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Operational Classification of Seizure Types by the International League Against Epilepsy

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Summary

The International League Against Epilepsy (ILAE) presents a revised operational classification of seizure types. The purpose of such a revision is to recognize that some seizure types can have either a focal or generalized onset, to allow classification when the onset is unobserved, to include some missing seizure types and to adopt more transparent names. Because current knowledge is insufficient to form a scientifically-based classification, the 2017 classification is operational (practical) and based upon the 1981 Classification, extended in 2010.

Changes include: 1. “partial” becomes “focal”; 2. Awareness is used as a classifier of focal seizures; 3. The terms dyscognitive, simple partial, complex partial, psychic, secondarily generalized are eliminated; 4. New focal seizure types include automatisms, behavior arrest, hyperkinetic, autonomic, cognitive and emotional. 5. Atonic, clonic, epileptic spasms, myoclonic, tonic seizures can be either of focal or generalized onset. 6. Focal to bilateral tonic-clonic seizure replaces secondarily generalized seizure. 7. New generalized seizure types are: absence with eyelid myoclonia, myoclonic absence, myoclonic-atonic, myoclonic-tonic-clonic. 8. Seizures of unknown onset may have features that can still be classified. The new classification does not represent a fundamental change, but allows greater flexibility and transparency in naming seizure types.

Introduction

The International League Against Epilepsy (ILAE), through the Commission for Classification and Terminology, has developed a working classification of seizures and epilepsy. Following the proposed reorganization in 2010^{1,2}, further clarification has been discussed and feedback sought from the community. One area that required further elucidation was the organization of seizure types. A Seizure Type Classification Task was established in 2015 to prepare recommendations for classification of seizure types, which are summarized in this document. A companion document guides the intended use of the classification.

Descriptions of seizure types date back at least to the time of Hippocrates. Gastaut^{3,4} proposed a modern classification in 1964. Various basic frameworks for seizure classification can be considered. Manifestations of certain seizures are age-specific and depend on maturation of the brain. Previous classifications have been based on anatomy, with temporal, frontal,

parietal, occipital, diencephalic or brainstem seizures. Modern research changed our view of the pathophysiological mechanisms involved and has shown epilepsy to be a network disease and not only a symptom of local brain abnormalities.⁵ From a network perspective, seizures could arise in neocortical, thalamo-cortical, limbic, and brainstem networks. Although our understanding of seizure networks is evolving rapidly⁶ it is not yet sufficient to serve as a basis for seizure classification. In 1981, an ILAE Commission led by Dreifuss and Penry⁷ evaluated hundreds of video-EEG recordings of seizures to develop recommendations that divided seizures into those of partial and generalized onset, simple and complex partial seizures and various specific generalized seizure types. This classification remains in widespread use today, with revisions in terminology and classification of seizures and epilepsy by the ILAE^{2; 8-13 14} and with suggested insights, modifications and criticisms by others.¹⁵⁻²⁴ We chose not to develop a classification based solely on observed behavior - instead, reflecting clinical practice, the 2017 classification is interpretive, allowing the use of additional data to classify seizure types.

The intention of the 2001¹² and 2006¹³ reports on reclassification was to identify unique diagnostic entities with etiologic, therapeutic, and prognostic implications, so that when a syndromic diagnosis could not be made, then the therapy and prognosis would be based on seizure type. Such a classification would permit grouping of reasonably pure cohorts of patients for discovery of etiologies, including genetic factors, research into fundamental mechanisms, involved networks and clinical trials. The ILAE Seizure Type Classification Task Force (hereafter called “the Task Force”) chose to use the phrase “operational classification,” because it is impossible at this time to base a classification fully on the science of epilepsy. In the absence of a full scientific classification, the Task Force chose to use the basic organization initiated in 1981 and subsequently modified^{1; 2} as a starting point for the revised operational classification.

Methods

What is a seizure type?

A seizure is defined as “a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain”²⁵ It is the clinician’s first task to

determine that an event has the characteristics of a seizure and not one of the many imitators of seizures.²⁶ The next step is classification into a seizure type.

The Task Force operationally defines a seizure type as a useful grouping of seizure characteristics for purposes of communication in clinical care, teaching and research. Mention of a seizure type should bring to mind a specific entity, albeit sometimes with subcategories and variations on a theme. Choices must be made by interested stakeholders to highlight groupings of seizure characteristics that are useful for specific purposes. Such stakeholders include patients, families, medical professionals, researchers, epidemiologists, medical educators, clinical trialists, insurance payers, regulatory agencies, advocacy groups, and medical reporters. Operational (practical) groupings can be derived by those with specific interests. A pharmacologist, for example, might choose to group seizures by efficacy of medications. A researcher doing a clinical trial might consider seizures as disabling or non-disabling. A surgeon might group by anatomy in order to predict the eligibility for and likely success of surgical therapy. A physician based in an ICU with predominantly unconscious patients might group seizures in part by EEG pattern²⁷. **The principal aim of this classification is to provide a communication framework for clinical use.** Seizure types are relevant to clinical practice in humans; whereas, it is acknowledged that seizure types in other species, experimental and natural, may not be reflected in the proposed classification. One goal was to make the classification understandable by patients and families and broadly applicable to all ages, including neonates. The Commission on Classification & Terminology recognizes that seizures ~~and epilepsies~~ in the neonate can have motor manifestations or alternatively little or no behavioral manifestations. A separate Neonatal Seizure Task Force is working to develop a classification of neonatal seizures. The 2017 seizure classification is not a classification of electroencephalographic ictal or subclinical patterns. The guiding principle of the Seizure Type Task Force was a quotation from Albert Einstein: “Make things as simple as possible, but no simpler.”

Motivation for change

Adapting to a change of terminology can be effortful and needs to be motivated by a rationale for change. Seizure type classification is important for several reasons. First, the classification becomes a worldwide shorthand communication among clinicians caring for

people with epilepsy. Second, the classification allows grouping of patients for therapies. Some regulatory agencies approve drugs or devices indicated for specific seizure types. A new classification should gracefully map to existing indications for drug or device usage. Third, the seizure type grouping might provide a useful link to specific syndromes or etiologies, for example, by noting an association between gelastic seizures and hypothalamic hamartoma or epileptic spasms with tuberous sclerosis. Fourth, the classification allows researchers to better focus their studies on mechanisms of different seizure types. Fifth, a classification provides words to patients to describe their disease. Motivations for revising the 1981 Seizure Classification are listed below.

1. Some seizure types, for example, tonic seizures or epileptic spasms, can have either a focal or generalized onset.
2. Lack of knowledge about the onset makes a seizure unclassifiable and difficult to discuss with the 1981 system.
3. Retrospective seizure descriptions often do not specify a level of consciousness, and altered consciousness, while central to many seizures, is a complicated concept.
4. Some terms in current use do not have high levels of community acceptance or public understanding, such as “psychic,” “partial,” “simple partial,” “complex partial” and “dyscognitive.”
6. Some important seizure types are not included.

Results

Classification of Seizure Types

Figure 1 depicts the basic and Figure 2 the expanded 2017 seizure classification. The two represent the same classification, with collapse of the subcategories to form the basic version. Use of one versus the other depends upon the desired degree of detail. Variations on the individual seizure theme can be added for focal seizure types according to level of awareness.

FIGURE 1 NEAR HERE

Structure of the classification

FIGURE 2 NEAR HERE

The classification chart is columnar, but not hierarchical (meaning that levels can be skipped), so arrows intentionally are omitted. Seizure classification begins with the determination of whether the initial manifestations of the seizure are focal or generalized. The onset may be missed or obscured, in which case the seizure is of unknown onset. The words “focal” and “generalized” at the start of a seizure name are assumed to mean of focal or generalized onset.

For focal seizures, the level of awareness optionally may be included in the seizure type. Awareness is only one potentially important feature of a seizure, but awareness is of sufficient practical importance to justify using it as a seizure classifier. Retained awareness means that the person is aware of self and environment during the seizure, even if immobile. A focal aware seizure (with or without any subsequent classifiers) corresponds to the prior term “simple partial seizure.” A focal impaired awareness seizure (with or without any subsequent classifiers) corresponds to the prior term “complex partial seizure.” **Impaired awareness during any part of the seizure renders it a focal impaired awareness seizure.** Focal seizures also are subgrouped as those with motor and non-motor signs and symptoms at the onset. If both motor and non-motor signs are present at the seizure start, the motor signs will usually dominate, unless non-motor (e.g., sensory) symptoms and signs are very prominent.

Focal aware or impaired awareness seizures optionally may further be characterized by one of the listed motor onset or non-motor onset symptoms, reflecting the first prominent sign or symptom in the seizure, for example, focal impaired awareness automatism seizure. Seizures should be classified by the **earliest** prominent motor onset or non-motor onset feature, except that a focal behavior arrest seizure is one for which cessation of activity is the dominant feature **throughout** the seizure, and any significant impairment of awareness **during the course** of the seizure causes a focal seizure to be classified as having impaired awareness. Classification according to onset has an anatomical basis; whereas, classification by level of awareness has a behavioral basis, justified by the practical importance of impaired awareness. Both methods of

classification are available and can be used in concert. Brief behavioral arrest at the start of a seizure often is imperceptible, and so it is not used as a classifier unless dominant throughout the seizure. The earliest (anatomical) classifier will not necessarily be the most significant behavioral feature of a seizure. For example, a seizure might start with fear and progress to vigorous focal clonic activity resulting in falling. This seizure would still be a focal emotional seizure (with or without impairment of awareness), but free text description of the ensuing features would be very useful.

A focal seizure name can omit mention of awareness when awareness is not applicable or unknown; thereby classifying the seizure directly by motor onset or non-motor onset characteristics. The terms motor onset and non-motor onset may be omitted when a subsequent term generates an unambiguous seizure name.

The classification of an individual seizure can stop at any level: a “focal onset” or “generalized onset” seizure, with no other elaboration, or a “focal sensory seizure,” “focal motor seizure,” “focal tonic seizure” or “focal automatism seizure,” etc. Additional classifiers are encouraged, and their use may depend upon the experience and purposes of the person classifying the seizure. The terms focal onset and generalized onset are for purposes of grouping. No inference is made that each seizure type exists in both groups; including absence seizures in the generalized onset category does not imply existence of “focal absence” seizures.

When the primacy of one versus another key symptom or sign is unclear, the seizure can be classified at a level above the questionably applicable term with additional descriptors of seizure semiology relevant to the individual seizure. Any signs or symptoms of seizures, suggested descriptor terms as listed in the companion paper or free text descriptions can optionally be appended to the seizure type as descriptions, but they do not alter the seizure type.

The seizure type “focal to bilateral tonic-clonic” is a special seizure type, corresponding to the 1981 phrase “partial onset with secondary generalization.” Focal onset bilateral tonic-clonic reflects a propagation pattern of a seizure, rather than a unitary seizure type, but it is such a common and important presentation that the separate categorization was continued. The term “to bilateral” rather than “secondary generalized” was used to further distinguish this focal onset seizure from a generalized onset seizure. The term “bilateral” is used for propagation patterns and “generalized” for seizures that engage bilateral networks from onset.

Seizure activity propagates through brain networks, sometimes leading to uncertainty about whether an event is a unitary seizure or a series of multiple seizures starting from different networks (“multifocal”). A single unifocal seizure can present with multiple clinical manifestations as a result of propagation. The clinician will have to determine (by observation of a continuous evolution or stereotypy from seizure-to-seizure) whether an event is a single seizure or a series of different seizures. When a single focal seizure presents with a sequence of signs and symptoms, then the seizure is named for the initial prominent sign or symptom, reflecting the usual clinical practice of identifying the seizure onset focus or network. For example, a seizure beginning with sudden inability to understand language followed by impaired awareness and clonic left arm jerks would be classified as a “focal impaired awareness (non-motor onset) cognitive seizure” (progressing to clonic left arm jerks). The terms in parentheses are optional. The formal seizure type in this example is determined by the cognitive non-motor onset and presence of altered awareness during any point of the seizure.

Generalized seizures are divided into motor and non-motor (absence) seizures. Further subdivisions are similar to those of the 1981 classification, with addition of myoclonic-atonic seizures, common in epilepsy with myoclonic-atonic seizures (Doose syndrome²⁸), myoclonic-tonic-clonic seizures common in juvenile myoclonic epilepsy,²⁹ myoclonic absence³⁰ and absence seizures with eyelid myoclonia seen in the syndrome described by Jeavons and elsewhere³¹. Generalized manifestations of seizures can be asymmetrical, rendering difficult the distinction from focal onset seizures. The word “absence” has a common meaning, but an “absent stare” is not synonymous with an absence seizure, since arrest of activity also occurs in other seizure types.

The 2017 classification allows appending a limited number of qualifiers to seizures of unknown onset, in order to better characterize the seizure. Seizures of unknown onset may be referred to by the single word “unclassified” or with additional features, including motor, non-motor, tonic-clonic, epileptic spasms and behavior arrest. A seizure type of unknown onset may later become classified as either of focal or generalized onset, but any associated behaviors (e.g., tonic-clonic) of the previously unclassified seizure will still apply. In this regard, the term “unknown onset” is a placeholder - not a characteristic of the seizure, but of ignorance.

Reasons for decisions

The terminology for seizure types is designed to be useful for communicating the key characteristics of seizures and to serve as one of the key components of a larger classification for the epilepsies, which is being developed by a separate ILAE Classification Task Force. The basic framework of seizure classification used since 1981 was maintained.

Focal vs. partial

In 1981, the Commission declined to designate as “focal” a seizure that might involve an entire hemisphere, so the term “partial” was preferred. The 1981 terminology was in a way prescient of the modern emphasis on networks, but “partial” conveys a sense of part of a seizure, rather than a location or anatomical system. The term “focal” is more understandable in terms of seizure onset location.

Focal vs. generalized

In 2010¹ the ILAE defined focal as “originating within networks limited to one hemisphere. They may be discretely localized or more widely distributed. Focal seizures may originate in subcortical structures.” Generalized from onset seizures were defined as “originating at some point within, and rapidly engaging, bilaterally distributed networks.” Classifying a seizure as having apparently generalized onset does not rule out a focal onset obscured by limitations of our current clinical methods, but this is more an issue of correct diagnosis than of classification. Furthermore, focal seizures may rapidly engage bilateral networks, while classification is based on unilateral onset. For some seizure types, for example epileptic spasms, the distinction of a focal versus generalized onset may require careful study of a video-EEG recording or the type of onset may be unknown. A distinction between focal and generalized onset is a practical one, and may change with advances in ability to characterize the onset of seizures.

Focal onset of seizure can be inferred by pattern matching to known focal onset seizures, even when the focal onset is not clear strictly in terms of observable behavior. A seizure is focal, for example, when it starts with déjà vu and then progresses to loss of awareness and responsiveness, lip-smacking and hand-rubbing for a minute. There is nothing intrinsically “focal” in the description, but video-EEGs of countless similar seizures have previously shown focal onsets. If the epilepsy type is known, then the onset can be presumed even if it is unwitnessed, for example, an absence seizure in a person with known juvenile absence epilepsy.

Clinicians have long been aware that so-called generalized seizures, for example, absence seizures with EEG generalized spike-waves, do not manifest equally in all parts of the brain. The Task Force emphasized the concept of bilateral, rather than generalized involvement of some seizures, since seizures can be bilateral without involving every brain network. The bilateral manifestations need not be symmetric. The term “focal to bilateral tonic-clonic” was substituted for “secondarily generalized.” The term “generalized” was maintained for seizures generalized from onset.

Unknown Onset

Clinicians commonly hear about tonic-clonic seizures for which the onset was unobserved. Perhaps, the patient was asleep, alone or observers were too distracted by the manifestations of the seizure to notice the presence of focal features. There should be an opportunity to provisionally classify this seizure, even in the absence of knowledge about its origin. The Task Force therefore allowed further description of seizures of unknown onset when key characteristics, such as tonic-clonic activity or behavior arrest are observed during the course of the seizure. The Task Force recommends classifying a seizure as having focal or generalized onset only when there is a high degree of confidence (e.g., $\geq 80\%$, arbitrarily chosen to parallel the usual allowable beta error) in the accuracy of the determination; otherwise, the seizure should remain unclassified until more information is available.

It may be impossible to classify a seizure at all, either because of incomplete information or because of the unusual nature of the seizure, in which case it is called an unclassified seizure. Categorization as unclassified should only be used for the exceptional situation in which the clinician is confident that the event is a seizure but cannot further classify the event.

Consciousness and awareness

The 1981 classification and the revision in 2010^{1; 10; 32} suggested a fundamental distinction between seizures with loss or impairment of consciousness and those with no impairment of consciousness. Basing a classification upon consciousness (or one of its allied functions) reflects a practical choice that seizures with impaired consciousness should often be approached differently from those with unimpaired consciousness, for example, with respect to allowing driving in adults or interfering with learning. The ILAE chose to retain impairment of consciousness as a key concept in the grouping of focal seizures. However, consciousness is a complex phenomenon, with both subjective and objective components.³³ Multiple different types of consciousness have been described for seizures.³⁴ Surrogate markers³⁵⁻³⁷ for consciousness usually comprise measurements of awareness, responsiveness, memory and a sense of self as distinct from others. The 1981 classification specifically mentioned awareness and responsiveness, but not memory for the event.

Retrospective determination of state of consciousness can be difficult. An untrained classifier might assume that a person must be on the ground, immobile, unaware and unresponsive (e.g., “passed out”) for a seizure to show impaired consciousness. The Task Force adopted state of awareness as a relatively simple surrogate marker for consciousness. “Retained awareness” is considered to be an abbreviation for “seizures with no impairment of consciousness during the event.” **We employ an operational definition of awareness as knowledge of self and environment.** In this context, awareness refers to perception or knowledge of events occurring during a seizure, not to knowledge of whether a seizure occurred. In several languages, “unaware” translates as “unconscious,” in which case changing the seizure designation from “complex partial” to “impaired awareness” will emphasize the importance of consciousness by putting its surrogate directly in the seizure title. In English, “focal aware seizure” is shorter than is “focal seizure without impairment of consciousness” and possibly better understood by patients. As a practical issue, retained awareness usually includes the presumption that the person having the seizure later can recall and validate having retained awareness; otherwise, impaired awareness may be assumed. Exceptional seizures present with isolated transient epileptic amnesia in clear awareness³⁸ but classification of an amnestic seizure

as a focal aware seizure would require clear documentation by meticulous observers. Awareness may be left unspecified when the extent of awareness cannot be ascertained.

Responsiveness may or may not be compromised during a focal seizure.³⁹

Responsiveness does not equate to awareness or consciousness, since some people are immobilized and consequently unresponsive during a seizure, but still able to observe and recall their environment. Additionally, responsiveness often is not tested during seizures. For these reasons, responsiveness was not chosen as a primary feature for seizure classification, although responsiveness can be helpful in classifying the seizure when it can be tested, and degree of responsiveness may be very relevant to the impact of a seizure. The term “dyscognitive” was not carried into the current classification as a synonym for “complex partial” because of lack of clarity and negative public and professional feedback.

Awareness is not a classifier for generalized onset seizures, because the large majority of generalized seizures present with impaired awareness or full loss of consciousness. However, it is recognized that awareness and responsiveness can be at least partially retained during some generalized seizures, for example, with brief absence seizures⁴⁰, including absence seizures with eyelid myoclonias or myoclonic seizures.

Etiologies

A classification of seizure types can be applied to seizures of different etiologies. A post-traumatic seizure or a reflex seizure may be focal with or without impairment of awareness. Knowledge of the etiology, for instance, presence of a focal cortical dysplasia, can aid in classification of the seizure type. Any seizure can become prolonged, leading to status epilepticus of that seizure type.

Supportive information

As part of the diagnostic process, a clinician will commonly use supportive evidence to help to classify a seizure, even though that evidence is not part of the classification. Such evidence may include videos brought in by family, EEG patterns, lesions detected by

neuroimaging, laboratory results such as detection of anti-neuronal antibodies, gene mutations, or an epilepsy syndrome diagnosis known to be associated with either focal or generalized seizures or both, such as Dravet syndrome. The seizures usually can be classified on the basis of symptoms and behavior, provided that good subjective and objective descriptions are available. Use of any available supportive information to classify the seizure is encouraged. Availability of supportive information may not exist in the resource-poor parts of the world, which may lead to a less specific, but still correct classification.

ICD 9, 10, 11, 12

The World Health Organization International Classification of Diseases (ICD) is used for inpatient and outpatient diagnoses, billing and many other purposes^{41; 42}. Concordance between ICD epilepsy diagnoses and ILAE seizure types is desirable for clarity and consistency. This is possible only to a limited extent with existing ICD terms, since ICD 9, 10, 11 are already formulated. The ILAE proposals will always lead ICD standards. ICD 9 and 10 make use of old seizure terminology, including terms such as petit mal and grand mal. ICD11 does not name seizure types at all, but focuses on epilepsy etiologies and syndromes, as do ILAE epilepsy classifications¹. For this reason, there is no conflict between our proposed seizure type classification and ICD11. Efforts can be made to incorporate new classifications of seizure types and syndromes into the development of ICD12.

Discussion

Discontinued terms

Simple/complex partial: After approximately 35 years of use, the terms “simple partial seizure” and “complex partial seizure” may be missed by some clinicians. The reasons for changing are three. First, a decision was previously made¹ to globally change partial to focal. Second, “complex partial” has no intrinsic meaning to the public. The phrase “focal impaired

awareness” can convey meaning to a lay person with no knowledge of seizure classification. Third, the words “complex” and “simple” can be misleading in some contexts. Complex seems to imply that this seizure type is more complicated or difficult to understand than other seizure types. Calling a seizure “simple” may trivialize its impact to a patient who does not find the manifestations and consequences of the seizures to be at all simple.

Convulsion: The term “convulsion” is a popular, ambiguous and unofficial term used to mean substantial motor activity during a seizure. Such activity might be tonic, clonic, myoclonic or tonic-clonic. In some languages, convulsions and seizures are considered synonyms and the motor component is not clear. The word “convulsion” is not part of the 2017 seizure classification, but will undoubtedly persist in popular usage.

Added terms

Aware/impaired awareness: As discussed above, these terms designate knowledge of self and environment during a seizure.

Hyperkinetic: Hyperkinetic seizures have been added to the focal seizure category. Hyperkinetic activity comprises agitated thrashing or leg pedaling movements. Hypermotor is an earlier term introduced as part of a different proposed classification by Lüders and colleagues in 1993.⁴³ The term hypermotor, which contains both Greek and Roman roots, was supplanted in the 2001 ILAE glossary⁴⁴ and 2006 report² by “hyperkinetic,” and to be both etymologically and historically consistent, “hyperkinetic” was chosen for the 2017 classification.

Cognitive: This term replaces “psychic” and refers to specific cognitive impairments during the seizure, for example, aphasia, apraxia or neglect. The word “impairment” is implied since seizures never enhance cognition. A cognitive seizure can also comprise positive cognitive phenomena, such as déjà vu, jamais vu, illusions or hallucinations.

Emotional: A focal non-motor seizure can have emotional manifestations, such as fear or joy. The term also encompasses affective manifestations with the appearance of emotions

occurring without subjective emotionality, such as may occur with some gelastic or dacrystic seizures.

New focal seizure types: Some seizure types that previously were described only as generalized seizures now appear under seizures of focal, generalized and unknown onset. These include epileptic spasms, tonic, clonic, atonic, myoclonic seizures. The list of motor behaviors constituting seizure types comprises the most common focal motor seizures, but other less common types, for example, focal tonic-clonic, may be encountered. Focal automatisms, autonomic, behavior arrest, cognitive, emotional and hyperkinetic are new seizure types. Focal to bilateral tonic-clonic seizure is a new type as the renamed secondarily generalized seizure.

New generalized seizure types: Relative to the 1981 classification, new generalized seizure types include: absence with eyelid myoclonia, myoclonic-atonic, and myoclonic-tonic-clonic (although clonic onset of tonic-clonic seizures was mentioned in the 1981 publication). Seizures with eyelid myoclonia could logically have been placed under the motor category, but since eyelid myoclonia are most significant as features of absence seizures, seizures with eyelid myoclonia were placed in the non-motor/absence category. Seizures with eyelid myoclonia may even rarely display focal features⁴⁵. Similarly, myoclonic absence seizures potentially have features of both absence and motor seizures, and could have been placed in either group. Epileptic spasms are seizures represented in focal, generalized and unknown onset categories, and the distinction may require video-EEG recording. The term “epileptic” is implied for every seizure type, but explicitly stated for epileptic spasms, because of the ambiguity of the single word “spasms” in neurological use.

What is different from the 1981 classification?

Table 1 summarizes the changes in the ILAE 2017 seizure type classification from the 1981 classification. Note that several of these changes were already incorporated into the 2010 revision of terminology and subsequent revisions.^{1; 32}.

TABLE 1 NEAR HERE

Compared to the 1981 classification, certain seizure types now appear in multiple categories. Epileptic spasms can be of focal, generalized or unknown onset. Represented both in focal and generalized columns are atonic, clonic, myoclonic and tonic seizures, although the pathophysiology of these seizure types may differ for the focal onset versus generalized onset seizure type of that name.

A companion paper provides guidance on how to apply the 2017 classification. Employment of the 2017 classification in the field for a few years likely will motivate minor revisions and clarifications.

Key Points

- The ILAE has constructed a revised classification of seizure types. The classification is operational and not based on fundamental mechanisms.
- Reasons for revision include clarity of nomenclature, ability to classify some seizure types as either focal or generalized, and classification when onset is unknown.
- Seizures are divided into seizures of focal, generalized, unknown onset, with subcategories of motor, non-motor, with retained or impaired awareness for focal seizures.

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We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

References

1. Berg AT, Berkovic SF, Brodie MJ, et al. Revised terminology and concepts for organization of seizures and epilepsies: report of the ILAE Commission on Classification and Terminology, 2005-2009. *Epilepsia* 2010;51:676-685.
2. Engel J, Jr. Report of the ILAE classification core group. *Epilepsia* 2006;47:1558-1568.
3. Gastaut H, Magnus O, Caveness W, et al. A proposed international classification of epileptic seizures. *Epilepsia* 1964;5:297-306.

4. Gastaut H. Classification of the epilepsies. Proposal for an international classification. *Epilepsia* 1969;10:Suppl:14-21.
5. Blumenfeld H. What is a seizure network? Long-range network consequences of focal seizures. *Adv Exp Med Biol* 2014;813:63-70.
6. Centeno M, Carmichael DW. Network Connectivity in Epilepsy: Resting State fMRI and EEG-fMRI Contributions. *Front Neurol* 2014;5:93.
7. Proposal for revised clinical and electroencephalographic classification of epileptic seizures. From the Commission on Classification and Terminology of the International League Against Epilepsy. *Epilepsia* 1981;22:489-501.
8. Berg AT. Classification and epilepsy: the future awaits. *Epilepsy Curr* 2011;11:138-140.
9. Berg AT, Scheffer IE. New concepts in classification of the epilepsies: entering the 21st century. *Epilepsia* 2011;52:1058-1062.
10. Korff CM, Scheffer IE. Epilepsy classification: a cycle of evolution and revolution. *Curr Opin Neurol* 2013;26:163-167.
11. Berg AT, Blackstone NW. Concepts in classification and their relevance to epilepsy. *Epilepsy Res* 2006;70 Suppl 1:S11-19.

12. Engel J, Jr. A proposed diagnostic scheme for people with epileptic seizures and with epilepsy: report of the ILAE Task Force on Classification and Terminology. *Epilepsia* 2001;42:796-803.
13. Engel J, Jr. ILAE classification of epilepsy syndromes. *Epilepsy Res* 2006;70 Suppl 1:S5-10.
14. Berg AT, Cross JH. Classification of epilepsies and seizures: historical perspective and future directions. *Handb Clin Neurol* 2012;107:99-111.
15. Luders HO, Amina S, Baumgartner C, et al. Modern technology calls for a modern approach to classification of epileptic seizures and the epilepsies. *Epilepsia* 2012;53:405-411.
16. Luders H, Acharya J, Baumgartner C, et al. Semiological seizure classification. *Epilepsia* 1998;39:1006-1013.
17. Panayiotopoulos CP. The new ILAE report on terminology and concepts for the organization of epilepsies: critical review and contribution. *Epilepsia* 2012;53:399-404.
18. Panayiotopoulos CP. The new ILAE report on terminology and concepts for organization of epileptic seizures: a clinician's critical view and contribution. *Epilepsia* 2011;52:2155-2160.

19. Gomez-Alonso J, Bellas-Lamas P. [The new International League Against Epilepsy (ILAE) classification of epilepsies: a step in the wrong direction?]. *Rev Neurol* 2011;52:541-547.
20. Shorvon SD. The etiologic classification of epilepsy. *Epilepsia* 2011;52:1052-1057.
21. Beghi E. The concept of the epilepsy syndrome: how useful is it in clinical practice? *Epilepsia* 2009;50 Suppl 5:4-10.
22. Tuxhorn I, Kotagal P. Classification. *Semin Neurol* 2008;28:277-288.
23. Luders H, Acharya J, Baumgartner C, et al. A new epileptic seizure classification based exclusively on ictal semiology. *Acta Neurol Scand* 1999;99:137-141.
24. Seino M. Classification criteria of epileptic seizures and syndromes. *Epilepsy Res* 2006;70 Suppl 1:S27-33.
25. Fisher RS, Boas WV, Blume W, et al. Epileptic seizures and epilepsy: Definitions proposed by the International League against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE). *Epilepsia* 2005;46:470-472.
26. Brodtkorb E. Common imitators of epilepsy. *Acta Neurol Scand Suppl* 2013;5-10.
27. Trinka E, Cock H, Hesdorffer D, et al. A definition and classification of status epilepticus--Report of the ILAE Task Force on Classification of Status Epilepticus. *Epilepsia* 2015;56:1515-1523.

28. Kelley SA, Kossoff EH. Doose syndrome (myoclonic-astatic epilepsy): 40 years of progress. *Dev Med Child Neurol* 2010;52:988-993.
29. Wolf P, Yacubian EM, Avanzini G, et al. Juvenile myoclonic epilepsy: A system disorder of the brain. *Epilepsy Res* 2015;114:2-12.
30. Verrotti A, Greco R, Chiarelli F, et al. Epilepsy with myoclonic absences with early onset: a follow-up study. *J Child Neurol* 1999;14:746-749.
31. Striano S, Capovilla G, Sofia V, et al. Eyelid myoclonia with absences (Jeavons syndrome): a well-defined idiopathic generalized epilepsy syndrome or a spectrum of photosensitive conditions? *Epilepsia* 2009;50 Suppl 5:15-19.
32. Scheffer IE. Epilepsy: a classification for all seasons? *Epilepsia* 2012;53 Suppl 2:6-9.
33. Ali F, Rickards H, Cavanna AE. The assessment of consciousness during partial seizures. *Epilepsy Behav* 2012;23:98-102.
34. Luders H, Amina S, Bailey C, et al. Proposal: Different types of alteration and loss of consciousness in epilepsy. *Epilepsia* 2014;in press.
35. Cavanna AE, Monaco F. Brain mechanisms of altered conscious states during epileptic seizures. *Nat Rev Neurol* 2009;5:267-276.
36. Blumenfeld H. Impaired consciousness in epilepsy. *Lancet Neurol* 2012;11:814-826.

37. Blumenfeld H, Taylor J. Why do seizures cause loss of consciousness? *Neuroscientist* 2003;9:301-310.
38. Felician O, Tramon E, Bartolomei F. Transient epileptic amnesia: Update on a slowly emerging epileptic syndrome. *Rev Neurol (Paris)* 2015;171:289-297.
39. Yang L, Shklyar I, Lee HW, et al. Impaired consciousness in epilepsy investigated by a prospective responsiveness in epilepsy scale (RES). *Epilepsia* 2012;53:437-447.
40. Porter RJ, Penry JK. Responsiveness at the onset of spike-wave bursts. *Electroencephalogr Clin Neurophysiol* 1973;34:239-245.
41. Bergen DC, Beghi E, Medina MT. Revising the ICD-10 codes for epilepsy and seizures. *Epilepsia* 2012;53 Suppl 2:3-5.
42. Jette N, Beghi E, Hesdorffer D, et al. ICD coding for epilepsy: past, present, and future--a report by the International League Against Epilepsy Task Force on ICD codes in epilepsy. *Epilepsia* 2015;56:348-355.
43. Luders HO, Burgess R, Noachtar S. Expanding the international classification of seizures to provide localization information. *Neurology* 1993;43:1650-1655.
44. Blume WT, Luders HO, Mizrahi E, et al. Glossary of descriptive terminology for ictal semiology: report of the ILAE task force on classification and terminology. *Epilepsia* 2001;42:1212-1218.

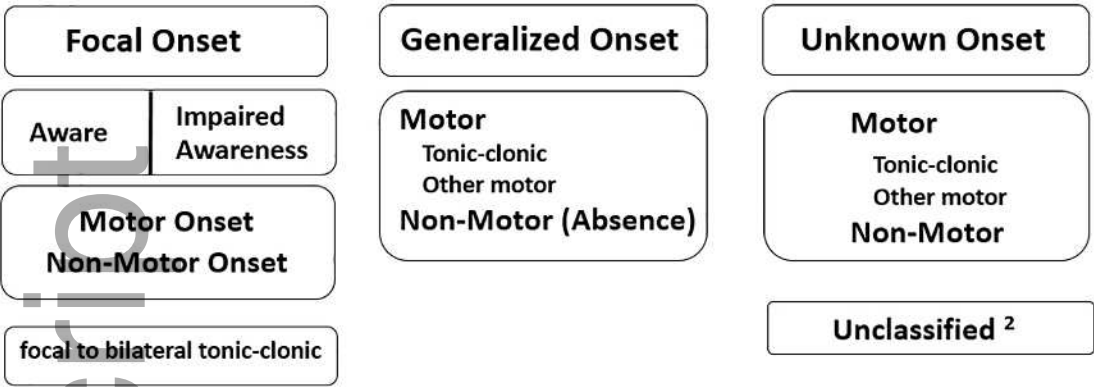
45. Mourente-Diaz S, Montenegro MA, Lowe JP, et al. Unusual focal ictal pattern in children with eyelid myoclonia and absences. *Pediatr Neurol* 2007;37:292-295.

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Table 1: Changes in Seizure Type Classification from 1981 to 2017

1. Change of “partial” to “focal.”
2. Certain seizure types can be either of focal, generalized or unknown onset.
3. Seizures of unknown onset may have features that can still be classified.
4. Awareness is used as a classifier of focal seizures.
5. The terms dyscognitive, simple partial, complex partial, psychic, secondarily generalized were eliminated.
6. New focal seizure types include automatisms, autonomic, behavior arrest, cognitive, emotional, hyperkinetic, sensory and focal to bilateral tonic-clonic seizures. Atonic, clonic, epileptic spasms, myoclonic and tonic seizures can be either focal or generalized.
7. New generalized seizure types include absence with eyelid myoclonia, myoclonic absence, myoclonic-tonic-clonic, myoclonic-atonic, epileptic spasms.

ILAE 2017 Classification of Seizure Types Basic Version ¹

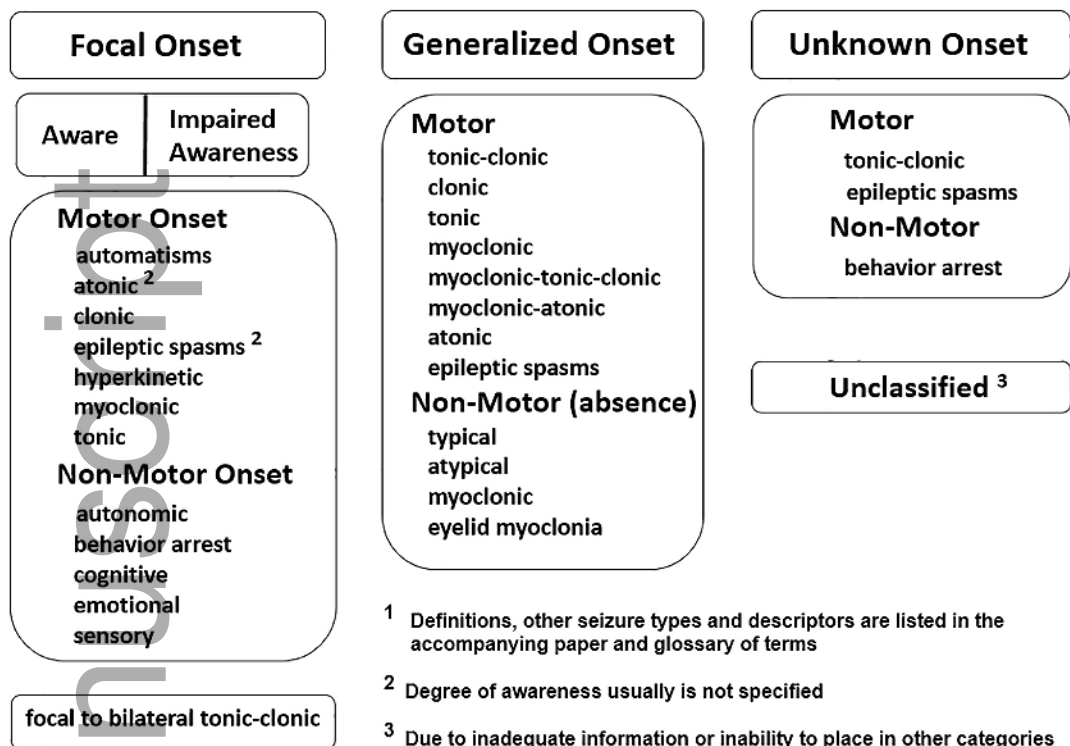


¹ Definitions, other seizure types and descriptors are listed in the accompanying paper & glossary of terms

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

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ILAE 2017 Classification of Seizure Types Expanded Version ¹



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