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CRITICAL REVIEW AND INVITED COMMENTARY

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Critique of the 2017 epileptic seizure and epilepsy classifications

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Summary

This article critiques the International League Against Epilepsy (ILAE) 2015-2017 classifications of epilepsy, epileptic seizures, and status epilepticus. It points out the following shortcomings of the ILAE classifications: (1) they mix semiological terms with epileptogenic zone terminology; (2) simple and widely accepted terminology has been replaced by complex terminology containing less information; (3) seizure evolution cannot be described in any detail; (4) in the four-level epilepsy classification, level two (epilepsy category) overlaps almost 100% with diagnostic level one (seizure type); and (5) the design of different classifications with distinct frameworks for newborns, adults, and patients in status epilepticus is confusing. The authors stress the importance of validating the new ILAE classifications and feel that the decision of *Epilepsia* to accept only manuscripts that use the ILAE classifications is premature and regrettable.

KEYWORDS
classification, epileptogenic zone, etiology, semiology

1 | INTRODUCTION

In 2015-2017, the International League Against Epilepsy (ILAE) published three new classification schemes for epilepsy,[1] epileptic seizures,[2,3] and status epilepticus.[4]

In the paragraphs below, we will point out the main features a rational classification of epileptic seizures and of epilepsies should satisfy and we will analyze whether the classifications listed above accomplish these conditions. The main part of the article discusses the fundamental deficiencies of the ILAE classifications. More detailed critiques are presented in Appendix 1.

Key Points

- The main shortcomings of the latest ILAE classification of seizures and epilepsies are presented
- The advantages of an alternative 4-dimension classification system are discussed
- The importance of using a similar framework for the classification of seizures in newborns and adults as also for status epilepticus is stressed

2 | CRITIQUE OF THE ILAE SEIZURE CLASSIFICATION

2.1 | A classification system should ideally be universally accepted

Classifications that are universally approved provide a common language facilitating communication among clinicians, researchers, and students. The ILAE is making great efforts to have their latest classifications widely adopted. However, that a classification is used universally does not necessarily mean that it is a good classification.

2.2 | Classifications should use the most important characteristics of the object to be classified as the basis of the classification

For example, Linnaeus in the 18th century realized that the most important information contained in plants and animals was evolution, and he used that characteristic to develop a highly successful biological classification of animals and plants. Epileptic seizures have several characteristics that provide essential information for the management of patients and should be used to classify epileptic seizures.

2.2.1 | Clinical characteristics of the seizure

Clinical features of the seizure onset and evolution (ictal and immediate postictal semiology) can provide valuable information about seizure type and location of the epileptogenic zone, and for this reason, they are commonly used in classification schemes. The 2017 ILAE seizure classification relies heavily on semiological parameters to classify seizures ("motor" and "nonmotor," "tonic," "myoclonic," "clonic," etc). In addition, it distinguishes seizures of "focal onset," "generalized onset," or "unknown onset." These last subdivisions, however, are not based on semiology but rather on electroanatomical characteristics (electroencephalogram [EEG], magnetic resonance imaging [MRI], and other tests). The ILAE classification of seizures[2,3] therefore mixes semiological data with information from other sources about location of seizure onset and epileptogenic zones, combining phenomenology with pathophysiology. This confusion can be easily avoided by including a pure semiological seizure classification[5–12] in a multidimensional epilepsy classification[13–15] in which seizure semiology and epileptogenic zone are independent dimensions classified by different parameters. In such a classification, semiological modifiers (including somatotopic modifiers such as right, left, bilateral, arm, leg, and face) always refer to the corresponding semiological category, and this can easily be differentiated from the conclusion about location of the epileptogenic zone.

2.2.2 | Seizure evolution

Another critically important characteristic of epileptic seizures is the evolution of symptomatology, as this has far-reaching implications for localizing the epileptogenic zone. In a standardized classification system, it is possible to describe seizure evolution in detail by dividing epileptic symptoms into distinct components and then listing the different components according to temporal occurrence, linked by arrows to show the order in which they occur.[15]

Example: (1) left visual aura → (2) left versive → (3) bilateral tonic clonic seizure.

However, the ILAE seizure classification[2] includes only a very limited repertoire of seizure evolutions, namely: focal to bilateral tonic–clonic seizure, tonic–clonic seizure, myoclonic–tonic seizure, myoclonic-atonic seizure, and clonic–tonic–clonic seizure.

This limited number of possible seizure evolutions in the ILAE seizure classification makes it impossible to express in detail the evolution of most focal epileptic seizures, and this important information is lost.

A true semiological seizure classification allows neurologists to already have an anatomofunctional perspective of seizure onset and evolution when taking the clinical history in the clinic. Over time, this is likely to provide a gestalt for surgical candidacy from the moment refractoriness is declared.

2.2.3 | Detailed seizure semiology

Management of epileptic seizures requires different degrees of semiological detail. Adequate prescription of antiepileptic drugs requires very limited semiological detail, as all seizures arising from a focal epileptogenic zone generally respond (or not) to the same antiepileptic drugs regardless of focus location. Seizures produced by a generalized epileptogenic zone differ in their response to antiepileptic medications according to some broad semiological features (tonic–clonic seizures vs tonic seizures vs myoclonic seizures vs absence seizures). On the other hand, surgical management requires detailed semiological description to adequately localize the epileptogenic zone.

It is possible to classify seizures initially into broad semiological groups that are then progressively subdivided into smaller subdivisions.[15] In such a system, it is possible for the clinician or investigator to classify seizures with the desired degree of precision depending on specific requirements tailored to the clinical situation. The ILAE seizure classification[2] uses this methodology for broad seizure groups ("motor" and "nonmotor" seizures) and "motor" seizures are then subdivided into six subgroups and nonmotor seizures into four subgroups. At that point, the ILAE classification stops further attempt at classification, arguing that "focal seizures provoke a variety of potential sensations and behaviors too diverse to be incorporated into a classification." In the management of seizures, all semiological data are important and there is no reason why these data should not be included in the classification of epileptic seizures. Rather than discard these details, it is possible to organize a classification that is designed from the outset with different levels of precision that can be used as necessary depending on the context. It is, however, essential to stress that including semiological details in an epilepsy classification is only practical if we adhere to the following guidelines:[2]

1. The classification should initially classify semiological features in a limited number of broad classes. These classes are then divided into subclasses, and these subclasses are again subdivided, and so on. In such a system, any seizure can be classified with the degree of precision the user feels necessary.
2. Semiological seizure features should be grouped into a class or subclass according to the following two criteria:
   a. Classes or subclasses group together semiologically similar features (eg, motor seizures, tonic seizures, somatosensory auras).
   b. At the same time, efforts should be made to group together semiological features that point to a common symptomatogenic zone. For example, seizures with distal automatisms tend to originate from the temporal lobe, whereas seizures with proximal automatisms usually arise from the frontal lobe.

Guidelines to identify a semiological seizure class are not specified anywhere in the report of the ILAE Commission.[2]

2.3 | Complicated and redundant terminology should be avoided

A seizure classification should use simple terminology and as far as possible only introduce new terms if absolutely necessary. The 2017 ILAE classification of seizures[2] uses terminology that is cumbersome and frequently more imprecise than traditional terminology. For example, the expression "aura" is replaced by "focal aware seizure." This change does not bring additional biological value but rather encumbers description. The definition of "focal aware seizure" has the same limitation as the terminology it has replaced ("simple partial seizures"), where objective definition of awareness is often challenging.

It is also important not to use redundant terms in the classification system. Unfortunately, the ILAE classification does not satisfy this requirement. For example, the traditional term "visual aura" is now replaced by "focal aware sensory (visual) seizure." In this case, "focal" is automatically redundant, because with very rare exceptions auras indicate that the patient has a focal epilepsy. The term "aware" is likewise redundant, because the patient could not possibly describe such an aura unless he or she were aware enough to have noticed and remembered it. Finally, "sensory" is also superfluous.

3 | CRITIQUE OF THE EPILEPSY CLASSIFICATION

1. We propose that the most efficient and meaningful way to classify epilepsy is to use a multiaxis (or multidimensional) system that describes each epilepsy according to a set of domains that are complementary and independent from one another.[15] One way to classify epilepsies would be a four-dimensional system that includes vital information using the following axes:
   a. Seizure type (defined exclusively by seizure semiology)
   b. Location of the epileptogenic zone (defined by all available information, particularly MRI and EEG)
   c. Etiology
   d. Comorbidities

This approach minimizes overlap (redundancy) by including only independent or mostly independent dimensions. The only potential overlap is that semiology may correlate with the location of the epileptogenic zone, but as described above, there are important advantages to reporting semiological data separately.

The ILAE "four diagnostic levels"[1] (seizure type, epilepsy type, epilepsy syndrome, and epilepsy with specific etiology) are redundant, overlapping, and confusing:

- Seizure type specifies both the seizure onset zone and seizure semiology. Neither of these two "dimensions" are properly defined, nor are they clearly differentiated from one another.
- Epilepsy type is redundant, because specifying the seizure type automatically defines the seizure category; patients with focal seizures have focal epilepsies, patients with generalized seizures have generalized epilepsies, patients with focal and generalized seizures have focal and generalized epilepsies, and patients with seizures of unknown origin have epilepsies of unknown origin. If the seizure type is known, the epilepsy type becomes a tautology.
- Epilepsy syndromes consist of specific constellations of:
  - Similar semiologies
  - Similar EEG abnormalities
  - Similar comorbidities
  - Similar type of etiologies

Syndromes were defined by astute epileptologists who realized that the correct identification of an epilepsy syndrome was often helpful to determine prognosis and treatment, but all syndromes are by definition empirical and artificial. Modern diagnostic techniques including MRI and genetic testing now allow precise diagnosis of epilepsy causes, so identification of syndromes is less important than it once was,[6] although several still impact therapy decisions (such as West syndrome, benign rolandic epilepsy, Dravet syndrome, juvenile myoclonic epilepsy) or have relevance to genetic research (such as Dravet syndrome).

As diagnostic technology and knowledge about epilepsy improve, it is likely that more syndromes will become obsolete in the near future. The emphasis of a classification scheme should not be to preserve a set of increasingly archaic conventions, but rather to define as precisely and objectively as possible the characteristics of each individual case of epilepsy to facilitate discovery of new etiologies.

- Regarding epilepsy with a specific etiology, the future of epilepsy treatment is appropriately anticipated in this last diagnostic category specified by the ILAE Commission.[1] We certainly agree with the elegant discussion of Scheffer et al[1] and the emphasis she places on etiology.

As we can see from the discussion above, the deficiencies of the diagnostic system proposed by the ILAE Commission are all resolved by adopting a multidimensional system that includes semiology, epileptogenic zone, etiology, and comorbidities.[15]

2. Etiology is increasingly becoming an essential component of epilepsy diagnosis. The ILAE Commission[1] stresses the importance of an etiological diagnosis, proposing five major etiological groups, with the understanding that

certain etiologies (such as tuberous sclerosis) may affect more than one of those five classes. However, from a practical point of view, it is usually much less valuable to know which of these five broad etiological groups are involved than it is to identify as precisely as possible what the etiological abnormality actually is. For example, in patients with *SCN1A* or *KCNT1* mutations, it is not helpful simply to know that these patients have a "genetic" etiology. Therefore, the ILAE Commission should not only discuss the five major etiological causes of the epilepsies but also indicate how to include a detailed etiological classification that, by definition, will evolve with time.

3. Seizures arising from different parts of the cortex differ considerably, and that is the reason most epilepsy classifications have included localization as an essential dimension or factor. In the current ILAE classification,[1] localization of the epileptogenic zone is extremely limited, including only "focal" versus "generalized" versus "focal and generalized" versus "unknown." Such a classification is certainly inadequate when considering surgical treatment, which requires exact localization of the epileptogenic zone. Even in nonsurgical contexts, however, a detailed semiological classification does have therapeutic relevance. When assessing the efficacy of antiepileptic drug regimens, it is expected that medication will control seizure components along an "axis" opposite to that of seizure evolution. Consider the following seizure example:

(1) Left visual aura → (2) left hand clonic → (3) left versive → (4) bilateral clonic seizure

Although the ideal goal is to achieve control of all components of a seizure in a given patient, in a large number of patients this is not feasible, and hence a realistic expectation should be that the drug regimen will, at least, suppress occurrence of the more severe components of the usual seizures. In the example above, a given antiepileptic drug regimen may be able to largely minimize bilateral clonic seizures, despite not controlling to the same degree the left visual aura and the left hand clonic movements. Careful consideration of the differential efficacy of antiepileptic medication in distinct seizure components may determine the ultimate functional impact of the recurrent seizures in a given patient. This, in turn, may help decide whether surgical remediation should be considered.

4. Finally, the exact localization of the epileptogenic zone may help in the diagnosis of the pathology causing the seizures and its treatment. For example, if limbic encephalitis is suspected, the occurrence of seizures from mesial temporal origin would strongly support the diagnosis. Localization is also important to determine whether a lesion visible on MRI is epileptogenic and can be essential for correct interpretation of subtle MRI abnormalities.

4 | THE QUEST FOR A UNIFIED CLASSIFICATION

The ILAE has established different commissions for the classification of epileptic seizures and epilepsies in adults[1–3] newborns, and for patients in status epilepticus.[4] Differences between these classifications do not refer only to details but include the main framework of the classifications. For example, as mentioned above, the classification of the epilepsies of Scheffer et al[1] calls for classification of progressively more detailed diagnostic categories (seizure type, epilepsy category, epilepsy syndrome, and epilepsy with specific etiology). On the other hand, the status epilepticus classification of Trinka et al[4] is a multidimensional classification including four dimensions: semiology, etiology, EEG, and age.[15]

We feel that to have completely different classifications using different frameworks to classify seizures occurring at different age groups and/or for status epilepticus adds unnecessary complexity to the classification system. This confusion can be avoided by using the same framework for adults and children and also for patients with status epilepticus.

The epilepsy itself (as defined by the epileptogenic zone, etiology, and comorbidities) will not vary just because the patient had status epilepticus or is a newborn rather than an adult, but there will likely be a difference in the seizure type. In a pure semiological classification, this can easily be resolved by replacing the expression "aura" by "aura status" and the expression "seizure" by "status." The semiological status is then added to the semiological dimension.

There are some seizure types that only infrequently occur in infants (such as "generalized" tonic–clonic seizures, automotor seizures, and auras), and other seizure types are seen mainly in infants (such as epileptic spasms).[16] The easiest way to resolve this complexity is to include within a unified framework all possible seizures (statuses) and have the user choose the seizure type that applies in any given situation. This is a much more straightforward solution than including multiple independent classification schemes.

DISCLOSURE

None of the authors has any conflict of interest to disclose. 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.

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APPENDIX 1

SPECIFIC COMMENTS ON THE ILAE SEIZURE CLASSIFICATION

The ILAE seizure classification[3] includes the following: 3 tables with a list of "common descriptors" (Table 1), a "glossary of terms used in this paper" (Table 2), and a table mapping old to new seizure-classifying terms (Table 3). "Common descriptors" are terms that the ILAE Classification Committee feels are not "seizure types" but are terms that the Committee encourages to be used in the description of seizures. The glossary, on the other hand, includes many of the terms listed as "common descriptors" also as "old terms for seizures," which presumably should now be replaced by the "new terms for seizures" in seizure descriptions. It is important to point out here that many of the terms used as "common descriptors" are not included in the glossary.

We feel it would be better to make a list of terms that the Committee encourages to use and include a corresponding glossary of all those terms. On the other hand, there should be a list of "old terminology," which the Committee feels are terms that should not be used anymore including also a mapping of old to new terminology. To include the old terminology in the glossary, mixing terms that should be used with those that are discouraged, is confusing.

Specific comments about Table 1 ("Common Descriptors")[3]

1. In addition to the six semiological seizure descriptors, there is also a laterality group (left vs right vs bilateral) in the "common descriptors." In the seizure type classification focal versus generalized, bilateral refers to the seizure onset zone or its spread (focal to bilateral). Therefore, the Committee must specify whether in the "common descriptors" the expression left versus right versus bilateral modifies the corresponding semiological descriptors or actually refers to the seizure onset zone.

Example A: A patient with left occipital epilepsy (left occipital lesion on MRI and left occipital spikes on the EEG) has a visual aura of flashing lights covering both visual fields. Should this be described as a "focal aware sensory (left visual) seizure" ("left" because the seizure onset is on the left) or as a "focal aware sensory (bilateral visual) seizure" (because the visual symptoms are bilateral)? In the semiological

classification,[16] semiology is an independent parameter of the epileptogenic zone. Therefore, we would simply "classify" the seizure as a "bilateral visual aura," because semiologically the patient has bilateral visual hallucinations, and in the epileptic zone dimension, we would classify the patient as having left occipital epilepsy.[15]

Example B: A patient with left occipital epilepsy (left occipital lesion on MRI and left occipital spikes on the EEG) has a visual aura of flashing lights in the right visual field. Should this be described as a "focal aware sensory (left visual) seizure" ("left" because the seizure onset is on the left hemisphere) or as a "focal aware sensory (right) seizure" (because the aura is in the right visual field)? In the semiological classification, we would simply classify this as a "right visual aura," because semiologically the patient has right visual hallucinations, and in the epileptic zone dimension, we would classify the patient as having left occipital epilepsy.

Example C: The patient described in Example B has a right visual aura evolving into a shaking of all extremities for 1 minute. This would be described as a focal to bilateral tonic–clonic seizure. The lateralizing and localizing value of the right visual aura would be lost. In the semiological seizure classification, this would be classified as "right visual aura → generalized tonic–clonic seizure."

2. In Table 1, hallucinations are included as a subgroup of cognitive seizures. All sensory seizures, however, are actually hallucinations according to the definition of hallucinations provided in the glossary.
3. Under autonomic, they list "respiratory changes" and also "hypo/hyperventilation." Actually, hyper/hypoventilation are relatively uncommon manifestations of seizures or are just secondary signs to the main clinical symptom (eg, irregular, hypoventilation during a generalized tonic–clonic seizure), whereas apnea (not listed specifically as a common descriptor) is the most frequent respiratory change seen as the dominant and not infrequently the only sign at the beginning of mesial temporal lobe epileptic seizures.

Specific comments about Table 2 ("Glossary of Terms")

Generalized seizure was defined as a seizure originating at some point within, and rapidly engaging bilateral distributed networks. Actually, focal seizures originating from midline structures (mesial frontal lobe, mesial paracentral structures, mesial occipital lobes) tend to spread to the contralateral hemisphere within 5-10 milliseconds and spread widely in the contralateral hemisphere.

Specific comments about Table 3 ("Mapping of Old to New Seizure Classifying Terms")[2,3]

1. Many of the old terms are mapped to new terms that include a "common descriptor" (dacrystic, gelastic, gustatory, Jacksonian, and uncinate), which according to the ILAE Committee is not "intrinsic to the classification." For example, in Table 3, these "common descriptors" that are "not intrinsic to the classification" are added in parentheses at the end of the seizure type (eg, "Focal [aware or impaired awareness] sensory [gustatory])." Mapping semiologically very different seizures to a common seizure type is problematic. Besides, it is difficult to understand how a patient with impaired awareness can have gustatory seizures. Notice that in the semiological classification,[16] the seizure would simply be classified as a "gustatory aura."[15]
2. In the "Summary of Rules for Classifying Seizures,"[3] the ILAE Committee encourages adding "common descriptors" as free text (eg, "*Focal emotional seizure* with tonic right arm activity and hyperventilation"). It is easy to agree that any classification of seizures should also be complemented by a detailed description of the actual seizure semiology. It certainly is also important to have a glossary for such a description to make sure we use clearly defined terms in the description. However, such a description is not part of a classification.

To comment specifically on this example, in Table 1,[3] *emotional seizures* are a subgroup of nonmotor seizures. The free text descriptor, however, includes tonic manifestations corresponding to motor seizures.

3. Many of the "old terms" are mapped to "new terms" that are of relatively little value because of their vagueness. Examples:

Old term	New term and criticism
Frontal, parietal, occipital seizure →	Focal seizure. Lumping together all focal seizures arising from different lobes under the term "focal seizures" is a gross simplification, neglecting important semiological information.
Fencer's posture →	Focal motor tonic seizure. Certainly, most focal tonic seizures are not "fencer's posture." The same is true in the next examples below.
Figure-of-4 →	"Focal motor tonic seizure."
Jacksonian seizure →	Focal aware motor.

4. Many of the "old terms" are mapped to new terms that are excessively restricted. Examples:

Old term	New term and criticism
Jacksonian seizure →	"Focal aware motor seizure (Jacksonian)." Jacksonian seizures, however, can occur in patients who are aware or are unaware.
Sylvian seizure →	Focal motor seizure. Sylvian seizures frequently are nonmotor.
Dacrystic or gelastic →	Focal emotional (dacrystic or gelastic) seizure. Frequently these seizures occur without mirth, ie, there is no emotional component.

5. Some of the seizures are mapped inaccurately.

Old term	New term and criticism
Astatic seizures →	[Focal/generalized] atonic seizures: "Astatic" means loss of posture, ie, falling down (a = not, status = standing). Focal loss of postural tone is only one mechanism that leads to a fall. Falls during a seizure only infrequently (less than one-third of falls) are produced by generalized or proximal muscle tone loss; usually, a generalized myoclonic seizure (frequently followed by a generalized atonic seizure) will produce the fall. Therefore, mapping it to [focal/generalized] atonic seizure is not accurate.
Dialeptic seizure →	Focal impaired awareness. In the design of the semiological classification, we strictly avoid any terminology that includes a mixture of seizure semiology and seizure onset (epileptogenic zone) description.[15] This led us to coin the term "dialeptic seizure," which refers to seizures manifested by an alteration of awareness (unresponsiveness to external stimuli) and amnesia for the event independent of whether the epileptogenic zone is focal or generalized.

6. Table 3 includes many "old" seizures that for over a century epileptologists have recognized may occur in patients with focal and generalized epilepsy (clonic seizures, myoclonic seizures, tonic seizure). There is no need to map these seizures from "old" to "new." However, in patients who have these seizures, the seizure origin may be unknown. It is unclear why these seizure types are not listed in Figure 2 of Fisher et al[3] under "Unknown Onset."

7. Absence seizures, since the introduction of the EEG, have also been considered to be typical examples of generalized epilepsy. To label these seizures just "absences" or "generalized absences" is not an innovation, except that the expression "generalized" is redundant.