Invited Editorial Comment

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The classification of status epilepticus

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The classification of disease has always been, and presumably will always remain, a controversial subject. It is therefore with some trepidation that I make this contribution to the pages of *Epileptic Disorders*. It is an almost universal truth that no single classification scheme in neurology will satisfy all persons, and indeed, most satisfy few, the field of epilepsy provides no exception. Clinicians, scientists, philosophers, bureaucrats and even governmental bodies have got in on the act, and as time has passed, more and more schemes have been presented. George Bernard Shaw thought crude classifications were "the curse of organized life", and it is hard not to sympathize with him or indeed with Disraeli, who rather more bluntly wrote "I hate definitions!". However, classification schemes are necessary in science and medicine. In this editorial, as requested, I shall comment critically on the new scheme of classification of *status epilepticus* proposed by Dr Rona and colleagues (Rona *et al.* pp. 5-12).

It seems appropriate to start with a few broad points. In science generally, classification schemes can be divided (*sic*) into what might be termed two distinct orders:

– **First-order classification schemes** are those which act as a structure on which to organise knowledge for its advancement and for research; and which, in turn are logically constructed according to scientific theory or hypothesis. Such schemes are explanatory, "true" systems of classification. The complexities and difficulties of constructing meaningful classificatory schemes were first explored in the 19th century, as part of the contemporary fashion for biological taxonomy and the rise of Darwinism: the concepts of classification still remain furthest advanced in biology and genetics. The division of plants and animals into kingdoms, phylum, class, order, family, genus and species is a good example of a first-order classification, stimulating as it has done major biological advance, and underpinning much scientific study. Such classification schemes are not arbitrary, but based on a meaningful scientific structure and with well-defined categories. The value of first-order schemes is in the extent to which they facilitate advance in human knowledge;

– **Second-order classification schemes** are altogether different. These schemes are not based on any synoptical, scientific hypothesis (evolution, phylogeny, speciation, underlying physiology, biochemical mechanism, etc), but are in essence, simply definitions – shorthand terms which simply categorise and describe clinical observations. Such schemes are less high-flown, and are needed for practical, everyday purposes, typically the communication of information (e.g. from doctor to doctor, doctor to patient, patient to employer etc). The value of second-order schemes is thus entirely in their practical utility.

This dichotomy has been long recognised in the field of epilepsy. In 1873, Hughlings Jackson (Jackson 1931, Wolf 2003) contrasted the classification of flowers by a botanist (first-order) and a gardener (second-order). He recognised that the second-order

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classifications schemes can be judged on their clinical utility, and that first-order schemes, although helpful to advance science, may be "absurd in asylums or the wards of a hospital".

A major landmark in epilepsy classification was the creation of an "official" classification of seizure type – the ILAE *Classification of Seizure Type* – published in 1970 and revised in 1981 (Gastaut 1970, Commission on Classification and Terminology of the International League Against Epilepsy 1981). The scheme has the outstanding virtues of being both the product of epilepsy's official bureaucracy, and also of being widely accepted. The success of this first exercise emboldened the ILAE to produce a second scheme, a *Classification of the Epilepsies and Epilepsy Syndromes* (Commission on Classification and Terminology of the International League Against Epilepsy 1989). Many other epilepsy classification schemes (mostly variations on the theme) have also been proposed, before and since (a new scheme almost every time a new book is written or a new researcher addresses the field). The current ILAE *Taskforce on Classification and Terminology* is itself wrestling with the task of devising a new scheme (Engel 2001), and recognising the problems, avoids the word classification altogether, referring to a "diagnostic scheme".

The history of the classification of status epilepticus has closely followed that of the classification of seizure type. The term (*état de mal épileptique*) first appeared in 1824, (although the condition itself is as old as time) and was originally used to signify what is now known as tonic-clonic status epilepticus. It was not until the early 20th century that other non-convulsive and myoclonic forms were included within the *status epilepticus* rubric [see for instance Clark and Prout (1903)]. In 1962, the Xth Marseilles Colloquium was the first major meeting to be devoted entirely to the subject of status epilepticus, and at this meeting the first modern definition and classification of status epilepticus were proposed. The classification closely mirrored the *Classification of Epileptic Seizures* newly devised by the same group. Gastaut, its chief instigator, yoked the two inextricably when he suggested that there was a status equivalent for every seizure type (Gastaut 1967). The classificatory scheme of status epilepticus was eventually published in 1970 as an appendix to the *Classification of Epileptic Seizures* (Gastaut 1970) and again as an appendix in the revision of the seizure-type classification published in 1981 (Commission on Classification and Terminology of the International League Against Epilepsy, 1981). In these schemes, seizure semiology and EEG appearance were used as the sole criteria for classification. A more synoptical scheme was proposed in 1994, which categorised status by age and epilepsy syndrome as well as by seizure type (Shorvon 1994), and more recently, a similar classification scheme of non-convulsive status epilepticus (Shorvon 2005). However, it remains true that no universally adopted scheme of classification is in use, and this hampers both clinical and clinical research work in epilepsy.

The dichotomous nature of classification schemes is well known, and the arguments well rehearsed. What should be immediately recognised is that all common current clinical epilepsy classifications are essentially second-order in nature – and thus their value lies almost entirely in their practical utility. It is therefore worth defining what aspects provide "utility":

– *There should be a lack of ambiguity in the definitions of the criteria used*. As second- order schemes are essentially shorthand descriptions, the terms (definitions) must be clear;

– *The criteria chosen should be of practical utility for the target audience*. Classifying by features which are of little clinical importance will undermine the value of the scheme;

– *The classification should convey information which is useful in the situation in which it is being applied*;

– *The classification scheme should be simple enough to be applied consistently and to be understood by its target audience*.

It is quite possible to construct a classification scheme without these features, but to do so reduces utility and, for most purposes, seems rather pointless.

It is against this background, that the new classification scheme for status epilepticus is proposed – the *Semiological Classification of Status Epilepticus* (SCSE) (Rona *et al.* 2005). It is a close adaptation of a similar scheme for classifying seizure type – the *Semiological Seizure Classification* (Lüders *et al.* 1998), just as was Gastaut's classification of status epilepticus a close adaptation of his seizure-type classification.

Like the 1962 classification, the SCSE is based on the observation and analysis of a large group of status episodes (237 episodes at the 1962 Marseilles Colloquium, and 100 EEG-video telemetry cases in the SCSE). It is based on the main clinical manifestations of the seizure and its evolution. Status epilepticus is defined as 10 minutes or more of continuous or intermittent seizure activity, and the classification has 3 "axes": the type of brain function compromised by the seizure (sensory, autonomic, cognitive, motor); the body part involved (somatotropic modifiers – generalised, bilateral asymmetric, axial, left or right, arm/leg/face, eyelid, etc); and the evolution over time (continuous/intermittent, temporal modifiers).

The classification is very much a second-order type, and proudly so. It makes no assumptions about underpinning theoretical structures, and is essentially a descriptive tool and "a standardised way to describe the multitude of semiological features of status epilepticus". The value of this effort therefore comes down to an assessment of its utility, and this can be measured by the four aspects listed above.

First is the question of lack of ambiguity. Here, the SCSE scores highly, although rather confusing new terminology is adopted (dialeptic status, delirious status, aura status etc). The critical issue of ambiguity, though, concerns the criteria used to define a case of status. Here the cases are defined entirely on semiological grounds, and there must be some uncertainty in some of the 100 cases cited (see accompanying CDRom) as to whether the semiology is in fact ictal at all. This problem relates particularly to non-convulsive cases, and it can be difficult to differentiate ictal symptomatology from that which is non-ictal due to encephalopathy or confusion, or to focal neurological symptoms reflecting simply the underlying cerebral pathology. Nonconvulsive status is, in my experience, commonly overdiagnosed in practice around the world, and the failure to address the diagnostic uncertainties is a weakness of the SCSE and a lost opportunity. Similar diagnostic problems are engendered in differentiating epilepsia partialis continua from disorders exhibiting non-epileptic subcortical jerking and in assessing PLEDs in comatose patients.

Second, the clinical utility of some criteria used in classification is open to question. In the SCSE, for instance, is it really important to have details of temporal evolution or to know what body part is involved? The latter of course, is the centre-piece of the semiological seizure classification (Lüders *et al.* 1998), and rightly so in view of the application of the semiological seizure classification in epilepsy surgery, but in status epilepticus, where there is no surgical interest, this seems rather superfluous. Other factors which would have much more utility for diagnostic, prognostic and therapeutic purposes (e.g. aetiology, syndromic type, etc) are not included as diagnostic criteria.

Third, the information conveyed should be clinically useful. The temporal evolution of status epilepticus is often due to factors which have little to do with the inherent processes of the status, but is more related to external events – for instance the effects of treatment, the effects of the level of oxygenation, co-morbidity, the underlying cause. In the SCSE, the emphasis placed on the temporal evolution (with the resulting terminological complexity), without acknowledging the importance of external aspects, seems to lessen the clinical utility.

Fourth, the SCSE, with its large number of categories and complex nomenclature, may prove too complicated to be either applied consistently or to be understood by any but the most experienced practitioner.

Finally, a word about the definition of status epilepticus. In the SCSE, status is defined as any seizure lasting longer than 10 minutes. This is a shorter period than that used in older classifications (usually 30 or 60 minutes), but reflects recent debate on this topic. The arguments in favour of this change are certainly not universally agreed, and to acquire wide adoption, a classification should be based on widely accepted definitions.

The classification and definition of status epilepticus thus remain controversial areas. The SCSE is a serious and original attempt at making sense of the varied clinical forms of status epilepticus. Future schemes might usefully address issues of definition, differential diagnosis, diagnostic criteria and clinical utility. What is also needed in status epilepticus, as in epilepsy generally is a first-order classification, but this is probably many years away. □

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