

ILAE Classification of Seizures and Epilepsies: An Update for the Pediatrician

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The International League Against Epilepsy (ILAE) recently published an updated classification of seizures and epilepsy. This updated classification aims to have a better organized classification and ensure better understanding of terms, in addition to including new seizure types. As both seizures and epilepsy are important childhood conditions, we herein list some of the important aspects of the updated classification for the benefit of the general pediatricians. The full classifications are available at the ILAE website.

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Seizures are a common pediatric problem, and of considerable interest to pediatricians [1,2]. Classification of seizures and epilepsy is the cornerstone in the evaluation and management of seizures. It helps in understanding the types of seizure, identifying and labeling the type of epilepsy, and grouping similar entities into specific syndromes – thereby guiding antiepileptic therapy and patient counseling. Epilepsy classification is primarily used for the diagnosis of patients, but it is also critical for research, communication among clinicians and researchers, and development of antiepileptic therapies [3].

The International League Against Epilepsy (ILAE) has been playing a pivotal role in classification of epilepsy [4-7]. The 1981 and 1989 ILAE classifications were a major breakthrough in understanding the types and classification of seizures and epilepsy [4,5]. Since then, multiple modifications and revisions have been proposed [6,7]. In 2017, ILAE published an updated classifications of both seizures [8] and epilepsies [3]; these being the “first new official papers on classification from the ILAE since 1989” [9]. With advancement in technology and research, new insight has been gained in understanding the phenotypic pattern and the basic mechanism of seizure, thus making revision a necessary process. The stated purpose of revisions is to include new seizure types, have a better organized classification and to enable usage of appropriate terms for better understanding [3].

We herein list some of the important aspects of the revised classification for the benefit of the readers – these include, among others, new focal seizure types which were

earlier in generalized category alone (*e.g.*, epileptic spasm could be focal as well as generalized), new generalized types (like myoclonic atonic and epileptic spasm), and classifying focal seizure by its “first clinical manifestation” [8]. The full documents are available at the ILAE website (<https://www.ilae.org/guidelines/definition-and-classification>).

The new classification framework of epilepsy is a multilevel classification with four main components; three of them sequential *viz.*, (i) the seizure type, followed by (ii) the epilepsy type, and then (iii) the epilepsy syndrome. The fourth component, identifying etiology, is an overarching activity, continuing at each individual step (**Fig. 1**). The framework is designed to enable classification of epilepsy in different clinical settings implying that patient characterization will be possible at every level, depending on the resources available to the clinician making the diagnosis [3]. Supporting information *viz.*, Video record, Electro-encephalography (EEG), neuroimaging, gene mutations and autoimmune panel, if available, are to be utilized in classifying epilepsy type and/or epilepsy syndrome [3].

CLASSIFYING SEIZURES

The new basic 2017 classification of seizure is based on three key features *viz.*, (i) locus of seizure origin in brain; (ii) level of awareness during seizure; and, (iii) other features. The basic operational seizure type classification includes [8]: Focal onset seizures (Aware/Impaired awareness; Motor/Non-motor onset; and Focal to bilateral tonic-clonic); Generalized onset (Motor/Non-

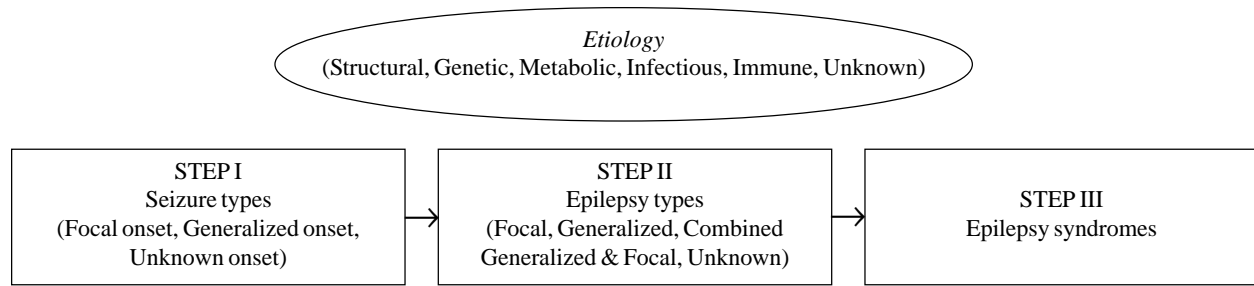


FIG. 1 Schematic representation of the multi-level classification of seizure and epilepsy (ILAE, 2017).

motor (Absence)); and Unknown onset (Motor/Non-motor; Unclassified).

Inclusion of new seizure types: The new seizure types included in the classification are enlisted in **Box 1**.

Clarification of impairment of consciousness: Consciousness is a complex phenomenon with both subjective and objective components. In the 2017 classification, awareness has been chosen to be the best surrogate marker of consciousness, and is simpler to evaluate [8]. Awareness is operationally defined as “knowledge of self and environment” [8].

Re-classification of certain seizure types into either focal or generalized onset: Seizures are classified by earliest prominent motor or non-motor features; even though, uncertainty is present in every seizure classification [8]. The term ‘bilateral’ is used for propagation patterns and ‘generalized’ for seizures that engage bilateral networks from onset. Few seizure types like epileptic spasms, myoclonic, tonic, tonic-clonic, clonic, atonic, which were previously included only in generalized-onset seizures are now also included in focal-onset seizures [8].

Classification of seizure of unknown onset: Situations in which patient is alone or asleep or the attender is not able to describe the seizure onset clearly is classified as Seizure of unknown onset. If clinician is confident that an

event is seizure but cannot classify it due to incomplete information, it is grouped under unclassified seizures. In the 2017 classification, Seizure of unknown onset is further classified as tonic-clonic, epileptic spasms or behavior arrest depending upon the predominant motor or non-motor activity noticed during the episode [8]. This is essential in guiding treatment, and for reclassification into focal or generalized onset with future episodes. In this regard, the term ‘unknown onset’ is a placeholder – not a characteristic of the seizure [8].

New terms for old ones: Some of the terms used in seizure-classification lack community acceptance or public understanding. The terms dyscognitive, simple partial, complex partial, psychic, and secondarily generalized were eliminated. The term ‘partial’ was replaced by ‘focal’, as the term partial conveys a sense of part of a seizure, rather than a location or anatomic system. ‘Focal to bilateral tonic-clonic’ replaced ‘focal seizure with secondary generalization,’ as this term can better reflect the propagation pattern of a seizure (**Table I**) [8].

Updated glossary of seizure terms: Glossary of seizure terms has also been updated and it includes new definition for terms like emotional seizure, eyelid myoclonia, myoclonic atonic, behavior arrest, unaware, and unclassified seizure [10]. Common descriptors used to describe seizures have also been standardized in the new 2017 classification [10].

BOX 1 NEWER TERMINOLOGIES FOR SEIZURE TYPE IN ILAE SEIZURE CLASSIFICATION, 2017

Focal seizures

Motor: Epileptic spasms, myoclonic, tonic, tonic clonic, clonic, atonic, hyper-kinetic, automatism

Non-motor: Behavior arrest, emotional

Focal to bilateral tonic-clonic

Generalized seizures

Absence with eyelid myoclonia, epileptic spasms, myoclonic-atonic, and myoclonic-tonic-clonic

CLASSIFYING EPILEPSY TYPES

A new group of combined generalized and focal epilepsy has been introduced in the epilepsy type. Idiopathic generalized epilepsy was renamed as Genetic generalized epilepsy, which includes Childhood absence epilepsy, juvenile absence epilepsy, Juvenile myoclonic epilepsy and Generalized tonic-clonic seizures alone [3]. The terms ‘Epileptic encephalopathy’ and ‘Developmental encephalopathy’ have been redefined, and ‘malignant’ and ‘catastrophic’ are omitted. The term ‘benign’ used in

TABLE I NEW TERMS INTRODUCED IN THE ILAE SEIZURE CLASSIFICATION, 2017

<i>Old terms</i>	<i>New terms</i>
Partial	Focal
Simple partial	Focal aware
Complex partial	Focal impaired awareness
Psychic	Cognitive
Secondary generalized tonic-clonic	Focal to bilateral tonic-clonic
Arrest, Freeze, Pause, Interruption	Behavior arrest
Dyscognitive	Focal impaired awareness
Astatic	(Focal or generalized) atonic
Grand mal	Generalized tonic clonic, Focal to bilateral tonic clonic, Unknown onset tonic clonic
Infantile spasm	Epileptic spasm
Psychomotor	Focal impaired awareness

some epilepsy syndromes like BECTS (Benign epilepsy with centro-temporal spikes) is now replaced by 'self-limited' or 'pharmaco-responsive' depending on the situations [3].

Epilepsy Syndromes

Diagnosis of an Epilepsy syndrome, if possible, is the third level of diagnosis [10]. An epilepsy syndrome is diagnosed on the basis of all or some of age at presentation, seizure type, EEG findings, etiological substrate, neuroimaging, genetic analyses, occurring in a typical pattern [10]. Recognition of a syndrome helps in determining etiology, evaluating for co-morbidity, deciding management, and conveying prognosis.

Labeling Etiology

This epilepsy classification lays stress on determining etiology at all stages along the diagnostic process, starting from the initial presentation and evaluation. Six major etiological categories have been recognized in the classification (**Fig. 1**), with an understanding that a patient's epilepsy may be classified into more than one etiological group. An example would be a 'structural' cause (cortical tuber) in a 'genetic' condition (tuberous sclerosis).

CONCLUSIONS

These new classifications of seizures and the epilepsies are likely to lead to improved understanding of seizure

etiology, making appropriate diagnosis and will guide targeted therapies to the patient. This classification may also help in greater ease of communication about seizure types among clinicians, the non-medical community, and researchers. The simultaneously published instruction manual on applying the seizure classification terminology will immensely assist in everyday clinical practice [9]. A companion piece to present these concepts for people with epilepsy and their caregivers is also available [10].

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