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The New Classification of Seizures by the International League Against Epilepsy 2017

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Abstract

Purpose of Review This review presents the newly developed International League Against Epilepsy (ILAE) 2017 classification of seizure types.

Recent Findings The fundamental distinction is between seizures that begin focally in one hemisphere of the brain, generalized onset seizures that apparently originate in both hemispheres, and seizures of unknown onset. Focal seizures optionally can be subclassified according to whether awareness (a surrogate marker for consciousness) is intact or impaired. The next level of classification for focal seizures is motor (with subgroups automatisms, atonic, clonic, epileptic spasms, hyperkinetic, myoclonic, tonic), non-motor (with subgroups autonomic, behavior arrest, cognitive, emotional, sensory), and focal to bilateral tonic-clonic. Generalized seizures are categorized as motor (tonic-clonic, clonic, tonic, myoclonic, myoclonic-tonic-clonic, myoclonic-atonic, atonic, epileptic spasms) and non-motor/absence (typical, atypical, myoclonic, eyelid myoclonia).

Summary The classification allows new types of focal seizures and a few new generalized seizures, and clarifies terms used to name seizures.

Keywords Epilepsy · Seizure · Classification · Focal seizure · Generalized seizure · Taxonomy

Introduction

Classification of seizure types is useful for several purposes. Classification allows grouping for applying or testing therapies for different types of seizures. It provides a foundation for basic and clinical research and epidemiological studies. Classification provides a common language for clinicians to use to collaborate on patient care. A classification system provides words to people with epilepsy and their families to name their seizure types. The classification used by most epileptologists was developed in 1981 by a committee of the International League Against Epilepsy (ILAE) [1•], based upon review and analysis of hundreds of videotaped seizures. This was, in turn, a revision of an earlier classification proposed by Gastaut [2].

Motivation for a New Classification

The 1981 classification, which promulgated the terms simple partial seizure, complex partial seizure, generalized tonic-clonic seizure, absence seizure, secondarily generalized tonic-clonic seizure, and others, has been useful, but it has limitations. The classification has no place for several focal motor seizure types such as tonic, clonic, atonic, or myoclonic other than in a general "partial motor" category. Seizures are unclassified if the beginning is missed, even though it may be obvious that the seizure is tonic-clonic. The 1981 classification does not specifically name certain important seizure types. The words used to describe seizures, for instance "complex partial" or "psychic," may not be well understood by the public or non-specialist physicians. The role of consciousness in defining a seizure type is complicated and difficult to apply in practice. For these reasons, the ILAE commissioned a revision of the seizure classification [3••, 4•] and epilepsy classification [5••].

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A scientific classification would be based upon the underlying pathobiology of seizures and would explain why there are different types of seizures. Unfortunately, our current level of understanding does not allow construction of such a true classification system. Therefore, any seizure type classification will be operational (practical) and observational. Different needs might mandate different classification systems. Neonatologists might develop a classification quite different from one formulated by neurosurgeons, neuropharmacologists, or laboratory researchers. The ILAE 2017 classification system is directed at practicing clinicians, but endeavors to be of use to a wider population.

The Basic Seizure Classification

The basic seizure classification (Fig. 1a) is intended for use by practitioners not specializing on epilepsy. Seizures are divided into those that begin with focal onset—limited to one hemisphere of the brain—or generalized onset—with apparent clinical or EEG onset in both hemispheres of the brain. The term "focal" replaces the previously used term "partial." The category of unknown onset seizures is a placeholder when the nature of seizure onset is known with less than 80% confidence by the clinician (the 80% level of confidence was arbitrarily chosen to match the usual acceptable false-negative beta error in an experiment). An unknown onset seizure later may be reclassified as focal or generalized as new information becomes available.

Focal onset seizures are subdivided into those with retained and impaired awareness. Focal aware seizures correspond to the 1981 classification "simple partial seizures" and focal impaired awareness seizures to the previous designation "complex partial seizures." Awareness is employed as a surrogate marker for consciousness. The level of consciousness during a seizure has great practical implication for predicting the impact of the seizure on behavior. Other potential surrogate markers might have included level of responsiveness or memory, but both of these are difficult to retrospectively assess. As a practical matter, most seizures with impaired memory also demonstrate impaired awareness, except for rare pure amnestic seizures. Classification by level of awareness is optional and may be omitted if the level of awareness is either unknown or does not apply, for example, with a focal myoclonic seizure.

The next classification level of focal onset seizures is motor versus non-motor. Subtypes of these categories are specified in the expanded classification system described below. A seizure that starts focally and then spreads to bilateral tonic-clonic movement is called a focal to bilateral tonic-clonic seizure, corresponding to the old phrase "secondarily generalized tonic-clonic seizure." The 2017 classification reserves the word "generalized" for seizures that are generalized from onset and the word "bilateral" for seizures that start one

hemisphere and then propagate to both hemispheres. A focal to bilateral tonic-clonic seizure is more of a propagation pattern than an individual seizure type, but it is such an important and common type of seizure that a distinct name is justified.

Generalized onset seizures are categorized as motor and non-motor (absence) seizures. The generalized onset tonic-clonic seizure, colloquially referred to as "grand mal," is well known to the general clinician, and therefore has a place in the basic classification.

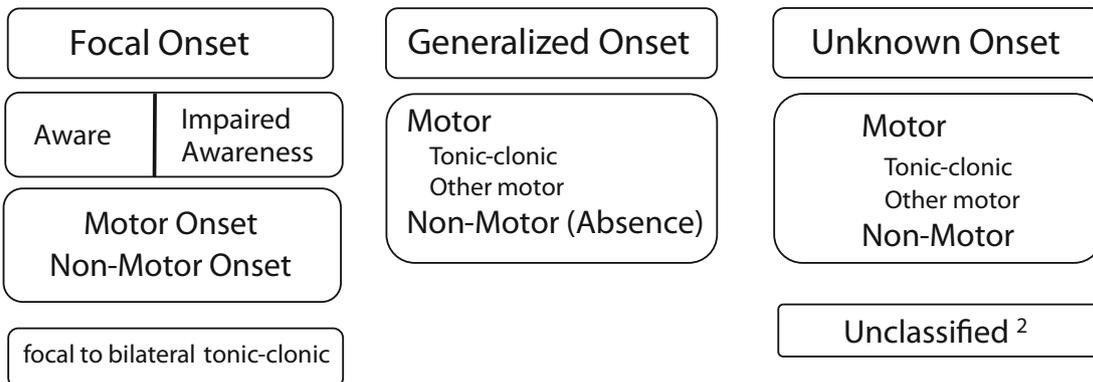## The Expanded Seizure Classification

The expanded seizure classification (Fig. 1b) is intended for use by clinicians with expertise in diagnosis and treatment of epilepsy. The framework is the same as that of the basic classification, but specific subheadings are expanded. Motor onset seizures optionally can be subdivided according to whether the first prominent manifestation is automatisms, atonic activity, clonic activity, epileptic spasms (previously called infantile spasms), hyperkinetic motions, myoclonic jerks, or tonic stiffening.

Focal Onset Seizures

Automatisms are robotic repetitive semi-purposeful movements during a seizure, such as lip-smacking, wandering, hand patting and rubbing, repetitive phrases, undressing, or other automatic activities. A focal atonic seizure manifests as focal loss of tone in a limb, whereas, a focal tonic seizure entail stiffening of the limb or the neck. Clonic implies sustained rhythmical jerking and myoclonic brief irregular jerking. A hyperkinetic seizure is synonymous with the term hypermotor seizure and may entail thrashing and pedaling. Epileptic spasms present with flexion at the waist and flexion or extension of the arms, usually in clusters in young children. Careful observation of the clinical and EEG pattern may be required to distinguish focal onset from generalized onset epileptic spasms. Epileptic spasms may be called "infantile spasms" when occurring during the infantile stage of life. Several of the focal motor categories also appear under the heading of generalized onset motor seizures; however, there is no implication that the pathophysiology or prognosis of seizures with corresponding names in each category is the same.

Focal non-motor onset seizures include autonomic seizures, behavioral arrest seizures, cognitive seizures, emotional seizures, and sensory seizures. A focal autonomic seizure is a seizure whose primary effect is on autonomic nervous system functions, such as heart rate, blood pressure, sweating, skin color, piloerection, gastrointestinal sensations. A focal behavior arrest seizure produces cessation of movement, sometimes called a freeze or a pause. Because brief behavior arrest is common and hard to recognize as being abnormal, a seizure
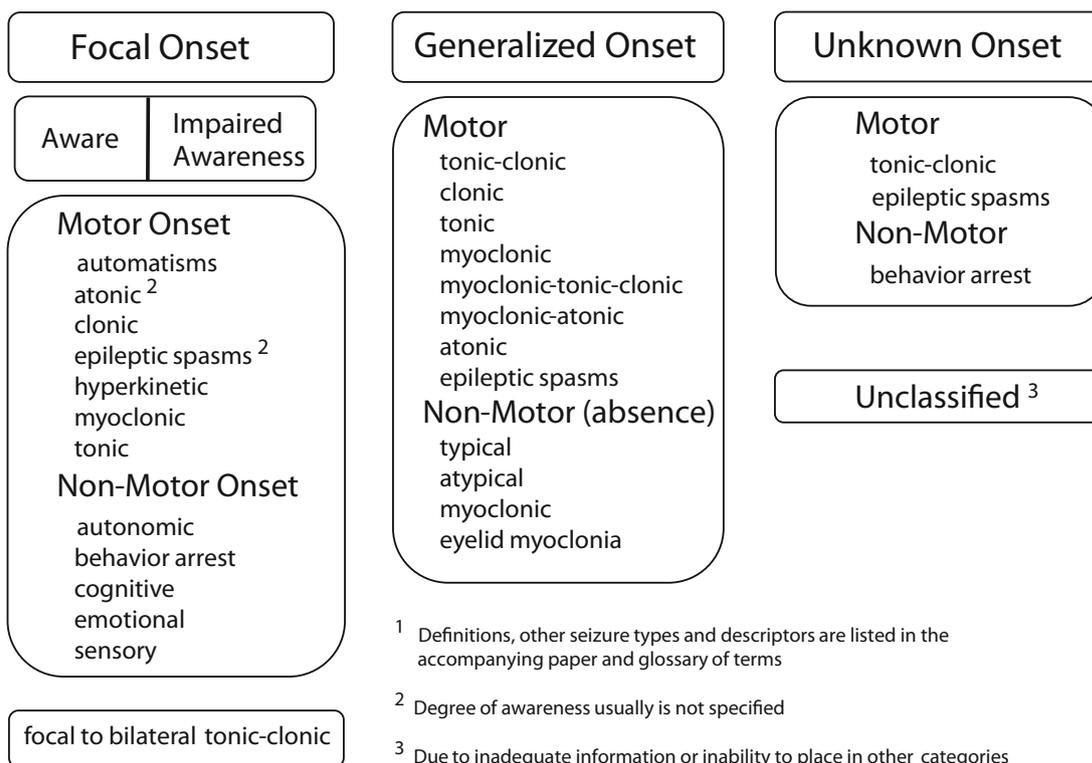
a   ILAE 2017 Classification of Seizure Types Basic Version [1]

Focal Onset

Aware	Impaired Awareness

Motor Onset
Non-Motor Onset

focal to bilateral tonic-clonic

Generalized Onset

Motor
  Tonic-clonic
  Other motor
Non-Motor (Absence)

Unknown Onset

Motor
  Tonic-clonic
  Other motor
Non-Motor

Unclassified [2]

[1] Definitions, other seizure types and descriptors are listed in the accompanying paper & glossary of terms

[2] Due to inadequate information or inability to place in other categories

b ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware	Impaired Awareness

Motor Onset
  automatisms
  atonic [2]
  clonic
  epileptic spasms [2]
  hyperkinetic
  myoclonic
  tonic
Non-Motor Onset
  autonomic
  behavior arrest
  cognitive
  emotional
  sensory

focal to bilateral tonic-clonic

Generalized Onset

Motor
  tonic-clonic
  clonic
  tonic
  myoclonic
  myoclonic-tonic-clonic
  myoclonic-atonic
  atonic
  epileptic spasms
Non-Motor (absence)
  typical
  atypical
  myoclonic
  eyelid myoclonia

Unknown Onset

Motor
  tonic-clonic
  epileptic spasms
Non-Motor
  behavior arrest

Unclassified [3]

[1] Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms

[2] Degree of awareness usually is not specified

[3] Due to inadequate information or inability to place in other categories

Fig. 1 a Basic seizure classification. **b** Expanded seizure classification. Figures reproduced with permission from the first author (Robert Fisher) and the publisher (Wiley) of reference [3••]

should only be classified as focal behavior arrest seizure if the behavior arrest is the dominant feature through the entire seizure. A focal cognitive seizure refers to impaired cognition during a seizure. The impairment might affect language, spatial perception, and ability to calculate arithmetic or other cognitive functions. A focal emotional seizure begins with spontaneous fear, anxiety or joy. An emotional seizure may involve involuntary laughing or crying, each of which might or might not be accompanied by a subjective emotion. Focal sensory seizures

can consist of tingling or numbness, visual symptoms, sounds, smells, tastes, vertigo, and hot-cold feelings. An unambiguous name, such as a focal sensory seizure, can omit the extra words from a full seizure name: focal onset non-motor sensory seizure.

Focal motor and non-motor seizures are classified by the first prominent sign or symptom, even if the first manifestation is not the most prominent feature during the course of a seizure. The initial manifestations mark the region or network of origin of the seizure. Impaired awareness is an exception, such that a focal seizure is classified as an impaired awareness seizure if awareness is impaired at any time during the seizure. A second exception is behavior arrest. Brief behavior arrest at the start of a seizure often is difficult to discern, so a focal behavior arrest seizure is one for which behavior arrest is the predominant manifestation throughout the entire seizure.

A seizure classification is far from a complete seizure description. Words following the declaration of seizure type can give crucial information about the seizure. These words may be drawn from other seizure classification terms, for example, "a *focal automatism seizure* with right arm tonic activity." Seizure descriptions also can be drawn from an ILAE-suggested common list of words called descriptors [4•]. These descriptors are cognitive: acalculia, aphasia, attention impairment, déjà vu or jamais vu, dissociation, dysphasia, hallucinations, illusions, memory impairment, neglect, forced thinking, responsiveness impairment; emotional or affective: agitation, anger, anxiety, crying (dacrystic), fear, laughing (gelastic), paranoia, pleasure; autonomic: asystole, bradycardia, flushing, gastrointestinal, hyper/hypoventilation, nausea or vomiting, pallor, palpitations, piloerection, respiratory changes, tachycardia; automatisms: aggression, eye-blinking, head-nodding, manual, oral-facial, pedaling, pelvic thrusting, perseveration, running (cursive), sexual, undressing, vocalization/speech, walking; motor: dysarthria, dystonic, fencer's posture (figure-of-4), incoordination, Jacksonian, paralysis, paresis, versive; sensory: auditory, gustatory, hot-cold sensations, olfactory, somatosensory, vestibular, visual; laterality: left, right, bilateral. Free text descriptions of seizures are also very useful.

Seizures evolve as they propagate and recruit brain networks not involved at the start of a seizure. A classification cannot easily distinguish a unitary seizure type with propagation patterns from sequential but different seizures. Rather, this is an issue of clinical diagnosis and one that may require observation of multiple seizures. A stereotyped pattern of signs and symptoms speak to a single seizure type, perhaps with multiple descriptors and free text appended to the seizure type. Varying patterns of signs and symptoms, accompanied by different time intervals between symptoms, might argue for separate multifocal seizures. This issue arises with both old and new seizure classifications.

Generalized Onset Seizures

Generalized seizure types are similar to those of the 1981 classification, with a few additions. Generalized onset seizures are not characterized by level of awareness, because awareness is usually impaired, although awareness can be hard to assess with myoclonic or atonic seizures. The primary division is into generalized onset motor and non-motor (absence) seizures.

A generalized tonic-clonic seizure produces immediate loss of awareness, accompanied by stiffening of all limbs (tonic phase), followed by sustained rhythmic jerking of limbs and

Table 1 Method for classifying a seizure

1. Onset	Decide whether seizure onset is focal or generalized, using an 80% confidence level.
2. Awareness	For focal seizures, decide whether to classify by degree of awareness or to omit awareness as a classifier.
3. Impaired awareness at any point	A focal seizure is a *focal impaired awareness seizure* if awareness is impaired at any point during the seizure.
4. Onset predominates	Classify a focal seizure by its first prominent sign or symptom. Do not count transient behavior arrest.
5. Behavior arrest	A *focal behavior arrest seizure* shows arrest of behavior as the prominent feature of the entire seizure.
6. Motor/non-motor	A *focal aware or impaired awareness seizure* maybe further subclassified by motor or non-motor characteristics. Alternatively, a focal seizure can directly be characterized by motor or non-motor characteristics, without specifying level of awareness. Example, a *focal tonic seizure*.
7. Optional terms	Terms such as motor or non-motor may be omitted when the seizure type is otherwise unambiguous.
8. Additional descriptors	It is encouraged to add descriptions of other signs and symptoms, suggested descriptors or free text. These do not alter the seizure type. Example, *focal emotional seizure* with tonic right arm activity and hyperventilation.
9. Bilateral vs. generalized	Use the term "bilateral" for tonic-clonic seizures that propagate to both hemispheres and "generalized" for seizures that apparently originate simultaneously in both.
10. Atypical absence	Absence is atypical if it has slow onset or offset, marked changes in tone or EEG spike-waves at less than 3 per second.
11. Clonic vs. myoclonic	Clonic refers to sustained rhythmical jerking and myoclonic to a regular unsustained jerking.
12. Eyelid myoclonia	Absence with eyelid myoclonia refers to forced upward jerking of the eyelids during an absence seizure.

face (clonic phase). Duration is typically 1–3 min. The seizure may produce a cry at the start, falling, tongue-biting, and incontinence. A generalized clonic seizure is associated with bilateral and sustained rhythmical jerking of limbs and/or head with no tonic stiffening phase. These seizures most often occur in young children. A generalized tonic seizure shows stiffening of all limbs, resolving without a clonic jerking phase. Generalized myoclonic seizures show unsustained irregular bilateral jerking of limbs, face, eyes, or eyelids. The jerking of generalized myoclonus may not always be left-right synchronous, but it occurs on both sides. Some clinicians also describe "negative myoclonus" as part of certain myoclonic seizures, although this may be difficult to distinguish from an atonic seizure or non-epileptic asterixis. A generalized myoclonic-tonic-clonic is like a tonic-clonic seizure, but it is preceded by a few myoclonic jerks on both sides of the body. Such seizures are commonly encountered in people with the syndrome of juvenile myoclonic epilepsy. Generalized myoclonic-atonic seizures present with a few myoclonic jerks, followed by a limp drop. Such seizures may be seen in children with Doose syndrome. A generalized atonic seizure is an epileptic drop attack, with sudden loss of muscle tone and strength and a fall to the ground or a slump in a chair. Atonic seizures usually last only seconds. Generalized epileptic spasms are brief seizures with flexion at the trunk and flexion or extension of the limbs. Generalized typical absence is associated with sudden onset of activity cessation, sometimes with eye fluttering and head nodding, or other automatic behaviors. If persistent for more than several seconds, awareness and memory are impaired. Recovery is immediate. The EEG during these seizures always shows generalized spike waves. Generalized atypical absence seizures are similar to typical absence seizures, but may have slower onset and recovery and more pronounced changes in tone. Atypical absence seizures can be difficult to distinguish from focal impaired awareness seizures, but absence seizures usually recover more quickly and the EEG patterns are different. A seizure with a few jerks and then an absence seizure is classified as a generalized myoclonic absence seizure. Eyelid myoclonia represents jerks of the eyelids and upward deviation of the eyes, often precipitated by closing the eyes or by light. Eyelid myoclonia may be associated with absence seizures. Although classifying eyelid myoclonia with non-motor (absence) seizures seems to be an oxymoron, the most important association is with absence seizures in people with Jeavon syndrome.

Supportive Information

Determination of seizure type usually can be made from signs and symptoms, but supplementary information can help to classify when such information is available. Supplementary information may include EEG, smartphone videos from home, neuroimaging, serum antibodies, genetic testing, and any other useful information. A generalized 3/s spike-wave pattern in an EEG will definitively classify a seizure with activity arrest and staring as a generalized absence seizure, rather than a focal behavior arrest seizure.

How to Use the Classification

Table 1, adapted from reference [4•], summarizes the practical approach to using the 2017 classification system.

Conclusions

The 2017 ILAE classification is an evolutionary advance over prior classifications, because a classification based on fundamental science is not yet possible. Some seizure types that were previously only generalized, such as tonic, atonic, myoclonic, clonic, and epileptic spasms, now have focal counterparts. New focal seizure types include automatism, behavior arrest, autonomic, cognitive, and emotional seizures. New generalized seizure types include myoclonic-tonic-clonic, myoclonic absence, and absence with eyelid myoclonia. Tonic-clonic, behavior arrest and epileptic spasm seizures can be provisionally classified even if onset is unknown. Level of awareness is employed as a surrogate for level of consciousness and optionally (if level of awareness applies and is known) subdivides focal seizures into aware and impaired awareness seizures.

The seizure classification is not a classification of the epilepsies or epilepsy syndromes. An epilepsy classification takes into account, not only seizure types, but also etiology, comorbidities and the broad clinical picture [5••]. The 2017 seizure classification will take some time to become common language, but ultimately should clarify what is meant by a particular type of seizure.

Human and Animal Rights and Informed Consent   This article does not contain any studies with human or animal subjects performed by any of the authors.

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