COMMENTARY

Epilepsia®

From theory to practice: Critical points in the 2017 ILAE classification of epileptic seizures and epilepsies

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Dr Fisher and colleagues have recently provided a commentary on our critique of the 2017 International League Against Epilepsy (ILAE) classification of epileptic seizures and epilepsies.[1] The commentary was disappointing in that it did not depart from their original views and suggests a lack of engagement to consider an alternative framework that might improve the ILAE classifications of epileptic seizures and the epilepsies.[2,3]

Epileptologists actively dealing with people with epilepsy must provide feedback and express their views on any official ILAE position, as the latter would only be valid if it succeeds in striking a balance between the perspectives of any ILAE commission and that of the practitioners. It is evident that thoughtful critiques of ILAE position papers can only be forwarded once a text is published and their directions put into use, which is what we did in our critique,[4] in an effort to achieve the best synthesis of accumulated knowledge that could result in a practical, useful, and forward-looking classification of epileptic seizures and the epilepsies.

1 | GENESIS OF CLASSIFICATIONS

In topics such as a classification of seizures, theoretical objective approaches need to be tempered with frontline experience dealing with patients, families, and referring doctors. A balanced view in a classification of epileptic seizures and epilepsies should seek to incorporate (1) nomenclature and concepts that are simple and easy to understand and communicate, (2) advances in technology and genetics, (3) new concepts, and especially (4) accumulated experience from

managing individuals with epilepsy. A successful classification is not imposed by diktat, but must evolve iteratively, taking into account feedback from the community.

2 | TERMINOLOGY AND THE WORLDWIDE USEFULNESS OF A CLASSIFICATION

If an ILAE classification is to target the international community, it must carefully choose its wording, both to facilitate communication and to encourage proper diagnosis. Terminology that is not easily understood by those for whom English is not their first language should be avoided. Terms such as "focal aware," "focal impaired awareness," and "nonmotor cognitive," for example, are complicated and nonintuitive. As a consequence, many general neurologists, nurses, and others working with persons with epilepsy will likely not incorporate this terminology and end up simply classifying a seizure as either focal or generalized. The result is that by shortcutting simple semiological features, a practitioner may misclassify focal seizures as generalized attacks, because features of a seizure with generalized movements or disconnection from the environment have the greatest importance for patients and relatives when they describe seizures to a doctor or a nurse, and this may lead to suboptimal treatment.

We suggest that ILAE should reflect upon this unfortunate consequence when choosing the framework of a seizure classification and its terminology. This situation could be circumvented by encouraging professionals dealing with people with epilepsy to add simple semiological features, as suggested by the four-dimension epilepsy classification (4D-EC) scheme.[5] In other words, if nurses, general practitioners, and neurologists are encouraged to report the order of events, and not focus purely on the most dramatic and obvious aspects of a seizure, diagnosis would be more precise and mistreatment would be less likely. Because listing the semiological sequence of seizures is part of the 4D-EC, health providers would be encouraged to identify the semiological evolution of epileptic seizures, which is not difficult; all that is needed is to ask patients or witnesses.

Terminology needs to be clear and to avoid confusion. Let us analyze this paragraph, from the ILAE seizure classification scheme 2: "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 (nonmotor onset) cognitive seizure' (progressing to clonic left arm jerks). The terms in parentheses are optional. The seizure type in this example is determined by the cognitive nonmotor onset and presence of altered awareness during any point of the seizure." This is an example of wording that may confuse more than help. This seizure consists of dysphasia, disconnection, and focal clonic movements of one arm. In the 4D-EC, it would be classified as follows: ictal aphasic→disconnection (loss of awareness)→left arm clonic seizure. This classification perspective gives the practitioner much more information than a "focal impaired awareness cognitive seizure" (which could mean any ictal cognitive abnormality and thus originate from very different cortical regions). Furthermore, because the patient evolved to disconnection from the environment, the 2017 ILAE nomenclature ignores that the patient was aware of a language difficulty at seizure onset. These problems are inevitable if one uses a single word or expression such as "focal impaired awareness (nonmotor onset) cognitive seizure" to designate different features. The choice of the ILAE classification commission to lump together focal seizures does not encourage a true understanding of what is going on and where in the brain. We think this reduces understanding and impoverishes doctor-patient interactions in epileptology, an area where patients are often perplexed with what suddenly happens to them and seek explanations. Furthermore, classifying the example above as "focal impaired awareness cognitive seizure" simply "crystallizes" something that is dynamic. This confuses even the treatment outcome, as an antiepileptic drug (AED) regimen may control the more disabling motor aspects of the seizure, which are not even mentioned in its classification. For instance, seizures could change after AED from aphasic→disconnection (loss of awareness)→left arm clonic seizure to aphasic seizure only, which changes the putative morbidity and practical impact of the seizures. With the ILAE epilepsy classification, seizures would appear unchanged or would need to be reclassified as "focal aware cognitive seizure." This conundrum can be avoided by using semiology evolution as proposed in the 4D-EC,[5,6] which is more helpful in understanding the dynamic nature of seizures, explaining the treatment goals, and assessing the results of treatment.

3 | FOCAL VERSUS GENERALIZED SEIZURES, LOCATION OF EPILEPTOGENIC ZONE, AND HOW TO ENCOURAGE REASONING

A pure semiological seizure classification, assessing what happens at seizure onset and during evolution, provides neurologists with an anatomofunctional perspective when taking the clinical history. This leads to one dimension of localization of the epileptogenic zone, which could raise potential surgical candidacy in the mind of a neurologist. This would naturally lead to a second dimension—or axis—with investigation with magnetic resonance imaging and electroencephalography (EEG). Furthermore, a dynamic perspective of seizure evolution is clearly important when trying to differentiate epileptic

seizures from other events such as syncope, psychogenic non-epileptic seizures, and parasomnia. We contend that including this critical, dynamic feature of seizures could help decrease misdiagnosis. It has been argued that such an anatomofunctional approach is too complicated. That may be true for nonneurologists and perhaps less-specialized nurses. To accommodate this, the 4D-EC can also classify seizures in very broad categories. Furthermore, neurologists and epileptologists should be trained to analyze seizures from an anatomofunctional perspective as outlined above.

Previous classifications, which align with the 2017 ILAE proposal, led to a way of thinking in which information from multiple dimensions or axes (such as semiology and EEG) is immediately integrated to provide a classification of the seizure type and direct treatment. There is nothing wrong with this when an EEG is available, abnormal, and correctly interpreted. This approach, however, may be confusing, because of variable availability of tests and variable quality of interpretation. Thus, it may be better to have neurologists think of the possible seizure scenarios involving "disconnection from the environment" (or loss of awareness) and do their best to use clinical features to differentiate a focal from a generalized seizure. That initial impression may then be confirmed or altered based upon EEG findings, which is common in practice and is a more straightforward proposition than, for instance, the ILAE-proposed terminology, "nonmotor behavioral arrest of unknown onset."[2]

4 | TIMING AND ADEQUACY FOR A CHANGE IN PERSPECTIVE

There have been major changes in epileptology over the past 2 decades. The first is the realization that epilepsy surgery is a highly underutilized method to treat a significant percentage of patients with refractory focal seizures.[7,8] This situation could be improved by a classification that encourages neurologists to consider localization of seizure onset, thus prompting a "gestalt" of surgical candidacy. In contrast, a classification system that leads neurologists away from the ingrained anatomofunctional thinking applied in their general clinical practice through the use of general terms such as "sensory," "motor," and "cognitive" may continue to lead to the underutilization of epilepsy surgery. Second, imaging, genetics, and neuroimmunology will continue to advance apace, and clarify the etiology of epilepsy of an increasing proportion of patients, and these data should be included in any classification with a multiaxial approach, a point on which both classification systems concur.[2,5]

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