

Commentary:

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 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.

The 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 (i) nomenclature and concepts that are simple, and easy to understand and communicate, (ii) advances in technology and genetics, (iii) new concepts and, especially, (iv) accumulated experience from managing individuals with epilepsy. A successful classification is not imposed by dictat, but must evolve iteratively, taking into account the feedback from the community.

Terminology and the world-wide 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 encourage proper diagnosis.

Terminology which 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 ‘non-motor cognitive’, for example, are complicated and non-intuitive. In 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 consequence 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 describing seizures to a doctor or a nurse, and which 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 4-dimension epilepsy classification (4D-EC) scheme⁵]. 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. Since 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 avoid confusion. Let us analyze this paragraph, from the ILAE seizure classification scheme²: “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 non-motor 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 ’(that 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 the fact that the patient was aware of a language difficulty at seizure onset. These problems are inevitable if one uses a single word or expression (ie, “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. Further, classifying the example above as ‘focal impaired awareness cognitive seizure ’simply ‘crystallizes ’something that is dynamic. In fact, this confuses even the treatment outcome, as an 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-EC, 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 to understand the dynamic nature of seizures, explain the treatment goals, and assess the result of treatment.

Focal versus generalized seizures, the location of the 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 anatomo-functional perspective when taking the clinical history. This leads to one dimension of localization of the epileptogenic zone, that could raise potential surgical candidacy in the mind of neurologists. This would naturally lead to a second dimension – or axis – with investigation with MRI and 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. Indeed, we contend that including this critical, dynamic feature of seizures could help decrease misdiagnosis. It has been argued that such anatomo-functional approach is too complicated. That may be true for non-neurologists and perhaps less specialized nurses. To accommodate this, the 4D-EC can also classify seizures in very broad categories. Further, neurologists and epileptologists should be trained to analyze seizures from an anatomo-functional 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) are 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 'non-motor behavioral arrest of unknown onset'².

Timing and adequacy for a change in perspective

There have been major changes in epileptology over the last two 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 anatomic-functional 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 multi-axial approach, a point on which both classification systems concur^{2,5}.

Disclosure and ethics

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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