



Review article

The new definition and classification of seizures and epilepsy

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ABSTRACT

This review discusses the updated classifications of seizures and the epilepsies, which were recently published by the International League Against Epilepsy (ILAE). While it is always a challenge to learn a new classification system, particularly one that has remained essentially unchanged for over three decades, these new classifications allow for the inclusion of some previously unclassifiable seizure types and utilize more intuitive terminology. In this review, we specifically discuss the use of these new classifications for patients, clinicians, and researchers.

1. Introduction

Classifications for seizures and epilepsy were previously constructed in 1981 (ILAE, 1981), 1985 (ILAE, 1985) and 1989 (ILAE, 1989). Having seizure and epilepsy classifications are exceedingly important for the clinicians and care teams, patients and families, and researchers. From a patient standpoint, it provides a namable diagnosis/etiology and improves understanding. For clinicians and the patient's care team, these classifications enhance communication and discussion. From a research standpoint, having these classifications enables investigation of drug or surgical treatments, responses, and typical clinical courses for different types of seizures and epilepsy.

Based on decades of accumulated clinical experience, the International League Against Epilepsy (ILAE) commissioned a new operational classification of seizure types and epilepsies. The new 2017 classifications, when compared to the 1981/1985/1989 classifications, utilize alternative terms and contain several important additions. These changes improve the intuitiveness, transparency and versatility of the classifications, and allow for inclusion/classification of previously unclassifiable seizure and epilepsy types.

2. History of seizure and epilepsy classifications

Seizure classifications have existed for centuries, with the first modern classification proposed in 1964 (Gastaut et al., 1964), and international use of this classification popularized in 1970 (Gastaut, 1970). Prior to 1970, distinction between seizure types and epilepsy types was not frequently made. This distinction is important as a large

percentage of patients with seizures are unclassifiable as to a specific epilepsy type according to the 1989 criteria (32% and 39% in two studies of 100 and 300 consecutive patients, respectively) (Seino, 2006). For these patients, their seizure type can and should still be classified, emphasizing the utility of having separate classifications.

The seizure classification was first updated in 1981, prompted by the widespread use of video electroencephalography (EEG), which had impacted clinical practice. The 1981 seizure classification promulgated the terms “simple partial”, “complex partial”, “generalized”, and “unclassifiable” that have been in use until today (ILAE, 1981). At the beginning of the 21st century, the ILAE sought to update the seizure classification again. Extensive discussions resulted in a decision in 2010 to maintain the 1981 classification of seizures with minor changes (Berg and Scheffer, 2011; Engel, 2001, 2006). The classification of the epilepsies were partially updated in 1985/1989 (ILAE, 1985, 1989) and then again in 2010 (Berg et al., 2010; Berg and Scheffer, 2011), with the 2010 revision being an intermediate stage aiming towards a final accepted epilepsy classification.

While it has been clear for some time that updated classifications were needed, consensus was difficult to reach. This need prompted the ILAE to assemble a new task force, which developed and published a new definition of epilepsy in 2014 (Fisher et al., 2014), then final classification of seizures (Fisher et al., 2017a,b) and the epilepsies (Scheffer et al., 2017) in 2017. Interestingly, some of the alternative terminology adopted in the 2017 seizure classification (focal instead of partial, and the term “aware”) were terms considered, debated, and contentious in 1981, and have continued to stir controversy (ILAE, 1985).

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ILAE 2017 Classification of Seizure Types Basic Version

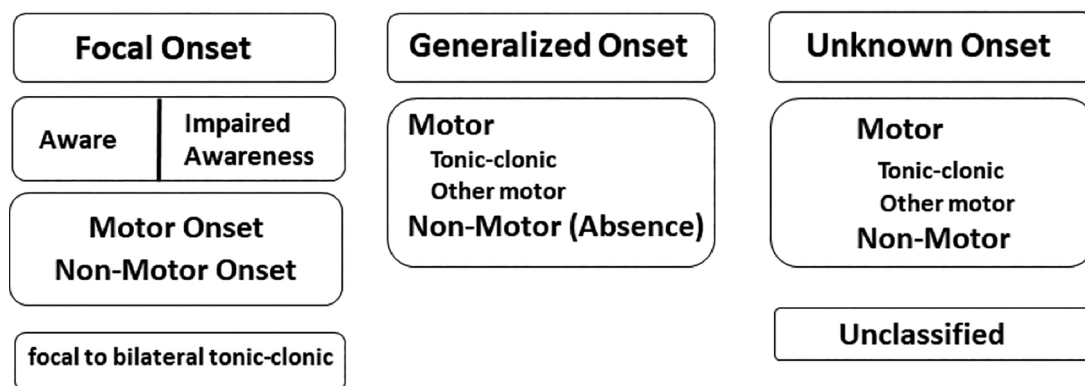


Fig. 1. ILAE 2017 classification of seizure types basic version.

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Given the advances in neuroimaging, genomic technologies and molecular biology, it was proposed to move towards a scientifically based classification of seizures, but after intense discussion it was felt this was not currently tenable. The new classification of seizures continues to rely on semiology, EEG and occasionally supplementary information from imaging. Classification of the epilepsy type and epilepsy syndrome does utilize more of these recent advances, being aided significantly by genetics, lab findings and neuroimaging findings, though still with a major focus throughout the diagnostic process on etiology.

3. Definition of seizure and epilepsy

An epileptic seizure is defined conceptually as: “a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.” This definition was updated most recently by the ILAE in 2005 (Fisher et al., 2005) and was not changed in 2014 when the definition of epilepsy was updated (Fisher et al., 2014).

Epilepsy exists when someone has an epileptic seizure and their brain “demonstrates a pathologic and enduring tendency to have recurrent seizures” (Fisher et al., 2014). More specifically, epilepsy is diagnosed when an individual has: 1) at least two unprovoked or reflex seizures > 24 h apart, 2) one unprovoked or reflex seizure and a probability of having another seizure similar to the general recurrence risk after two unprovoked seizures ($\geq 60\%$) over the next 10 years, or 3) an epilepsy syndrome (Fisher et al., 2014). Greater than 60% was chosen as this is the lower limit of the confidence interval for someone with two unprovoked seizures having another seizure (Hauser et al., 1982). Examples of evidence that increases the probability of having additional seizures include: 1) epileptiform activity on EEG or 2) a potential epileptogenic abnormality on brain imaging. A recent study (PRO-LONG) evaluated patients who were diagnosed with epilepsy according to this new definition (2014) vs. the old definition. PRO-LONG found the long-term recurrence of seizures in patients diagnosed with the new definition was 83.6% at 10 years, thus supporting treatment and a diagnosis of epilepsy after only one seizure (Beretta et al., 2017). Thus, if the clinical picture, EEG or imaging findings increase the probability of another seizure to $\geq 60\%$, then these individuals are defined as having epilepsy, and should be, as clinically they are statistically equivalent in their recurrence risk to those who have had two or more unprovoked seizures.

Epilepsy is considered “resolved” under the following circumstances: 1) in a patient with an age-dependent epilepsy syndrome who is older than the age in which this syndrome is active, or 2) a patient who has been seizure free for ≥ 10 years and has been off all anti-

seizure medications for ≥ 5 years.

Epilepsy was redefined by the ILAE as a “disease” and not a “disorder” (Fisher et al., 2014). The term “disease” better emphasizes to patients, clinicians and society the importance and impact of epilepsy. Although some might worry that “disease” carries more stigma, important diseases such as cancer, diabetes, and heart disease, are all diseases, not disorders.

The definition of epilepsy, a diagnosis for an individual, and a decision about whether to treat, are linked, but diagnosis of epilepsy does not mandate treatment and not all epilepsy requires treatment. Treatment decisions must be individualized. The changes made in 2014, as outlined above, compared to the prior definition from 2005, align the definition and resolution of epilepsy with clinical practice.

4. Classification of seizure types

The new classification has both a basic and expanded version, depending upon the needs and expertise of the individual utilizing the classification. The basic version is a contracted form of the expanded version. The expectation is the basic version will be more useful for doctors in general practice, pediatricians, non-neurologists and general neurologists, nurses and health care workers, while the expanded version will aid epileptologists/neurophysiologists and researchers. Though, clearly there will be researchers who only want the information the basic classification contains, and others mentioned above may prefer the detail of the expanded classification. It should be noted that one should be reasonably sure an event is epileptic prior to attempting to classify a seizure and epilepsy type.

The classification applies to seizures in adults as well as children, except for neonatal seizures—for which there is a separate classification. This classification does not attempt to classify subclinical electrographic seizures (Fig. 1).

4.1. Basic classification

Seizures are defined by onset as: focal, generalized, unknown, or unclassifiable. “Focal” is synonymous with the old term “partial.” The term “generalized” has been retained unchanged. A generalized onset seizure is when both hemispheres (potentially asymmetrically) are activated at onset of the seizure, according to behavior and EEG. “Unknown” onset refers to when the onset is unknown but other manifestations are known. This is clarified further below. “Unclassified” remains as a category, although usage may decrease given the addition of additional seizure types and the “unknown onset” category. Few events are clearly seizures, yet unclassifiable (Fig. 2).

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