Classification of Status Epilepticus

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Historical: Introduction

- Status epilepticus (SE), considered the most extreme form of a seizure, included in classification of seizures of ILAE of 1970 and 1981
  - The first ILAE Classification of Seizures, which was developed in 1964 and approved in 1970
- SE was defined as a "seizure that persists for a sufficient length of time or is repeated frequently enough to produce a fixed and enduring condition"
- SE was divided into partial, generalized, or unilateral types, and basically mirrored the seizure classification

Classification of Seizures 1970

- Definition of SE: "... a seizure persists for a sufficient length of time or is repeated frequently enough to produce a fixed and enduring epileptic condition ("status" implies a fixed or enduring state)."
- Classification of SE: “Status may be divided into
  - Partial (e.g., Jacksonian)
  - Generalized (e.g., absence status or tonic–clonic status), or unilateral (e.g., hemi-clonic) types.”

Historical: Introduction

- In the revision of 1981, definition was minimally changed into a “seizure” that “persists for a sufficient length of time or is repeated frequently enough that recovery between attacks does not occur
- The distinction between partial, generalized, and epilepsia partialis continua (EPC) was mentioned
Classification of Seizures Revised 1981

- **Definition:** "... a seizure persists for a sufficient length of time or is repeated frequently enough that recovery between attacks does not occur."
- **Classification:** "Status may be divided into
  - Partial (e.g., Jacksonian)
  - Generalized (e.g., absence status or tonic–clonic status). When very localized motor status occurs, it is referred to as epilepsia partialis continua."

Glossary of descriptive terms 2001

- **Definition of status epilepticus:**
  - No clinical signs of arresting after a duration encompassing the great majority of seizures of that type in most patients
  - Recurrent seizures without interictal resumption of baseline central nervous system function.

Diagnostic scheme for people with epileptic seizures and with epilepsy 2001

Classification: Continuous seizure types:

- **Generalized status epilepticus**
  - Generalized tonic–clonic status epilepticus
  - Clonic status epilepticus
  - Absence status epilepticus
  - Tonic status epilepticus
  - Myoclonic status epilepticus
- **Focal status epilepticus**
  - Epilepsia partialis continua (EPC) of Kojevnikov
  - Aura continua
  - Limbic status epilepticus (psychomotor status)
  - Hemi convulsive status epilepticus with hemiparesis

Definition of epileptic seizures and epilepsy 2005:

- **Definition of seizure:** "An epileptic seizure is a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain. The term transient is used as demarcated in time, with a clear start and finish."
- **Definition of status epilepticus:** Though there is no formal definition in the Report,
  - SE is defined as "a special circumstance with prolonged or recurrent seizures."
Report of the ILAE Classification
Core Group 2006:

- **Definition:** There is no formal definition of SE in the 2006 report, instead SE is described as “the failure of natural homeostatic seizure-suppressing mechanisms responsible for seizure termination . . . .

- Additional factors that need to be considered in determining criteria for classification include:
  - **Different mechanisms** that can prevent seizure termination, i.e,
    - mechanisms that prevent active inhibition, desynchronization of hypersynchronous discharges, and depolarization block
  - **Progressive**
  - **Maturation factors**

Report of the ILAE Classification
Core Group 2006:

- Classification: The proposal of the core group recognizes nine types of SE
  - Epilepsia partialis continua (EPC) of Kojevnikov
  - Supplementary motor area (SMA) status epilepticus
  - Aura continua
  - Dyscognitive focal (psychomotor, complex partial) status epilepticus
  - Tonic–clonic status epilepticus
  - Absence status epilepticus
  - Myoclonic status epilepticus
  - Tonic status epilepticus
  - Subtle status epilepticus

New classification of SE:
Introduction

- Purpose of *diagnostic axes* to provide framework for clinical diagnosis, investigation, and therapeutic approaches for each patient

- In 1970, the axes encompassed
  - Clinical seizure type
  - EEG: ictal and interictal expression
  - Anatomic substrate
  - Etiology
  - Age

- In the 1981 revision, the axes were limited to the *seizure type* and EEG expression (ictal and interictal) (Classification 1981)

- At least half of the patients with SE do not have epilepsy or specific epilepsy syndromes—they have SE due to acute or remote central nervous system or systemic illness. Therefore, the axes used previously in seizure classification need to be modified

New classification of SE:
Introduction

For new classification of SE using four axes:

- **Semiology**
- **Etiology**
- **EEG correlates**
- **Age**
### Axis 1: Semiology

(A) With prominent motor symptoms

A.1 Convulsive SE (CSE, synonym: tonic–clonic SE)
   A.1.a. Generalized convulsive
   A.1.b. Focal onset evolving into bilateral convulsive SE
   A.1.c. Unknown whether focal or generalized
A.2 Myoclonic SE (prominent epileptic myoclonic jerks)
   A.2.a. With coma
   A.2.b. Without coma
A.3 Focal motor
   A.3.a. Repeated focal motor seizures (Jacksonian)
   A.3.b. Epilepsia partialis continua (EPC)
   A.3.c. Adversive status
   A.3.d. Oculoconlonic status
   A.3.e. Ictal paresis (i.e., focal inhibitory SE)
A.4 Tonic status
A.5 Hyperkinetic SE

(B) Without prominent motor symptoms (i.e., nonconvulsive SE, NCSE)

B.1 NCSE with coma (including so-called “subtle” SE)
B.2 NCSE without coma
   B.2.a. Generalized
      B.2.a.a Typical absence status
      B.2.a.b Atypical absence status
      B.2.a.c Myoclonic absence status
   B.2.b. Focal
      B.2.b.a Without impairment of consciousness (aura continua, with autonomic, sensory, visual, olfactory, gustatory, emotional/psychic/experiential, or auditory symptoms)
      B.2.b.b Aphasic status
      B.2.b.c With impaired consciousness
   B.2.c Unknown whether focal or generalized
      B.2.c.a Autonomic SE

### Axis 2: Etiology

- The term “known” or “symptomatic” is used—consistent with the common neurologic terminology—for SE caused by a known disorder, which can be
  - Structural
  - Metabolic
  - Inflammatory
  - Infectious
  - Toxic
  - Genetic

Based on its temporal relationship, the subdivisions acute, remote, and progressive

- Epileptic encephalopathies
- Coma with non evolving epileptiform EEG patterns
- Behavioral disturbance (e.g., psychosis) in patients with epilepsy
- Acute confusional states, (e.g., delirium) with epileptiform EEG patterns
Axis 2: Etiology

**Etiology of status epilepticus**

- Known (i.e., symptomatic)
  - Acute (e.g., stroke, intoxication, malaria, encephalitis, etc.)
  - Remote (e.g., posttraumatic, postencephalitic, poststroke, etc.)
  - Progressive (e.g., brain tumor, Lafora’s disease and other PMEs, dementias)
  - SE in defined electroclinical syndromes
- Unknown (i.e., cryptogenic)

**Axis 2: Etiology**

- The term “idiopathic” or “genetic” is not applicable to the underlying etiology of SE
- In idiopathic or genetic epilepsy syndromes, the cause of status is not same as for disease, but some metabolic, toxic, or intrinsic factors (like sleep deprivation) may trigger SE in these syndromes
- Therefore, the term “idiopathic” or “genetic” is not used
- SE in a patient with juvenile myoclonic epilepsy (“idiopathic” or “genetic”) can be symptomatic, due to inappropriate antiepileptic drug (AED) treatment, abrupt drug withdrawal, or drug intoxication

**Axis 2: Etiology**

- The term “unknown” or “cryptogenic” used in its strict original meaning: unknown cause
- The assumption that it is “presumably” symptomatic or genetic is inappropriate
- SE in its varied forms has a plethora of causes;

**List of Etiologies That May Cause Status Epilepticus**

<table>
<thead>
<tr>
<th>Etiology of status epilepticus</th>
</tr>
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<tbody>
<tr>
<td>Cerebrovasculardiseases (Ischemic stroke)</td>
</tr>
<tr>
<td>CNS infections</td>
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<tr>
<td>Neurodegenerative diseases</td>
</tr>
<tr>
<td>Intracranial tumors (Gliat tumors)</td>
</tr>
<tr>
<td>Cortical dysplasias</td>
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<tr>
<td>Head trauma</td>
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<tr>
<td>Alcohol related</td>
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<tr>
<td>Intoxication</td>
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<tr>
<td>Withdrawal of or low levels of AEDs</td>
</tr>
<tr>
<td>Cerebral hypoxia or anoxia</td>
</tr>
<tr>
<td>Metabolic disturbances</td>
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<tr>
<td>Autoimmune disorders</td>
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<tr>
<td>Mitochondrial diseases</td>
</tr>
<tr>
<td>Chromosomal aberrations and genetic anomalies</td>
</tr>
<tr>
<td>Neurocutaneous syndromes</td>
</tr>
<tr>
<td>Metabolic disorders</td>
</tr>
<tr>
<td>Others</td>
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</tbody>
</table>
Axis 3: Electroencephalographic correlates

- None of the ictal EEG patterns of any type of SE is specific.
- Epileptiform discharges are regarded as the hallmark, but with increasing duration of SE, the EEG changes and rhythmic nonepileptiform patterns may prevail.
- Similar EEG patterns, such as **triphasic waves**, can be recorded in various pathologic conditions, leading to substantial confusion in the literature
- Although EEG is overloaded with movement and muscle artifact in the convulsive forms of SE and thus of limited clinical value
- **It is indispensable diagnosis of NCSE**, as clinical signs are often subtle and non-specific

**Axis 3: Electroencephalographic correlates**

**Terminology to describe EEG patterns in SE:**

- **Location**: generalized (including bilateral synchronous patterns), lateralized, bilateral independent, multifocal
- **Name of the pattern**: Periodic discharges, rhythmic delta activity or spike-and-wave/sharp-and-wave plus subtypes
- **Morphology**: sharpness, number of phases (e.g., triphasic morphology), absolute and relative amplitude, polarity
- **Time-related features**: prevalence, frequency, duration, daily pattern duration and index, onset (sudden vs. gradual), and dynamics (evolving, fluctuating, or static)
- **Modulation**: stimulus-induced vs. spontaneous
- **Effect of intervention** (medication)onEEG.

**Axis 3: Electroencephalographic correlates**

- Advances in electrophysiologic techniques may provide with increased capability to utilize EEG in emergency setting and allow better delineation of highly dynamic changes of EEG patterns in near future
- Currently **no evidence-based EEG criteria for SE**. Based on large descriptive series and consensus panels

**Axis 3: Electroencephalographic correlates**

**NCSE**: bi-PLEDs
Ambiguous, asymmetric triphasic waves in a patient with subarachnoid hemorrhage

Generalized triphasic waves in a 79-year-old man with renal failure. Note the prominent second positive phase and the anterior–posterior lag

Axis 4: Age
- Neonatal (0 to 30 days)
- Infancy (1 month to 2 years)
- Childhood (> 2 to 12 years)
- Adolescence and adulthood (> 12 to 59 years)
- Elderly (≥ 60 years)

Axis 4: Age
- SE in neonates may be subtle and difficult to recognize
- Some forms of SE are seen as an integral part of the electroclinical syndrome; others can occur in patients within
  - Certain electroclinical syndrome
- Trigger factors or precipitating causes are present, such as sleep deprivation, intoxication, or inappropriate medication