

A semiological classification of status epilepticus

To the Editor: We read with great interest the thoughtful review of Prof Simon Shorvon of our paper titled «A semiological classification of status epilepticus» (*Epileptic Disord* 2005; 7: 1-3). Prof Shorvon is one of the world experts in status epilepticus, and this is certainly reflected in this insightful review.

We agree wholeheartedly with Prof Shorvon that classifications can be divided into first-order and second-order systems, each one satisfying different needs. It is important to stress however, that second-order classification systems are not only important practical tools but may also facilitate research by providing an objective description of a biological variable, in this case the semiology of epileptic status. For example, such an objective classification may be essential for the discovery of modifier genes that affect the semiological expression of epilepsy. Moreover, the scientific development of a first-order classification almost invariably depends on a carefully designed, objective, second order classification system. In other words, the second-order classification system permits the scientific revelation of relationships between categories that is essential for the establishment of a first order classification system. Darwin hypothesised evolution by analyzing the relationship between different second-order classification categories of animals and plants with other second-order classification categories of environmental factors. In general, first-order classification systems require, at an initial stage as a building block, an objective, second-order classification system.

We strongly agree with Prof Shorvon on his assessment that essentially all available epilepsy classification systems represent second-order classification systems. Unfortunately, even modern epileptology has not sufficient insight into epilepsy to permit a reliable first-order classification. Therefore, it is essential that at this point we develop an objective, clearly defined second-order classification system that can be used effectively in future research endeavors for a better understanding of epilepsy, and hopefully, the eventual discovery of a first-order classification system. On the other hand, assuming that we have such an understanding, developing a so-called «first-order classification system», based on pure assumptions may be very misleading. For example, the current *Taskforce on Classification and Terminology* has submitted a proposal of a «first-order» classification of epileptic seizures, presumably based on the pathophysiology of

the seizures. However, there is absolutely no objective evidence which documents that the different semiological seizure types described in this classification have, indeed, a different, underlying pathophysiology.

We also agree with the criteria that Prof Shorvon outlined as the essential points in deciding the «utility» of a second-order classification system. In essence, Prof Shorvon concludes that the semiological classification of status epilepticus (SCSE) scores high in point 1, but relatively lower in points 2 to 4. Let us discuss these points.

With respect to point number one, we certainly made every effort to define, as clearly as possible, all of the semiological features that we observed in status epilepticus. Unfortunately, classification of clinical signs and symptoms is always difficult, and even the most classical signs or symptoms (as for example the Babinski sign), are always somewhat ambiguous. Prof Shorvon is correct in that some of the terms used in the classification are new. However, as explained elsewhere, these new terms were actually chosen to avoid ambiguity and confusion. For example, *diabetic status* is a new concept, defined as an alteration of consciousness produced by an epileptic condition. It is not equivalent to absence status (which by definition refers to patients who have a generalized epilepsy), or *complex partial status* (which applies to patients with partial epilepsy). There is no term in the current literature that can be applied to those patients in whom it is uncertain whether or not they have focal or generalized epilepsy.

Prof Shorvon is also correct when he indicates that sometimes it is unclear if the observed symptomatology is due to the epileptic condition or is just the expression of the underlying neurological condition. It is important, however, to stress here that in all the cases discussed in our manuscript, we made sure that the patient had an associated epileptiform EEG and that there was a clear temporal correlation between the presence of epileptiform discharges and the symptomatology we called «epileptic». In other words, we used the EEG to determine if the observed condition was epileptic or not, even if we did *not use the EEG to classify* the seizure semiology. This approach minimized the error that Prof Shorvon points out in his discussion, (for example, this approach avoids the mislabeling of non-epileptic subcortical jerking as *epilepsia partialis continua*). On the other hand, we agree with Prof Shorvon that in some comatose patients, who actually show epileptiform discharges, it may sometimes

be difficult to define if any given symptoms or signs are related to the epileptiform discharges or are just an expression of the underlying neurological deficit (coma and PLEDs are the typical example).

Points 2 and 3 highlighted by Prof Shorvon are actually similar. In point 2 he requires that the criteria of classification are of practical utility and in point 3 that the information conveyed by the classification is clinically useful. Prof Shorvon is entirely correct when he points out that often, a detailed description of the semiology of the status epilepticus or of the evolution of these symptoms is not clinically relevant. This is, however, not a weakness but actually the strength of the proposed classification system. As for most first-order classification systems, we have divided the semiological features into broad categories that can be subdivided into progressively smaller and smaller subgroups according to the need of the user. Besides, the user of the classification system can use only one or more axes. For example, depending on the detail you feel is clinically relevant, you can talk about motor status, simple motor status, clonic status, hand clonic status or, in with maximum detail, right hand clonic status. The same is true regarding evolution. In the manuscript published in *Epileptic Disorders*, we used *the maximum possible detail* because we wanted to illustrate the ability of the system to define semiological detail for those situations in which it could be relevant. It is important to stress however, that in no way do we imply that this should be common clinical practice. On the other hand, we disagree with Prof Shorvon's assessment that a detailed description of the semiology of the status is only important in those cases in which surgical treatment is being considered. For example, in many patients with a neurological disease, precise definition of the status epilepticus may be helpful in localizing the lesion, (for example, an elderly patient who presents with sudden onset, left hand, clonic status epilepticus). Let us also point out here, that the same system of different categories with different degrees of precision is actually defined by the first-order classification of biological systems. However, in everyday practice, we almost never use the detailed classification available, but only the subgroup which fulfills our needs (kingdom, phyla, class, order, family, genus or species).

In point 3, Prof Shorvon indicates that the SCSE has only limited clinical usefulness. He indicates, correctly, that other factors such as underlying cause, co-morbidity, etc., may be more relevant. At this point we would like to stress that the SCSE is only a classification of the symptoms and signs of the status epilepticus and not a classification of the status epilepticus itself. It is obvious that a classification of the semiology of status cannot provide other information such as etiology, associated medical conditions, etc., which in many cases may be more relevant from a management rather than etiological point of view. This is exactly the reason why, in the manuscript published in *Epileptic Disorders*, we included an epilepsy classifica-

tion. This classification consists of different axes, one is the SCSE, but the other axes define the etiology, associated medical conditions, and other pertinent variables.

Finally, in point number 4, Prof Shorvon points out that the SCSE is too complex for the majority of the targeted audience. As we mentioned above, this is certainly the case for the detailed classification presented in the manuscript we published in *Epileptic Disorders*. However, as outlined before, the actual intention of the classification is to provide different levels of complexity, according to the needs of the practitioner. It is interesting to observe that the first order biological classification is also extremely complex. However, in one way or another we all use the subgroups that give us the precision we may need in any given situation.

Prof Shorvon also indicates that he does not necessarily agree with the definition we used for status epilepticus. The 10 minutes used in this manuscript is only an arbitrary limit. The same discussion would certainly apply if we set the limit to the standard 30 minutes.

Summarizing, the main objective of our paper was to describe a semiological classification of status epilepticus (SCSE). This classification defines with precision the main classes of symptoms and signs observed during status epilepticus. In addition, by providing different degrees of semiological precision and complexity, it is a highly flexible system that can be used by different groups of practitioners depending on their needs and their level of familiarity with epilepsy. In addition, its application is greatly simplified because it contains the same categories and follows the same general principles used for the classification of epileptic seizures. The SCSE, as for the semiological classification of epileptic seizures, only provides information about the semiology of seizures (status epilepticus), which is only one dimension of epilepsy classification and usually only a small part of the relevant information necessary for managing patients in status epilepticus. Practitioners who are looking for a comprehensive classification will primarily be concerned with the classification of the epilepsies which includes other essential factors such as etiology and related medical conditions. □

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