Chapter 2

Classification and terminology to organise seizures and epilepsies

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Classification schemes can appear dull and cumbersome, but are crucial to facilitate diagnosis of epilepsy, communication between health professionals, and communication between professionals and people with epilepsy.

Because there is not a one-to-one mapping between underlying disease and clinical phenomenology in the epilepsies, there is a need for at least two separate schemes: a classification of *seizures* (i.e. a scheme based on clinically observable phenomenology) and an organisation* of *epilepsies* (i.e. a scheme that considers aspects beyond seizure semiology such as age at onset, neuroimaging and genetic aspects, aetiology, and prognosis). Even more ambitiously, the International League against Epilepsy (ILAE) has recently proposed a set of classification schemes based on five axes: ictal phenomenology, seizure type, syndrome, aetiology, and impairment. However, this approach has not yet achieved consensus approval.

The ILAE commission on classification and terminology has created an online manual at https://www.epilepsydiagnosis.org/index.html. This practical tool provides a three-tiered structure of seizure classification, epilepsy syndromes, and aetiologic categories.

The ILAE established a standardised classification and terminology for epileptic seizures in 1981. It is important to recognise that this scheme utilised seizure manifestations and EEG findings only. In the 30 plus years since then, advances, especially in neuroimaging and genetics, have had a major impact on our understanding of epilepsy. The ILAE has recognised the pressing need for a new scheme, and the proposal from 2010 will be discussed here alongside the 1981 scheme (Tables 1 and 2).

Both classification schemes are based on the widely accepted concept that seizures can be focal or generalised. A *generalised* seizure is conceptualised as originating within, and rapidly engaging neuronal networks in both hemispheres (though not necessarily the entire cortex). Nonetheless, generalised seizures can be asymmetric to some extent in their clinical and EEG manifestations. A *focal* seizure is thought to arise within an area confined to one hemisphere. Focal seizures can spread within the same hemisphere, and/or to areas in the contralateral hemisphere, and evolve into a generalised convulsive seizure (i.e. adopt the semiology of a primary generalised tonic-clonic seizure). The (initial) seizure semiology in focal seizures often reflects the functional role of the cortex that is involved at the onset and early evolution of the seizure.

*The ILAE commission uses the term *organisation* to recognise that knowledge about underlying seizure mechanisms is evolving and often still inadequate to allow a scientifically based *classification*.

Table 1. 1981 International classification of seizures.

Partial seizures beginning locally
Simple (consciousness not impaired)
• With motor symptoms
• With somatosensory or special sensory symptoms
• With autonomic symptoms
• With psychic symptoms
Complex (with impairment of consciousness)
• Beginning as simple partial seizure (progressing to complex seizure)
• Impairment of consciousness at onset
a) Impairment of consciousness only
b) With automatism
Partial seizures becoming secondarily generalised
Generalised seizures
Absence seizures
• Typical
• Atypical
Myoclonic seizures
Clonic seizures
Tonic seizures
Tonic-clonic seizures
Atonic seizures

Objective support for this dichotomy of focal and generalised seizures is provided by EEG findings in patients with generalised and focal seizures. In generalised seizures, there is immediate (within milliseconds) EEG involvement of both hemispheres. Inter-ictal abnormalities likewise involve both hemispheres simultaneously. In focal seizures, the ictal discharge is initially confined to one hemisphere. Inter-ictal abnormalities are also confined to individual areas.

An inherent limitation to this scheme is that seizures are not classified by their semiologic features alone, but in combination with EEG findings. It has to be recognised that the *semiological* features seen in the generalised seizure types described above can be seen in focal seizures as well (e.g. clonic movements, atonic head drop, brief loss of awareness).

Generalised seizures (ILAE 2010)

Tonic-clonic seizures (in any combination)
In its classic form, this seizure is a sequence of events that often begins with bilateral myoclonic jerks. This is then followed by a tonic contraction of the extremities and axial trunk muscles, resulting in extension of the neck and extension of the extremities. The tonic contraction of the diaphragm and abdominal and chest wall muscles against the contracted glottis causes the characteristic tonic cry. The patient may become cyanotic during this phase. The generalised tonic activation can only be sustained for a short period of time. The

Table 2. Proposed new ILAE classification scheme

Generalised seizures

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

Focal seizures

- *Without impairment of consciousness or awareness*
  - With observable motor or autonomic components
    - ‘Focal motor’ and ‘autonomic’ can be used
  - Involving subjective sensory or psychic phenomena only
    - ‘aura’ can also be used
  - Replaces term ‘simple partial seizure’

- *With impairment of consciousness or awareness*
  - ‘Dyscognitive’ can also be used.
    - It is understood that dyscognitive may not always mean altered awareness but it is used here to denote altered consciousness or awareness which may be response tested
  - Replaces term ‘complex partial seizure’

- *Evolving to a bilateral convulsive seizure*
  - May include tonic, clonic or tonic and clonic components in any order
  - Replaces term ‘secondarily generalised seizure’

Unknown

- Epileptic spasms

Continuous seizure types

- *Generalised status epilepticus*
  - Generalised tonic-clonic status epilepticus
  - Clonic status epilepticus
  - Absence status epilepticus
  - Tonic status epilepticus
  - Myoclonic status epilepticus

- *Focal status epilepticus*
  - Epilepsia partialis continua of Kojevnikov
  - Aura continua
  - Limbic status epilepticus (psychomotor status)
  - Hemiconvulsive status with hemiparesis

contractions become progressively longer and interrupted, resulting in clonic jerking of the extremities. There may be reflectory emptying of the bladder and bowels, and the patient may bite their tongue. There are variations on this theme: the tonic or the clonic phase may be skipped, and asymmetric features (e.g. forced head turn to one side, or asymmetric or asynchronous jerking) can be seen. After the seizure, the patient is in a deep coma, and it usually takes 15–60 minutes to regain consciousness. Patients often feel utterly exhausted, and report diffuse muscle aches, headache, or depressed mood for up to several days after a generalised tonic-clonic seizure.

Typical absence seizures
This seizure type is characterised by a behavioural and mental arrest for a few seconds. After the seizure, the patient typically resumes their activity as if nothing had happened. There may be subtle jerky movements of facial muscles, however the lapse of awareness with immediate post-ictal recovery is the characteristic feature. The typical absence seizure is seen in developmentally normal subjects and associated with rhythmic 3 Hz spike and wave complexes in the EEG.

Atypical absence seizures
These are seen in patients with developmental delay. The behavioural arrest may be longer, and it may be more difficult to ascertain the arrest as such, compared to the patient's behaviour at baseline. The EEG in these patients usually shows background abnormalities (diffuse slowing), and the spike and wave complexes are of less than 3Hz in frequency.

Absence seizures with special features
Eyelid myoclonia with associated interruption of awareness is the characteristic seizure type in Jeavon's syndrome. Myoclonic seizures (discussed below) can also be associated with momentary loss of awareness.

Myoclonic seizures
These are brief jerks of the extremities and/or axial trunk muscles. A myoclonic jerk generated in the cortex can be distinguished from subcortically generated movements by its brief duration (usually less than 50 msec).

Atonic seizures
The ictal phenomenon in these seizures is a sudden generalised loss of tone. This may manifest as a head drop, or, if the patient is standing, as a forward fall that the patient cannot mitigate, often resulting in head and facial injuries.

Focal seizures (ILAE 2010)

It is well recognised that seizure manifestations in focal seizures often reflect activation of underlying cortical areas. This may result in a subjective alteration of experience (aura), transient impairment of cognition or language, behavioural alterations (e.g. clonic movements of one part of the body), and autonomic manifestations (e.g. brady- or tachycardia, pallor, hypersalivation). Complex behaviours such as semi-purposeful automatisms (e.g. chewing, lip smacking, fumbling, pedalling), kicking and thrashing movements, and shouting, humming, or laughter may be seen as well. These automatic behaviours likely reflect activation or disinhibition of a specific cortical/subcortical network. Consciousness may be preserved, altered, or lost during a seizure. The listed features may occur in any combination.

The 1981 proposal emphasised alteration of awareness ('simple' partial seizure reflecting retained awareness, and 'complex' partial seizure implying impaired awareness). It has been recognised since that awareness cannot be easily judged during a seizure, unless the patient

is actively tested for it. On the one hand, the patient may not be aware that he loses consciousness during a seizure. On the other hand, the presence of an automatic behaviour that is out of context for the situation does not necessarily imply that the patient has lost consciousness (because he lost 'control'). Although it is often the impairment of awareness that defines the disability resulting from a seizure (e.g. implications for driving), other ictal manifestations can be equally disabling (even when consciousness is preserved). Furthermore, the terms *simple* and *complex* are somewhat intransparent. *Simple* also has the connotation of 'not serious'. It is for all these reasons that the current ILAE proposal has abandoned the terms *simple* and *complex partial seizure*.

Consequently, the term *simple partial seizure* has been replaced by 'focal seizure without loss of awareness', and *complex partial seizure* has been replaced by 'focal seizure with alteration of awareness'. The term *secondarily generalised tonic-clonic seizure* has been abandoned as well, and is replaced by *evolving into a bilateral convulsive seizure with tonic and/or clonic components*.

Epileptic spasms

Epileptic spasms are placed separately (i.e. aside from generalised and focal seizures). This seizure type occurs in infancy and is characterised by tonic flexion of the head, neck and trunk, with circumflexion of the upper extremities. It is usually seen in infants with extensive brain abnormalities (e.g. diffuse tuberous sclerosis). Epileptic spasms were considered a type of generalised seizure. However, they can be seen in children with gross structural lesions confined to one hemisphere, and surgical treatment can be curative in this setting (implying a focal aetiology in at least some cases).

Classification of epilepsies

A classification of epilepsies (as opposed to seizures) combines information about seizure semiology and EEG findings with information from neuroimaging, aetiology and associated conditions. From an intellectual perspective, it makes sense to classify these parameters independently.

The 2010 classification scheme streamlines the terminology for aetiology. In particular, the terms *idiopathic*, *symptomatic* and *cryptogenic* have been abandoned, since it was felt that these are not always used precisely, and may have different connotations. For example, idiopathic epilepsies are thought to have a good prognosis and respond well to anticonvulsants, whereas symptomatic epilepsies are often thought to have a poor prognosis. The new scheme proposes unidimensional terms for aetiology (*genetic, structural/metabolic, unknown*). This subclassification has been criticised as overly simplistic.

Status epilepticus

Seizures are almost always self-limiting. Rarely one may follow another in close succession (without complete recovery in between seizures), or the ictal activity may be ongoing. Status epilepticus has been traditionally defined as ongoing seizure activity for 30 minutes or more. However, most seizures self-limit within five minutes or less. From a pragmatic point of view, a seizure that lasts longer than five minutes often warrants pharmacological intervention. Principally, any seizure type listed in Tables 1 and 2 may occur as status epilepticus.

Convulsive status
This is a state of recurrent tonic-clonic seizures without recovery of consciousness between attacks. It represents a medical emergency with a high morbidity and mortality. Status may

occur in approximately 3% of people with epilepsy but it is most common in patients with severe epilepsy who are non-compliant with drug therapy. It may also occur in alcohol withdrawal, in acute meningitis or encephalitis, and in acute metabolic disturbances.

Nonconvulsive status
This term is used imprecisely for the following two very different scenarios:

a) Motor manifestations in convulsive status inevitably cease at some point, however the cerebral cortex may continue to generate ictal discharges ('no longer convulsive status epilepticus'). This represents the most severe form of status, with ongoing excitotoxicity on a cellular level and high morbidity and mortality from a clinical perspective.

b) Ictal activity that from the onset was not associated with motor manifestations. Usually, the leading symptom is a change in the patient's cognitive state (confusion, disorientation with subsequent amnesia). This kind of status epilepticus is thought to have focal origin, though this may no longer be evident once the ictal activity has been ongoing.

Focal status
The ongoing seizure activity that defines status epilepticus may be restricted to a confined brain area. In this setting, the ictal symptoms reflect the cortical area affected (e.g. aura continua, aphasia). One classic example is epilepsia partialis continua of Kojevnikov. This refers to repetitive jerking of muscles or muscle groups in the face, arm or leg, originally described in association with epidemic encephalitis in Russia. Nowadays, the most common aetiologies are vascular disease, Rasmussen encephalitis, and tumours.

Relative frequency of seizure types

Data on the relative frequency of seizure types is unsatisfactory, and is largely based on populations of patients with relatively severe epilepsy, including large numbers of patients with partial epilepsies. Furthermore, the milder the epilepsy the more difficult it is to determine on clinical and electroencephalographic grounds whether it is of primary generalised or partial type. With these restrictions in mind, most series would suggest that approximately one-third of epilepsies may be of a generalised type, while two-thirds are partial, most commonly with a temporal lobe origin.

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