idiopathic, symptomatic and cryptogenic** with **genetic, structural/metabolic and unknown** were particularly egregious examples (thus, the syndrome Idiopathic Generalized Epilepsy was changed to Genetic Generalized Epilepsy).

The substitution of 'genetic' for 'idiopathic' is not only unnecessary but also misleading. The genetic basis of the great majority of idiopathic epilepsies is not known, and these epilepsies are likely to have multifactorial causal influences encompassing environmental, developmental, provoking and genetic factors. It is a simplification to label these as just 'genetic' (and in this sense, everything we are or do is 'genetic' including our opinions, our physical characteristics, our intelligence, and so on). The term 'idiopathic' implies a wider and more complex scope incorporating genetic factors, epigenetic

Table 1.10 The 2010 ILAE Commission proposal for the listing of electroclinical syndromes and other epilepsies arranged by age at onset.

Neonatal period
Benign familial neonatal epilepsy (BFNE)
Early myoclonic encephalopathy (EME)
Ohtahara syndrome

Infancy
Epilepsy of infancy with migrating focal seizures
West syndrome
Myoclonic epilepsy in infancy (MEI)
Benign infantile epilepsy
Benign familial infantile epilepsy
Dravet syndrome
Myoclonic encephalopathy in non-progressive disorders

Childhood
Febrile seizures plus (FS+) (can start in infancy)
Panayiotopoulos syndrome
Epilepsy with myoclonic atonic (previously astatic) seizures
Benign epilepsy with centrotemporal spikes (BECTS)
Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)
Late-onset childhood occipital epilepsy (Gastaut type)
Epilepsy with myoclonic absences
Lennox–Gastaut syndrome
Epileptic encephalopathy with continuous spike–wave during sleep (CSWS)
Landau–Kleffner syndrome (LKS)
Childhood absence epilepsy (CAE)

Adolescence to adult
Juvenile absence epilepsy (JAE)
Juvenile myoclonic epilepsy (JME)
Epilepsy with generalized tonic–clonic seizures alone
Progressive myoclonus epilepsies (PME)
Autosomal dominant epilepsy with auditory features (ADEAF)
Other familial temporal lobe epilepsies

Less specific age relationship
Familial focal epilepsy with variable foci (childhood to adult)
Reflex epilepsies

Distinctive constellations
Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
Rasmussen syndrome
Gelastic seizures with hypothalamic hamartoma
Hemiconvulsion–hemiplegia–epilepsy
Epilepsies that *do not* fit into any of these diagnostic categories can be distinguished first on the basis of the presence or absence of a known structural or metabolic condition (presumed cause) and then on the basis of the primary mode of seizure onset (generalized versus focal)

Epilepsies attributed to and organized by structural–metabolic causes
Malformations of cortical development (hemimegalencephaly, heterotopias, etc.)
Neurocutaneous syndromes (tuberous sclerosis complex, Sturge–Weber, etc.)
Tumour
Infection
Trauma
Angioma
Perinatal insults
Stroke

Epilepsies of unknown cause
Conditions with epileptic seizures that are traditionally not diagnosed as a form of epilepsy per se
Benign neonatal seizures (BNS)
Febrile seizures (FS)

Source: Berg *et al.* 2010 [26]. Reproduced with permission from John Wiley & Sons.

factors, epistatic factors, the influence of the temporal aspects of cerebral development and the (probably great) influence of chance.

The substitution of 'unknown' for 'cryptogenic' creates no new concept but appears wholly to be a change for change's sake.

Replacing 'symptomatic', a universally understood term used throughout medicine, with 'structural/metabolic' is clumsy linguistically, and ignored the many symptomatic epilepsies that have no macroscopic structural or measurable metabolic change, such as those resulting from immunological, inflammatory, degenerative, toxic or biochemical causes. This deficiency was recognized in the most recent pronouncements from the Commission which widen the categories further; although simply returning to the term 'symptomatic' would be simpler. This would not matter so much if there were not disadvantages to changing terminologies:

1 The fact that new terminologies are potentially confusing to the wider medical community, especially those not intimately involved in classification (including general neurologists, paediatricians, general practitioners).
2 There are consequences to changing terms in non-medical arenas (for instance, the press, the courts, social services) where guidelines, regulatory definitions and case law will become redundant and need a long process of revision.
3 There are social consequences to introducing new terms. An example is the substitution of 'genetic' for 'idiopathic' – which will cause difficulties for patients in countries where the term 'genetic' can be highly stigmatizing (for instance, for marriage prospects in many countries).
4 Changing terminology can also lessen the credibility and authority of any classification scheme. New terms used without any strong intellectual underpinning tend to fall out of usage quickly, causing further confusion.
5 The purpose of classification is to facilitate research, diagnosis, investigation and treatment. The crucial test of any change in terminology is the extent to which this facilitation has been achieved, and to which the terms change the mindset and framework of clinical or research effort. The substitution of the term 'idiopathic' with 'genetic', for instance, has the reverse effect, and the change of 'cryptogenic' to 'unknown' and 'symptomatic' to 'structural/metabolic' has no benefit.

What was also confusing was the publication of articles entitled 'Revised classification of seizures and epilepsy' (given that the scheme was not intended to be a 'classification') [26], and their statement that the 'classification structure has been formally abandoned' as there was not an adequate knowledge base to propose a new classification (in the sense of organization) of epilepsies [27].

Attempts to explain the changes have not helped. The 2010 paper ends with the obscure proposal:

> The various forms of epilepsy (at all levels of specificity) will be organized according to those dimensions that are most relevant to a specific purpose. These may be comparable to those in the 1989 classification (seizure onset, 'aetiology,' and age at onset), a different hierarchical arrangement of these same dimensions, a more detailed version of these dimensions, or by entirely different dimensions as needed. [26]

A version written in plain English would have been much more helpful.

The Commission was reconstituted in 2013 and currently is working to revise the 2010 paper with a different approach, and, it is to be hoped the new Commission will make it more meaningful and intelligible. A diagnostic manual is being developed which should be helpful (one of the original objectives of the 1997 taskforce). The concepts of generalized and focal epilepsies, which were removed in the 2010 classification, have been reinstated, and aetiological categories expanded to incorporate genetic, structural, metabolic, immune, infectious and unknown. The disadvantages of changing terminology, however, were unfortunately not addressed.

It is now quite unclear, at least to this author, whether there is intended to be a 'classification of epilepsy' now or not. Quite why it was thought necessary to alter the status of the 1981 and 1989 classification schemes is also unclear, especially as it was the stated view that knowledge was not advanced enough to do so. The provision of a diagnostic manual and of lists for various 'axes' has been very beneficial, but much confusion has been caused in relation to classification and especially terminology. Furthermore, the efforts in the past 5 years have become mired in politics, personal vanities and promotional activities, none of which assist in clinical or scientific goals (and as such probably mirror the situation in 1970).

The future of classification schemes of epilepsy

We all aspire to a classification that is scientifically meaningful – in Jackson's words, a botanist's not a gardener's classification – where the categorization is on the basis of pathophysiology, neurochemical systems, or physiological or anatomical networks. This would be the 'paradigm shift' sought by the Core Group. We are a long way from this goal. Interestingly, similar considerations are current in the field of psychiatry as a reaction about the Diagnostic and Statistical Manual of Mental Disorders (DSM) categories (nothing could be more 'gardening' in nature than this). We are not there yet, and it is to be hoped that by the time of the next edition, the new taskforce will have revised the current position and will have settled upon a system that is more scientifically based.

Classification by aetiology

With the advent of MRI scanning and also more sophisticated biochemical and genetic screening, uncovering the underlying aetiology of an epilepsy has become increasingly feasible [36,37]. Aetiology often dictates prognosis, severity, response to treatment and other clinical features and so is of fundamental importance in any scientific or utilitarian classification (actually, in many ways more so than seizure type). For this reason, the possibility of classifying epilepsy primarily by aetiology is of obvious interest. However, assigning a cause to epilepsy is not straightforward for a number of reasons, limiting the usefulness of any aetiological scheme. These reasons include the following points (for more detailed discussion see [38]).

The cause of epilepsy is often multifactorial Epilepsy is, in the majority of cases, the combined result of genetic and acquired influences, provoking factors and also of the vicissitudes of the process of development (especially in the idiopathic epilepsies).

Problems in assigning cause arise particularly in 'idiopathic epilepsy' where there is no obvious genetic cause, as is the case in the great majority of patients with idiopathic generalized epilepsy for instance. Although this condition is likely to have a genetic predisposition, the phenotype probably depends as much on developmental factors as on genetic predisposition (the analogy is a person's height – there are a variety of known genetic factors, but of equal importance is nutrition, physical environmental factors, psychical stress during development, and also chance). The influence of any individual genes or genetic mechanism (sometimes called

susceptibility) is, in the majority of idiopathic cases, likely to be relatively small. Indeed, in the vast majority of idiopathic cases to date, no genetic susceptibilities at all have been identified. In a small proportion of cases, a mutation with a large effect is present, but even here, there are often major genetic or environmental modifiers influencing the phenotype.

In view of the multifactorial nature of epilepsy, it is useful to consider aetiologies as 'causal factors' rather than as 'causes', and to assign aetiology using 'odds ratios' statistics. The estimation of the odds ratio of any particular causal factor gives it a weighting of its importance. Thus, open head injury as a causal factor of epilepsy has a high odds ratio – and it can be considered statistically likely that this 'cause' contributes a great deal to the development of epilepsy, whereas a mild head injury has a low odds ratio, and can be considered not likely to contribute a major susceptibility. The problem with this approach is that it has a statistical basis, derived at population level, and for any individual patient the weighting may depend on many other individual factors. In some patients, a minor head injury may have greater significance than the population odds ratio might suggest.

Cause versus causal mechanism ('remote' versus 'proximate' cause) A powerful way of classifying cause would be to do so according to the mechanisms by which a remote cause results in a seizure (i.e. by the causal mechanism of a seizure). For instance, trauma and tumour might cause seizures by membrane effects or by the deposition of haematin – and in this sense the membrane dysfunction or haematin deposition is the immediate (proximate) 'cause' not the trauma/tumour (which is the remote cause). This distinction was first pointed out by Jackson. He postulated that classification by proximate cause would allow a 'botanical' rather than a 'gardening' approach. Unfortunately, in many instances, knowledge is not sufficiently advanced to attempt this. As most future advances in the field of causation of epilepsy are likely to be in the field of molecular science, it may be that a classification based on molecular mechanisms will prove possible in the future. This would lead to a radically different approach to classification, and is the sort of paradigm shift that epilepsy classification is sorely in need of.

Epilepsy is a process, and the 'cause' of seizures differs in new-onset and established epilepsy There is considerable evidence of molecular changes occurring after the onset of epilepsy, which may in themselves contribute to the evolution of a new-onset epilepsy into an established, or chronic epilepsy. In such cases, the internal process can be considered at least in part as the 'cause' of the chronic epilepsy. The molecular nature of these processes at present is not clearly understood.

A related issue is the differentiation of 'early' and 'late' seizures after acute cerebral injury (e.g. trauma, stroke). These two types of seizures are physiologically and clinically different. There is also often a latent period which can extend for months or even years between the acute insult (e.g. a head injury) and the onset of late seizures and this is further evidence of a prolonged process of epileptogenesis, and which might continue further after epilepsy has developed. The physiological bases of this process are not known.

Role of investigation in defining the range of causes The identification of cause in any individual of course depends on how thoroughly investigations have been carried out. The range of 'causes' identified in clinical practice alters when new investigatory modalities become available, as happened for instance with EEG, neuroimaging, clinical chemistry and molecular histochemistry. It is probably true to say now that all or almost all of the causal conditions of epilepsy that cause gross structural change, and all monogenic conditions, have been identified, but the mechanisms of genetic influences and the proximate molecular mechanisms remain to be discovered. A new molecular or clinical genetic investigatory modality might have a big impact here in the future.

It should also be noted that not all 'symptomatic' epilepsy is 'acquired'. There are many congenital and innate causes of epilepsy that are developmental or genetic in origin and yet which belong in the 'symptomatic' category. These include for instance cortical dysplasias, neurocutaneous syndromes (e.g. tuberous sclerosis), monogenic diseases (e.g. Rett syndrome, Angelman syndrome), chromosomal disorders (e.g. ring chromosome 20 syndrome) and progressive myoclonic epilepsies (e.g. mitochondrial disease or the neuronal lipofuscinoses).

A final difficulty in any aetiological classification is the nosological position of provoked epilepsy. Provoking factors are often ignored in a classification of cause. However, if for instance a person with idiopathic generalized epilepsy has awakening seizures only after lack of sleep and not at other times, is it not logical to consider the presence of the provoking factor as at least as important in terms of susceptibility as the putative genetic or congenital basis?

With these caveats in mind, a listing of causes (or more accurately causal factors) has been attempted, and a classification of aetiologies is shown in Table 1.11. This ignores seizure type and syndrome altogether, and the mapping across of aetiology/syndrome/seizure type is inexact. A book also cataloguing all the causes of epilepsy (or at least the 'remote' causes) has also been published [39].

Classification by semiology and anatomical site

One other major insight of Jackson (and his colleagues Sir David Ferrier and Sir Victor Horsley) in the 1860s was the recognition that the clinical form of an epileptic seizure gives a clue to the position of the epileptic focus in the brain. Jackson realized that: (i) some brain functions were localized within the cerebral cortex; (ii) a seizure discharge in these areas will produce symptoms of 'overactivity' of these functions; and (iii) the analysis of the symptoms occurring in the initial phase of a seizures (before seizure spread had occurred) would allow the location of the epileptic focus to be surmised and thus provide a target for surgical intervention. This logic remains the basis for all clinical localization for epilepsy surgery to this day. This was also Jackson's reason for focusing on the seizure type imprecisely now named semiology) as a basis for categorizing an individual epilepsy, and again this focus has remained to this day. As Jackson wrote in 1868, 'One of the most important questions we can ask an epileptic patient is "How does the fit begin?"', and, in 1873, 'There is nothing more important than to note where a convulsion begins, for the inference is, that the first motor symptom is the sign of the beginning of the central discharge' and 'The mode of onset is the most important matter in the anatomical investigation of any case of epilepsy.'

In 1993, Lüders *et al.* published a preliminary paper concerning a new approach to classification, which they named a 'semiological classification scheme', and in 1998 presented this in its final form (Table 1.12) [40]. This was based on a detailed analysis of the ictal clinical features, and was proposed as an alternative to the ILAE-sanctioned electroclinical schemes. The rational is that the ictal/interictal EEG, which is an integral part of the ILAE system, correlates poorly with clinical features. The semiological classification stresses that EEG, and other investigatory modalities such as

Table 1.11 Aetiological classification of epilepsy (currently accepted listing of causes of epilepsy).

Main category	Subcategory	Examples[a]
Idiopathic epilepsy	Pure epilepsies due to single gene disorders	Benign familial neonatal convulsions; autosomal dominant nocturnal frontal lobe epilepsy; generalized epilepsy with febrile seizures plus; severe myoclonic epilepsy of childhood; benign adult familial myoclonic epilepsy
	Pure epilepsies with complex inheritance	Idiopathic generalized epilepsy (and its subtypes); benign partial epilepsies of childhood
Symptomatic epilepsy – predominately genetic or developmental causation	Childhood epilepsy syndromes	West syndrome; Lennox–Gastaut syndrome
	Progressive myoclonic epilepsies	Unverricht–Lundborg disease; Dentato-rubro-pallido-luysian atrophy; Lafora body disease; mitochondrial cytopathy; sialidosis; neuronal ceroid lipofuscinosis; myoclonus renal failure syndrome
	Neurocutaneous syndromes	Tuberous sclerosis; neurofibromatosis; Sturge–Weber syndrome
	Other neurological single gene disorders	Angelman syndrome; lysosomal disorders; neuroacanthocytosis; organic acidurias and peroxisomal disorders; prophyria; pyridoxine-dependent epilepsy; Rett syndrome; urea cycle disorders; Wilson disease; disorders of cobalamin and folate metabolism
	Disorders of chromosome function	Down syndrome; fragile X syndrome; 4p-syndrome; isodicentric chromosome 15; ring chromosome 20
	Developmental anomalies of cerebral structure	Hemimegalencephaly; focal cortical dysplasia; agyria–pachygyria band spectrum; agenesis of corpus callosum; polymicrogyria; schizencephaly; periventricular nodular heterotopia; microcephaly; arachnoid cyst
Symptomatic epilepsy – predominately acquired causation	Hippocampal sclerosis	Hippocampal sclerosis
	Perinatal and infantile causes	Neonatal seizures; postneonatal seizures; cerebral palsy; vaccination and immunization
	Cerebral trauma	Open head injury; closed head injury; neurosurgery; epilepsy after epilepsy surgery; non-accidental head injury in infants
	Cerebral tumour	Glioma; ganglioglioma and hamartoma; DNET; hypothalamic hamartoma; meningioma; secondary tumours
	Cerebral infection	Viral meningitis and encephalitis; bacterial meningitis and abscess; malaria; neurocysticercosis, tuberculosis; HIV
	Cerebrovascular disorders	Cerebral haemorrhage; cerebral infarction; degenerative vascular disease; arteriovenous malformation; cavernous hemangioma
	Cerebral immunological disorders	Rasmussen encephalitis; SLE and collagen vascular disorders; inflammatory and immunologic disorders
	Degenerative and other neurological conditions	Alzheimer disease and other dementing disorders; multiple sclerosis and demyelinating disorders; hydrocephalus and porencephaly
Provoked epilepsy	Provoking factors	Fever; menstrual cycle and catamenial epilepsy; sleep–wake cycle; metabolic and endocrine-induced seizures; drug-induced seizures; alcohol and toxin-induced seizures
	Reflex epilepsies	Photosensitive epilepsies; startle-induced epilepsies; reading epilepsy; auditory-induced epilepsy; eating epilepsy; hot water epilepsy
Cryptogenic epilepsies		

Source: Data from Shorvon 2011 [36] and Shorvon 2011 [37].
DNET, dysembryoplastic neuroepithelial tumour; SLE, systemic lupus erythematosus.
[a]These examples are not comprehensive, and in every category there are other causes.

Table 1.12 Semiological classification of epilepsy (this is an alternative classification of seizure type which has not been formally adopted by ILAE).

Epileptic seizure
Aura
Somatosensory aura
Visual aura
Auditory aura
Gustatory aura
Olfactory aura
Autonomic aura
Abdominal aura
Psychic aura
Autonomic seizure
Dialeptic seizure
Typical dialeptic seizure
Motor seizure
Simple motor seizure
Tonic seizure
Myoclonic seizure
Epileptic spasm
Clonic seizure
Tonic–clonic seizure
Versive seizure
Complex motor seizure
Hypermotor seizure
Gelastic seizure
Automotor seizure'
Special seizure
Atonic seizure
Astatic seizure
Hypomotor seizure
Akinetic sieuzre
Negative myoclonic seizures
Aphasic seizure
Paroxysmal events (of non-epileptic origin)

Source: Lüders *et al.* 1998 [40]. Reproduced with permission from John Wiley & Sons.

neuroimaging, should be analysed separately and integrated with the clinical findings only after the clinical findings have been categorized.

In this classification, the ictal symptoms were divided into sensory, consciousness and motor categories. There is an emphasis on the aura (demonstrating the Jacksonian principle that the first symptom of a seizure gives away its cerebral location) and also on the temporal sequence of events in a seizure. An example of a seizure description using this scheme is olfactory aura → automotor seizure → left versive seizure → generalized tonic–clonic seizure.

In 2005, the authors went further and proposed a five-tier classification system. Two tiers (semiology and frequency) define the symptoms {the epileptic seizure] and three tiers (aetiology, associated neurological deficits and location of the epilepsy) define what is producing the epilepsy and the location of the brain abnormality [41].

The analysis of an epilepsy, according to this scheme, goes forward in the following tiers: identification of brain location → seizure semiology → aetiology → seizure frequency → related medical conditions (later modified to a four-dimensional system, dropping seizure frequency).

The semiological classification embedded within this scheme has a number of drawbacks. It introduced new terms (seizure descriptors) which some find unnecessary and obscure, such as dialeptic, automotor, hypomotor and hypermotor. This classification was devised by a unit focusing on epilepsy surgery, and hence the prominence given to structural aspects of epilepsy and aspects most pertinent to surgery. There is no doubt that it works best for focal lesional epilepsies but less well for the common or garden varieties. Its major role is in presurgical assessment, where the meticulous unpackaging of seizure semiology provides useful information in some cases [41].

Lüders resigned from the ILAE Taskforce on Classification, as he disagreed with the ILAE approach, and the ILAE have similarly not endorsed Lüder's scheme. However, its use even for surgical assessment is limited by the fact that partial seizures, as Gastaut himself realized, are often not localized to one area of cortex but are formed by neuronal networks which can be extensive. This is a fact often swept under the carpet by enthusiasts of seizure localization, hunting for an illusory 'focus' using semiological clues. No amount of semiological analysis in this significant number of patients will be able to overcome this essentially insurmountable obstacle.

Definition

Acute symptomatic seizures

Another rather simpler classification system has been used, especially in epidemiological work, which divides epilepsies and seizures into acute symptomatic, remote symptomatic and idiopathic categories. This schema seems first widely applied in the 1970s in the landmark epidemiological work from Rochester. It then fell from fashion, and interestingly neither acute symptomatic nor remote symptomatic are terms included in the 2001 glossary. The main reason for categorizing epilepsy in this way was to ensure that 'acute symptomatic seizures' were not included within the term 'epilepsy', as these seizures differ in context and prognosis from those in 'genuine' epilepsy. Recently, the Epidemiology Commission of the ILAE convened a subgroup to reconsider the definition of acute symptomatic seizures for epidemiological studies. This group has modified the meaning of the original terms and defined an acute symptomatic seizure as 'a clinical seizure occurring at the time of a systemic insult or in close temporal association with a documented brain insult' [42].

There are two problems with the usage proposed by the Commission. First, the term covers two quite different clinical situations: (i) The 'early seizures' in acute brain insults; and (ii) the seizures provoked by reversible environmental metabolic disturbances or toxins. The two clinical categories could not be more different and should not be intermixed. In the first there is severe destruction of brain tissue and many patients progress to develop chronic epilepsy and neurological deficits. In the second category there is no underlying brain pathology and there are no known physiological differences from patients with existing epilepsy who experience seizures provoked by the same cause or indeed from individuals who do not have seizures when exposed to the same precipitant. The provoking factors probably light up what is in effect a low 'seizure threshold' (the 2001 glossary defines a 'provocative factor' as a 'transient and sporadic endogenous or exogenous element capable of augmenting seizure incidence in persons with chronic epilepsy

and evoking seizures in susceptible individuals without epilepsy'). Once the metabolic or toxic exposure is reversed, the prognosis is excellent and none of these patients progress to have chronic epilepsy. It makes no sense to lump these two totally disparate types of seizure together.

The second problem is the arbitrary nature of the criteria for inclusion. The ILAE scheme for instance categorizes a seizure as 'acute symptomatic' within 1 week of trauma and stroke, but longer (not specified) for a subdural haematoma or infections. Parasitic infections are included but congenital toxoplasmosis excluded. Alcohol withdrawal seizures are included but not alcohol-induced seizures. Seizures caused by environmental triggers such as visual stimulation are not included but seizures induced by hypoglycaemia are included. Furthermore, the term has even been extended to include seizures that lead to the diagnosis of progressive conditions such as tumours (primary and secondary) which are in fact 'remote' symptomatic seizures'. In the metabolic conditions, arbitrary cut-off levels are cited despite the fact that the rate of change of metabolic parameters is as important as the extent of change.

For these reasons, in the author's opinion, the classification of epilepsy into remote symptomatic, acute symptomatic and idiopathic seizures, and especially the term 'acute symptomatic seizure', should be abandoned in the meaning given by the ILAE Commission. If the term 'acute symptomatic' is to be retained, it should be restricted to the physiologically distinct 'early seizures' after acute brain injury. Acute seizures caused by metabolic disturbance or toxins should be simply referred to as 'provoked seizures' [43].

Whatever term is used, it is important to point out that the early seizures after acute brain injury are quite different from the late post-injury seizures. In early seizures, the epilepsy may be caused by contusions, haemorrhage, metabolic change, endocrine change, hypotension, and so on. These are mechanisms that have nothing in common with the late seizures of post-traumatic epilepsy. Thus, it makes sense to differentiate the two.

Epilepsy in remission

Another important distinction for clinical practice is the difference between epilepsy in which seizures are controlled on or off treatment, and epilepsy in which seizures continue despite treatment. Studies have shown that after 10–20 years after the onset of epilepsy, >70% of individuals no longer have seizures (i.e. are in remission). Of course, the actual number of cases in remission depends on how long the seizure-free period must be to qualify as a remission. All studies in this field have shown that the longer the period of seizure freedom, the less likely is subsequence recurrence. However, even after long periods of remission, seizures do occasionally recur. For most studies, remission has been defined as a 2 or 5-year period without seizures.

In the 2014 ILAE definition of epilepsy [6], the condition is considered to be 'no longer present' when either: (i) individuals who had an age-dependent epilepsy syndrome but are now past the applicable age; or (ii) those who have remained seizure free for at least 10 years off antiseizure medicines, provided that there are no known risk factors associated with a high probability (>75%) of future seizures.

The 2014 taskforce added that the term 'resolved' was considered not necessarily identical to the conventional view of 'remission' or 'cure'. However, as all three terms can be only demonstrated retrospectively, there is no practical difference between them. The taskforce also recognized that different practical definitions (e.g. different durations of seizure freedom) can be formed and used for different specific purposes. This is an important consideration, as, for instance, the legal requirements for driving are not necessarily the same as the clinical requirements in terms of treatment.

Provoked epilepsy and reflex epilepsy

It has been known for centuries that seizures can be 'provoked' by various factors [43]. In the nineteenth century, all seizures were considered to have both predisposing and also exciting components, and the production of seizures was considered invariably the result of both influences acting together (Jackson repeated the commonly used analogy of gunpowder and the spark). This re-emphasizes the point that epilepsy has a multifactorial causation, and really the differentiation of 'underlying cause' from a 'seizure precipitant' is simply one of degree. In a recent survey, it was found that 97% of patients with epilepsy believe that there is at least one precipitant for some of their seizures, and 28% believe that there is a precipitant for all of their seizures [44].

A distinction is sometimes made between 'provoking' (precipitating) factors and 'reflex' epilepsy. The line between the two is not easy to define, and to do so is to apply largely arbitrary criteria. Gastaut [45] defined reflex epilepsies as those in which all seizures, or a large part of them, are reliably provoked by naturally occurring or artificial stimulation of a certain receptor or group of receptors, and a similar formulation was given by the 2001 glossary. In a recent textbook, the working definition of reflex epilepsy was 'an epilepsy in which seizures are reliably provoked by a specific identifiable environmental trigger' [43].

Currently, the reflex epilepsies are commonly subdivided into two categories.

1 *Simple reflex epilepsies* – where the seizures are precipitated by simple sensory stimuli (e.g. flashing lights, startle). Photosensitive epilepsy is by far the most common type and has been extensively studied. The frequency and type of visual stimulation can be highly specific in individuals, and there is also a genetic predisposition in some cases.
2 *Complex reflex epilepsy* – where the stimuli are more integrative and complicated. Examples include musicogenic epilepsy, in which sometimes a highly specific piece of music triggers the seizures, or seizures induced by thinking, reading, eating or sometimes highly specific cognitive tasks.

Reflex epilepsies can be either focal or generalized. Internal triggers, such as the effects of menstruation of fatigue, are not usually included in the category, nor are more indirect external triggers such as alcohol intake.

Definition and classification – status epilepticus

Status epilepticus is a type of epilepsy that has been recognized since the beginning of recorded medical history [46,47]. The term '*état de mal*' though was coined by Calmeil in 1824 in his doctoral thesis, where he notes it was used by patients in the Parisian asylums. The first detailed modern medical description was by Bourneville in 1869. At that time, the usage of the term status epilepticus was confined to what is now known as tonic–clonic status epilepticus, and it was only after the advent of EEG that it was realized that continuing or prolonged seizure activity could take various forms.

The first major conference devoted solely to the topic of status epilepticus was the Xth Marseille Colloquium, held in 1962 led by Henri Gastaut [48]. A total of 103 participants presented 237 cases with both clinical and EEG findings of abnormally

prolonged or serially repeated seizures. A new definition of status was proposed: 'status epilepticus is a term used whenever a seizure persists for a sufficient length of time or is repeated frequently enough to produce a fixed or enduring epileptic condition'. Although no duration was specified in the definition, Gastaut later specified a duration of 60 minutes to define status epilepticus. Another development of great importance coming from the colloquium was the concept, as Gastaut put it, that 'there were as many types of status as there were types of epileptic seizure' [49]. Gastaut was, at that time, also leading the formulation of the ILAE seizure type classification, and he envisaged that the classification of status epilepticus could take the same form. Status was thus subdivided then into three types: generalized status epilepticus, partial status epilepticus and unilateral status epilepticus. This categorization appeared in a addendum to the 1969/1970 ILAE classification [12,13].

In the revision of 1981, status epilepticus was relegated to the addendum where the definition was minimally changed into a 'seizure that persists for a sufficient length of time or is repeated frequently enough that recovery between attacks does not occur' [16], and was subdivided in the addendum to partial (e.g. Jacksonian) or generalized (e.g. absence status or tonic–clonic status) categories, and that 'when very localized motor status occurs, it is referred to as epilepsia partialis continua'.

Definition and classification were the subject of detailed consideration in the monograph on status epilepticus published in 1994, where a definition was proposed: 'Status epilepticus is a condition in which epileptic activity persists for 30 minutes or more, causing a wide spectrum of clinical symptoms, and with a highly variable pathophysiological, anatomical and aetiological basis' [46]. A detailed hybrid classification was proposed, which attempted comprehensively to incorporate all types of status epilepticus, subdivided by age, and also a group of conditions, termed 'boundary syndromes' in which it was not clear to what extent these conditions were due to epileptic activity (Table 1.13). In this monograph, the problems of classification were discussed at length and the difficulties of differentiating various forms of non-convulsive status epilepticus were particularly noted, not least because both diagnosis and categorization are particularly reliant on EEG patterns which could be variable and non-specific.

Tonic–clonic status epilepticus (convulsive status epilepticus) was in this book also divided into four temporal stages, which was considered important in order to organize an appropriate treatment protocol: the stage of premonitory status epilepticus (treated with benzodiazepines, often out of hospital) the stage of early status epilepticus (0–30 minutes: treated with IV benzodiazepines), the stage of established status epilepticus (30–60/90 minutes: treated with IV antiepileptics) and the stage of refractory status epilepticus (after 60/90 minutes: treated IV anaesthesia). The emphasis on timing of treatment led Lowenstein to propose that any convulsive seizure continuing for more than 5 minutes in duration should be considered a case of status epilepticus. This 'operational definition'

Table 1.13 1994 Definition and classification of status epilepticus.

Definition

Status epilepticus (SE) is a condition in which epileptic activity persists for 30 minutes or more, causing a wide spectrum of clinical symptoms, and with a highly variable pathophysiological, anatomical and aetiological basis

Classification

SE occurring in the neonatal and infantile epilepsy syndromes
West syndrome
Ohtahara syndrome
Severe myoclonic encephalopathy of infancy (SMEI; Dravet syndrome)
SE in other forms of neonatal or infantile epilepsy

SE occurring only in childhood
SE in early-onset benign childhood occipital epilepsy (Panayiotopoulos syndrome)
SE in other forms of childhood epileptic encephalopathies, syndromes and aetiologies (e.g. ring chromosome X and other karyotype abnormalities, Angelman syndrome, Rett syndrome, myoclonic–astatic epilepsy), other childhood myoclonic encephalopathies
Electrical status epilepticus in slow-wave sleep (ESES)
Landau–Kleffner syndrome

SE occurring in both childhood and adult life
Convulsive SE
Tonic–clonic SE
Epilepsia partialis continua
Myoclonic SE in coma (after severe brain injury)
Myoclonic SE
NCSE with epileptic encephalopathy
NCSE in Lennox–Gastaut syndrome
  Atypical absence status epilepticus
  Tonic status epilepticus
Other forms of NCSE in patients with learning disability or disturbed cerebral development (cryptogenic or symptomatic)
NCSE without epileptic encephalopathy
Typical absence status epilepticus in idiopathic generalized epilepsy
Complex partial status epilepticus
  Limbic
  Non-limbic
NCSE in the aftermath of tonic–clonic seizures
Subtle status epilepticus (myoclonic SE occurring in the late stage of convulsive SE)
Aura continua with sensory, special sensory, autonomic or cognitive symptoms

SE occurring in late adult life
De novo absence status epilepticus of late onset

Boundary syndromes
Some cases of epileptic encephalopathy
Some cases of coma due to acute brain injury with epileptiform EEG changes
Some cases of epileptic behavioural disturbance or psychosis
Some cases of drug-induced or metabolic confusional state with epileptiform EEG changes

Source: Shorvon 1994 [46]. Reproduced with permission from Cambridge University Press.
NCSE, non-convulsive status epilepticus.

was formulated so that there was no delay in initiating emergency therapy in patients with prolonged seizures [50].

In 2001, the ILAE glossary of terms [4] defined status epilepticus in a rather obtuse way: 'a seizure that shows no clinical signs of arresting after a duration encompassing the great majority of seizures of that type in most patients or recurrent seizures without interictal resumption of baseline central nervous system function'. The ILAE Core Group of the Commission on Terminology and Classification [25] in 2006 included status epilepticus in its listing of 'seizure type', albeit in a rather incomplete fashion Table 1.7.

In 2014, a taskforce of the ILAE Commission on Classification and Terminology produced a proposal for a new definition of status epilepticus [51]: in which two time points are mentioned – the time point at which seizure activity can be considered continuous (t1) and the time point that might lead to long-term consequences (t2) including neuronal death, neuronal injury, and alteration of neuronal networks.

As the taskforce put it, this is a 'conceptual' definition with two operational dimensions. In the case of convulsive (tonic–clonic) status epilepticus, both time points (t1 at 5 minutes and t2 at 30 minutes) are based on some evidence from animal experiments and (albeit minimal) clinical research, but data on these time points were not available for most other forms of status epilepticus, and it was hoped that the proposal would stimulate research to define these.

The taskforce also devised a new classification scheme. This took the form of four axes: (i) semiology; (ii) aetiology; (iii) EEG correlates; and (iv) age. Axis 1 (semiology) lists different forms of status epilepticus divided into those with prominent motor systems, those without prominent motor systems, and currently indeterminate conditions (such as acute confusional states with epileptiform EEG patterns). The category of non-convulsive status epilepticus (i.e. without prominent motor features) was divided into those cases in coma and those not in coma. Axis 2 (aetiology) is divided into subcategories of known and unknown causes. Axis 3 (EEG correlates) adopts the latest recommendations by consensus panels to use the following descriptors for the EEG: name of pattern, morphology, location, time-related features, modulation and effect of intervention. Finally, Axis 4 divides age groups into neonatal, infancy, childhood, adolescent and adulthood, and elderly (Table 1.14). This is

Table 1.14 The four axes in the classification of status epilepticus (SE) proposed by the 2014 ILAE Taskforce.

Axis 1. Classification according to semiology

A. With prominent motor symptoms

1. Convulsive SE (CSE, synonym: tonic–clonic SE)
   (a) Generalized convulsive
   (b) Focal onset evolving into bilateral convulsive SE
   (c) Unknown whether focal or generalized
2. Myoclonic SE (prominent epileptic myoclonic jerks)
   (a) With coma
   (b) Without coma
3. Focal motor
   (a) Repeated focal motor seizures (Jacksonian)
   (b) Epilepsia partialis continua (EPC)
   (c) Adversive status
   (d) Oculoclonic status
   (e) Ictal paresis (i.e. focal inhibitory SE)
4. Tonic status
5. Hyperkinetic SE

B. Without prominent motor symptoms (i.e. non-convulsive SE, NCSE)

1. NCSE with coma
2. NCSE without coma
   (a) Generalized
      i. Typical absence status
      ii. Atypical absence status
      iii. Myoclonic absence status
   (b) Focal
      i. Without impairment of consciousness (aura continua, with autonomic, sensory, visual, olfactory, gustatory, emotional/psychic/experiential, or auditory symptoms)
      ii. Aphasic status
      iii. With impaired consciousness (i.e. dyscognitive SE)
   (c) Unknown whether focal or generalized
      i. Autonomic SE

Boundary syndromes (currently indeterminate conditions)

1. Epileptic encephalopathies
2. Coma with non-evolving epileptiform EEG pattern[a]
3. Behavioural disturbance (e.g. psychosis) in patients with epilepsy
4. Acute confusional states (e.g. delirium) with epileptiform EEG patterns

Axis 2. Classification according to aetiology

1. Known (synonymous: symptomatic)
   (a) Acute (e.g. stroke, intoxication, malaria, encephalitis)
   (b) Remote (e.g. post-traumatic, post-encephalitic, post-stroke)
   (c) Progressive (e.g. brain tumour, Lafora disease and other PMEs, dementias)
   (d) SE in defined electroclinical syndromes
   (e) Unknown (synonymous: cryptogenic)

Axis 3. Classification according to EEG correlates

Currently there are no evidence-based EEG criteria for SE. The terminology was proposed to describe EEG patterns in status epilepticus:

1. *Name of the pattern:* periodic discharges, rhythmic delta activity or spike–wave/sharp–wave plus subtypes
2. *Morphology:* sharpness, number of phases (e.g. triphasic morphology), absolute and relative amplitude, polarity
3. *Location:* generalized (including bilateral synchronous patterns), lateralized, bilateral independent, multifocal
4. *Time-related features*: prevalence, frequency, duration, daily pattern duration and index, onset (sudden or gradual) and dynamics (evolving, fluctuating or static)
5. *Modulation:* stimulus-induced or spontaneous
6. *Effect of intervention (medication) on EEG*

Axis 4. Classification according to age

1. Neonatal (0–30 days)
2. Infancy (1 month to 2 years)
3. Childhood (3–12 years)
4. Adolescence and adulthood (13–60 years)
5. Elderly (>60 years)

Source: unpublished data [51].

NCSE, non-convulsive status epilepticus; PME, progressive myoclonus epilepsy.

[a] e.g. Periodic lateralized and generalized periodic discharges.

work in progress and it will be interesting to see to what extent this new scheme is helpful in practice.

Afterthought

In this chapter, I have tried to sketch out the evolution over time of definition and classification of epilepsy since the work of Jackson (whose oeuvre marks the dawn of modern epilepsy), to provide a summary of current classifications and provide a brief critique. Since the 1960s, these topics have become synonymous with the ILAE, and the ILAE classification structures created between 1969 and 1989 are amongst the organization's greatest achievements. A number of general observations concerning classification and terminology in epilepsy become apparent when an historical approach is taken, and I end this chapter with a brief consideration of these.

What is abundantly clear from the historical perspective is that the state of knowledge in the field of epilepsy is such that our current and past classifications schemes have been by necessity utilitarian and not scientific (i.e. gardening in nature and not botanical to use Jackson's analogy). Although much effort has been expended, and with boring regularity unsubstantiated claims to the contrary have been made, we are as far as ever from being able to devise a suitably scientific schema.

In devising a utilitarian classification, a few other points become apparent. First, there is a danger that, through a desire to be too all-inclusive, the schemes become too complex and unwieldy, thereby losing their utilitarian value. Given that they are gardening in character and thus derive their worth entirely from their utility in common practice, complexity is an enemy. The failure of uptake of the 1989 *Classification of the Epilepsies and Epileptic Syndromes* into widespread usage, for instance, was not due to its poor quality (indeed, on the contrary, it is of excellent quality) but due to its complexity. Similarly, Gastaut's summary classification of 1964 (Table 1.3) became more popular and more widely cited and utilized that the full 1969/1970 classification which was the official version (Table 1.4). As knowledge advances, the field becomes more complex, but the difficult trick for classificationists is to maintain a balance between the Scylla of superficiality and the Charybdis of intricacy. A very useful method of avoiding this is to consider each 'axis' of classification separately – and thus produce unidimensional lists or databases. This was what Engel postulated in 2001 and what the current Commission is also pursuing. Of course, these 'axes', known then as 'criteria' or 'parameters', were used in all the previous schemes, but were not there separated so conclusively as is the current trend. Even with such databases, there is a need for a single simple scheme, and the main categories of the 1989 classification are difficult to beat in this regard.

Another essential feature, often overlooked, is the extraordinary care needed when devising terminology. Jackson's invention of the term 'discharge' for an epileptic seizure is a good example of a term that continues to be used and which captures the essence and the **nature** of a seizure. It has also, simply through its linguistic implications, directed scientific research. Modern sloppiness in the use of terminology, which is currently a widespread problem, has the absolutely opposite effect.

Furthermore, the current fashion for continual change, often spuriously justified as being needed by advances in science, should be deprecated. All that is achieved by continuous tinkering of definition and classification is chaos and confusion, and this erodes the authority of any scheme. We have been particularly guilty of this in the past decade. Terminology matters, and it is a mistake continually to modify this. To do so is damaging as well as unnecessary, and there are disadvantages not only for epileptologists, but also for those engaged in more general medical practice and in legal, regulatory and societal arenas. The WHO realized the importance of having an authoritative dictionary, but having formed such a dictionary, its pages should not be torn up with every change of leadership.

Finally, and quite remarkably, history shows that classification, perhaps more so than all other topics in epilepsy, has been often the cause of dissention and conflict – a true minefield – for the foot soldiers of epilepsy. This is perhaps because at one level it is the work of gardeners not botanists, and thus has been usually a matter of opinion (assertion) and not of fact. The flare-up of passions in 1969/1970, in 1989 and now in 2010–2014 are evidence of this. Everyone can have an opinion, and it seems that everyone does, often from a limited viewpoint (the current author is guilty of this), and it is sad to see the intrusion of politics and personal opinions and vanities into a field that should be dry and academic. One lesson is that the schemes, being largely of an opinionated nature, must win the approval of the community at large and cannot be forced through on to unwilling recipients. To try to do so simply causes conflict, as has been evident on several occasions. One way of garnering wide support is for committees or panels to devise the systems and for the drafts to be sent out for wide consultation. However, this only works if the results of the consultation are heeded, as Gastaut found to his cost in 1969/1970. The best classification schemes have also depended on an acceptance of the authority of their authors, and the widespread respect that Jackson, Gastaut and Dreifuss commanded was instrumental in the success of their schema.

What about the future? Let us hope a botanical scheme, fully scientifically justified, based on such aspects as pathophysiology, neurochemical systems, or physiological or anatomical networks, and with carefully chosen terminology, will eventually be possible, and, it is hoped, one which remains tagged with the ILAE name.

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