Nuova proposta di classificazione degli SE

Paolo Tinuper

IRCCS Institute of Neurological Sciences of Bologna, Italy.
Department of Biomedical and Neuromotor Sciences,

University of Bologna, Italy

STELE NEO-BABILONESE 718-612 AC
XXV/XXVI TAVOLE DI SAKIKKU
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… «se il demonio lo possiede molte volte nella notte, e se nel momento della possessione le sue mani e i suoi piedi sono caldi ed è scuro il viso, se apre e chiude la bocca …. può continuare per qualche tempo…ma morirà»…

1ª referenza all'epilessia e allo SDM

Ci sono delle volte che non appena una crisi finisce, un' altra inizia, una di seguito all' altra in successione, cosicchè si possono contare 40 , 60 crisi senza interruzione : i pazienti lo chiamano " etat de mal". Il pericolo è imminente , molti pazienti muoiono

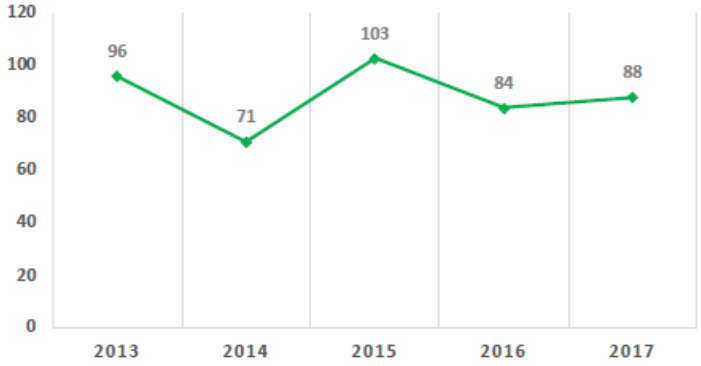
L.F. Calmeil De l' epilepsie, Parigi 1824

Lo Stato Epilettico (SE) è una condizione grave, caratterizzata da un'alta mortalità e morbilità.

Incidenza annuale attorno a 10–20 per 100,000 nell'Europa centrale (Jallon et al., 1999; Knake et al., 2001; Rosenow et al., 2007).

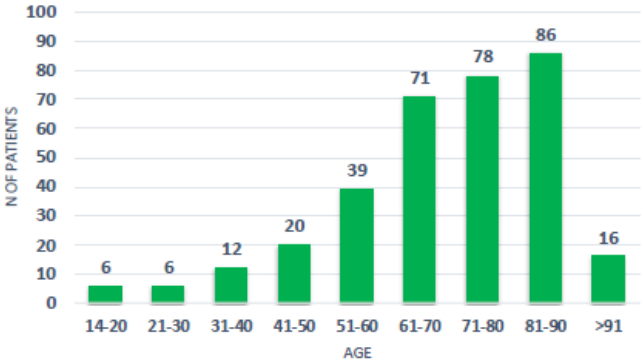
- Periodo di osservazione: 2013 - 2017

Distribuzione degli SE per anno



Media: 88 casi/anno = 1-2 casi/settimana

Distribuzione degli SE in base all'eta'



SDM : DEFINIZIONE

una situazione caratterizzata da una crisi epilettica che e' sufficientemente prolungata o ripetuta a intervalli di tempo cosi' brevi tali da produrre una "condizione epilettica" duratura.

Dizionario dell' Epilessia 1973

PROBLEMI DI DEFINIZIONE

- durata delle crisi?
- perdita di coscienza ?
- ripristino di coscienza ?
- crisi subentranti o "in serie"

CLASSIFICAZIONE (1) (Gastaut e Tassinari, 1975)

GENERALIZZATO	CONVULSIVO	TONICO - CLONICO TONICO CLONICO MIOCLONICO
GENERALIZZATO	NON CONVULSIVO	ABSENCE STATUS ATONICO
PARZIALE	ELEMENTARE	SOMATOMOTORE SOMATOSENSITIVO ADVERSIVO DISFASICO / AFASICO VISIVO
	COMPLESSO	
UNILATERALE		
ERRATICO		

CLASSIFICAZIONE (2)

CONVULSIVO :

- **GENERALIZZATO**
  - **TONICO-CLONICO**
  - **TONICO**
  - **CLONICO**
- **UNILATERALE**
- **FOCALE**

NON CONVULSIVO :

- **CON DISTURBO DELLA COSCIENZA**
- **SENZA DISTURBO DELLA COSCIENZA**

SDM : DEFINIZIONE 2

una condizione in cui l' attivita' epilettica persiste per almeno 30 minuti causando un ampio spettro di sintomi clinici, e con una variabile base neurofisiologica, anatomica ed eziologica (Shorvon 1994)

una crisi che non si arresta dopo una durata che supera quella di gran parte delle crisi nella maggior parte dei pazienti o crisi ripetute senza recupero delle funzioni cerebrali di base fra una e l'altra (Glossario ILAE 2001)

ALTRI : 20 min.?
10 min.?
50% del periodo di registrazione?

Epilepsia, 47(Suppl. 5):9–15, 2006
Blackwell Publishing, Inc.

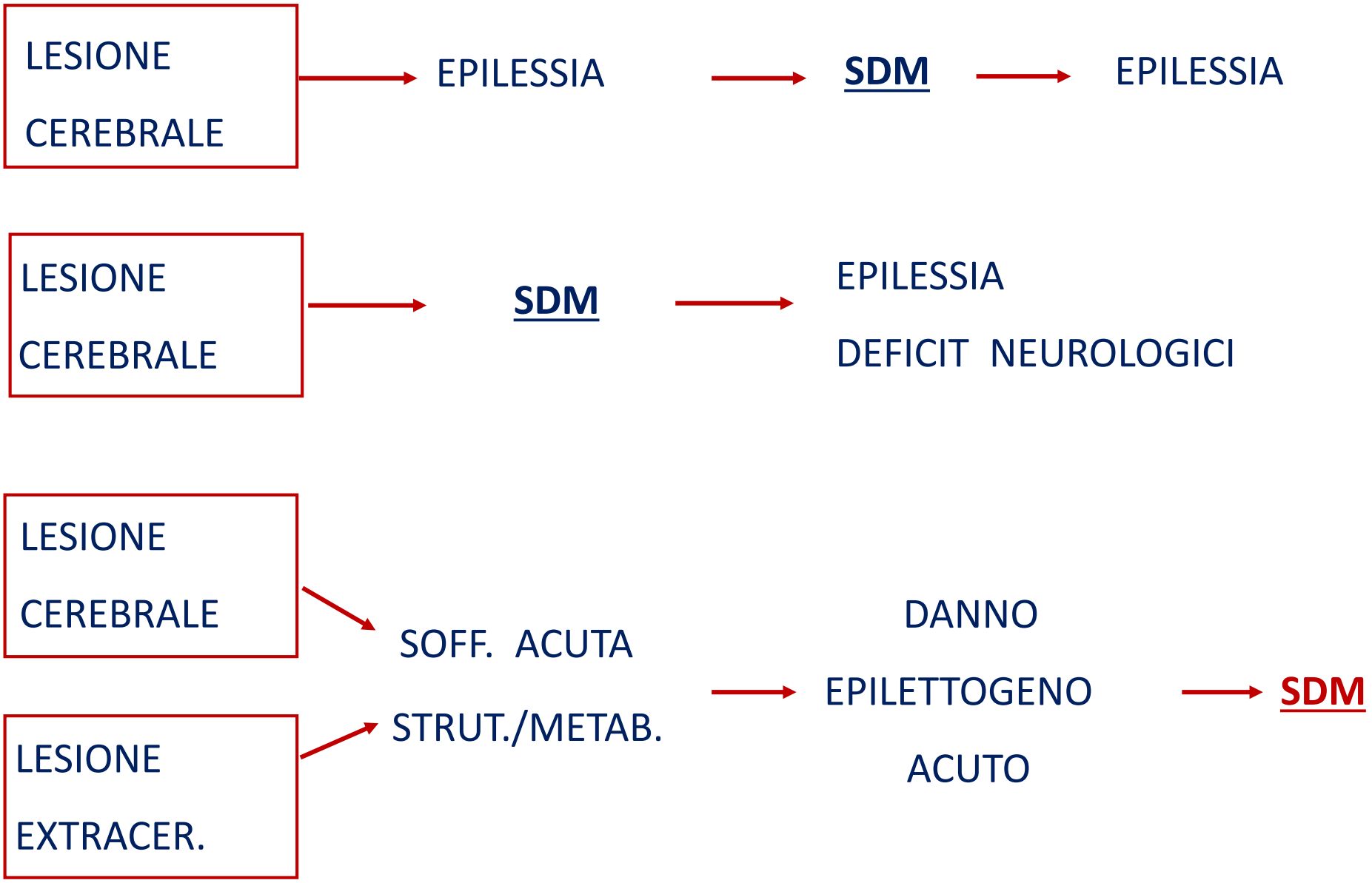
Treatment of Status Epilepticus in Adults: Guidelines of the Italian League Against Epilepsy

*Fabio Minicucci, †Giancarlo Muscas, ‡Emilio Perucca, §Giuseppe Capovilla, ‖Federico Vigevano, and ¶Paolo Tinuper

**Clinical Neurophysiology, San Raffaele Hospital, Milan; †Epilepsy Center, Department of Neurology, Careggi Hospital, Florence; ‡Institute of Neurology, IRCCS C. Mondino Foundation and Clinical Pharmacology Unit, University of Pavia, Pavia; §Department of Child Neuropsychiatry, C. Poma Hospital, Mantova; ‖Neurology Division, Bambino Gesu Children's Hospital, Rome; and ¶Department of Neurological Sciences, University of Bologna, Bologna, Italy*

Uno stato di male epilettico e' una situazione clinica nella quale una crisi epilettica (generalizzata o focale, motoria o no) si prolunga per più di 20 minuti o nella quale le crisi si ripetono a brevissimi intervalli tali da rappresentare una condizione epilettica continua

SDM : POSSIBILI SCENARI



Commenti alle definizioni

- ✓ Dagli anni ‘80 le definizioni di SDM erano basate sul tipo di crisi (convulsive/non convulsive) e sulle caratteristiche EEG (critiche /intercritiche)

- ✓ Almeno la metà dei pazienti con SE non hanno l'epilessia o una specifica sindrome epilettica, ma hanno avuto un danno acuto o pregresso sul sistema nervoso centrale o presentano una malattia sistemica.

- ✓ Pertanto, le definizioni utilizzate in precedenza nella classificazione delle crisi nello SDM dovevano essere modificate.

SPECIAL REPORT

A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus

*†‡Eugen Trinka, §Hannah Cock, ¶Dale Hesdorffer, #Andrea O. Rossetti, **Ingrid E. Scheffer, ††Shlomo Shinnar, ‡‡Simon Shorvon, and §§Daniel H. Lowenstein

Epilepsia, 56(10):1515–1523, 2015
doi: 10.1111/epi.13121

Condizione risultante sia dal fallimento dei meccanismi responsabili della terminazione delle crisi sia dall'attivazione di meccanismi che conducono alla comparsa di crisi abnormemente prolungate .

Si tratta di una condizione che può avere conseguenze a lungo termine, incluse la morte neuronale e l'alterazione di reti neurali, che variano in base alla tipologia e alla durata delle crisi

SPECIAL REPORT

A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus

*†‡Eugen Trinka, §Hannah Cock, ¶Dale Hesdorffer, #Andrea O. Rossetti, **Ingrid E. Scheffer, ††Shlomo Shinnar, ‡‡Simon Shorvon, and §§Daniel H. Lowenstein

Epilepsia, 56(10):1515–1523, 2015
doi: 10.1111/epi.13121

quattro assi per la nuova classificazione dello SE

1: semeiologia
2: eziologia
3: caratteristiche eeg
4: eta' del paziente

1. SEMEIOLOGIA

2 criteri:
presenza/assenza di manifestazioni epilettiche motorie

grado (qualitativo/quantitativo) di compromissione della coscienza

Le forme con prominenti sintomi motori e compromissione della coscienza sono indicate come
SE CONVULSIVO (CSE)
in opposizione alle forme di
SE NON CONVULSIVO (NCSE).

Axis 1: Classification of status epilepticus (SE)

(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. Oculoclonic 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

2. EZIOLOGIA

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)

NB: Non si parla più di SE «IDIOPATICO» o GENETICO», queste definizioni rientrano nel temine «UNKNOWN»

Vengono contemplate poi delle condizioni indeterminate «Boundary syndromes»

Table 3. Currently indeterminate conditions (or "boundary syndromes")

Epileptic encephalopathies
Coma with non evolving epileptiform EEG pattern[a]
Behavioral disturbance (e.g., psychosis) in patients with epilepsy
Acute confusional states, (e.g., delirium) with epileptiform EEG patterns

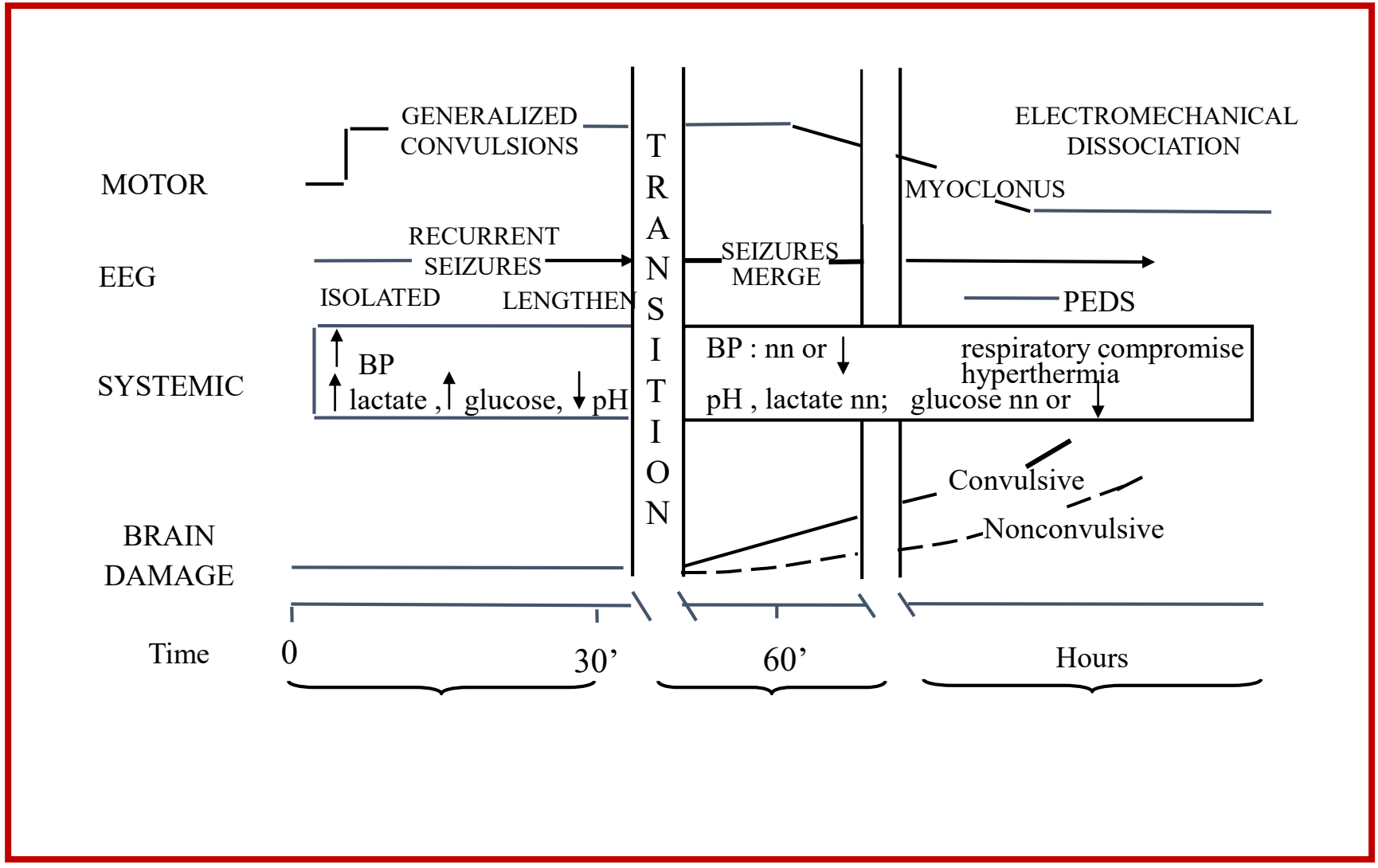
[a]Lateralized and generalized periodic discharges with monotonous appearance are not considered as evolving EEG patterns.[26,27]

3. EEG

1. Localizzazione anomalie (generalizzato, lateralizzato, bilaterale indipendente, multifocale)
2. Denominazione anomalie (scariche periodiche, attività delta ritmica, PO/SW…)
3. Morfologia anomalie (es. onde trifasiche)
4. Decorso temporale delle anomalie
5. Modulazione anomalie (indotte da stimoli/spontanee)
6. Effetti dei farmaci sull'EEG

4. ETA'

1. Neonatale (0-30 giorni)
2. Infantile (30 giorni-2 anni)
3. Fanciullezza (>2 anni-12 anni)
4. Adolescenza ed Età adulta (>12-59 anni)
5. Anziani (>/= 60 anni)


MOTOR
EEG
SYSTEMIC
BRAIN DAMAGE
GENERALIZED CONVULSIONS
RECURRENT SEIZURES
ISOLATED
LENGTHEN
BP
lactate , glucose, pH
TRANSITION
SEIZURES MERGE
ELECTROMECHANICAL DISSOCIATION
MYOCLONUS
PEDS
BP : nn or
respiratory compromise
hyperthermia
pH , lactate nn; glucose nn or
Convulsive
Nonconvulsive
Time
0
30'
60'
Hours

Definizione operativa, che tiene conto della progressione dello SE e quindi anche dei tempi di intervento

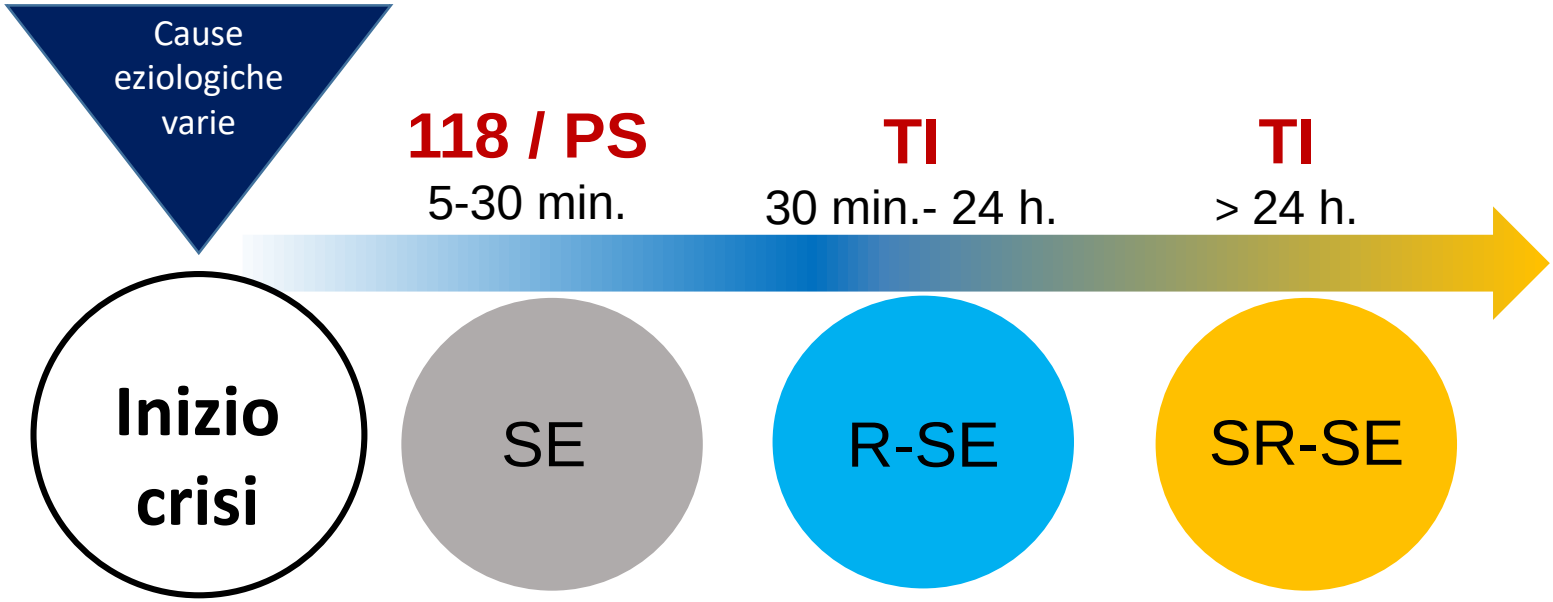
SE “incombente o iniziale” (impending SE): quando una crisi perdura per più di 5 min.

SE “stabilito” (established SE): quando le crisi continuano fino a 30-60 min.

SE “refrattario” > 60 min. Res a T di 1° e 2° linea

Subtle SE: fase in cui le manifestazioni motorie si fanno sempre meno intense, pur essendo evidente il prolungarsi ed il ripetersi delle scariche EEG

SE “super-refrattario” resistente agli anestetici



SE definito: necessita di terapia con AEDs per poter essere controllato
SE refrattario : necessita di ammissione in TI e di t. con anestetici
SE Super-Refrattario: persiste dopo 24 h o ricompare alla sospensione della t. con anestetici

Dimensione operazionale

T1= tempo in cui il trattamento deve essere iniziato
T2= tempo dopo il quale l'attività critica comporta il rischio di conseguenze a lungo termine

Table 1. Operational dimensions with t_1 indicating the time that emergency treatment of SE should be started and t_2 indicating the time at which long-term consequences may be expected

Type of SE	Operational dimension 1 Time (t_1), when a seizure is likely to be prolonged leading to continuous seizure activity	Operational dimension 2 Time (t_2), when a seizure may cause long term consequences (including neuronal injury, neuronal death, alteration of neuronal networks and functional deficits)
Tonic–clonic SE	5 min	30 min
Focal SE with impaired consciousness	10 min	>60 min
Absence status epilepticus	10–15 min[a]	Unknown

[a]Evidence for the time frame is currently limited and future data may lead to modifications.

Table 4. 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)

ILAE 2015

Relevance of clinical context in the diagnostic-therapeutic approach to status epilepticus

U. Aguglia, C.Sueri, S.Gasparini,E.Beghi, A.Labate, A. Gambardella,L.M. Specchio, E. Ferlazzo.
and the Epilepsy Study Group of the Italian Neurological Society and of the Subcommission on Status Epilepticus of the Italian League Against Epilepsy

Epilepsia, 57(9):1521–1530, 2016

Table 1. Etiologies of SE according to different clinical contexts

(1) Etiologies of SE in patients with known epilepsy
- (a) *Triggering factors*: AED withdrawal, febrile illnesses, sleep deprivation, inappropriate AED prescription, reduced AED plasmatic levels (vomiting/diarrhea, drug–drug interactions)
- (b) SE as an integral part of different electroclinical syndromes:
  - (i) Neonatal period and infancy:
    1. Ohtahara syndrome
    2. West syndrome
  - (ii) Childhood and adolescence:
    1. Dravet syndrome
    2. Angelman syndrome
    3. Lennox-Gastaut syndrome
    4. Panayiotopoulos syndrome
    5. Ring chromosome 20 syndrome
    6. Electrical SE during slow wave sleep (ESES) and Landau-Kleffner syndrome
  - (iii) Adolescence and adulthood:
    1. Absence status epilepsy
  - (iv) Any age:
    1. Progressive myoclonic epilepsies
    2. Rasmussen encephalitis
    3. Others (epilepsy associated with *POLG I* mutations, etc)

(2) Etiologies of SE common to patients with and without known epilepsy
- (a) *Acute* (i.e., stroke, intoxication, infectious or autoimmune encephalitis, metabolic disturbances, abrupt drug or alcohol withdrawal, etc.) (see Appendix I in Trinka et al.[1])
- (b) *Remote* (i.e., posttraumatic, postencephalitic, poststroke, etc.) (see Appendix I in Trinka et al.[1])
- (c) *Progressive* (i.e., brain neoplasms, dementias, etc.) (see Appendix I in Trinka et al.[1])

(3) Unknown etiologies ("cryptogenic SE")

International League Against Epilepsy

Nosology and Definitions Task Force (2017-2020)

Chair E.Wirrel, P.Tinuper

Sub- Task Force on Status Epilepticus

Eugen Trinka

grazie per l'attenzione

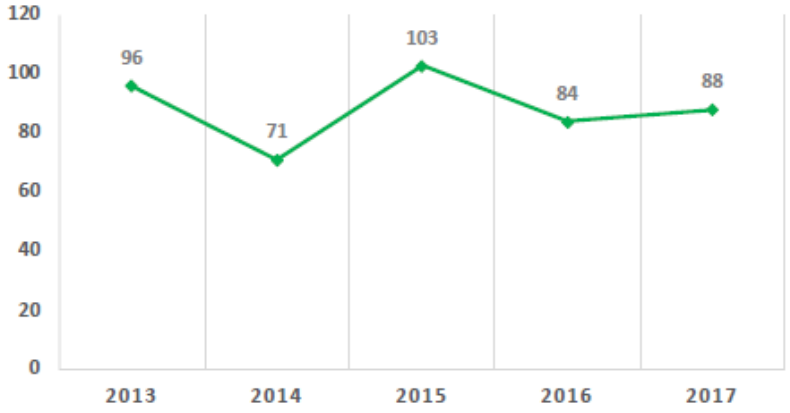
- Periodo di osservazione: 2013 - 2017

Contents lists available at ScienceDirect
Epilepsy & Behavior
journal homepage: www.elsevier.com/locate/yebeh

A one-year prospective study of refractory status epilepticus in Modena, Italy

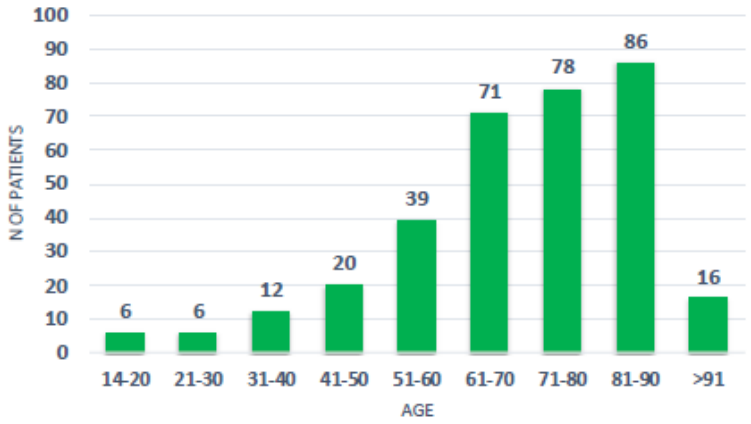
Giada Giovannini [a,b,*], Giulia Monti [a,b], Michela M. Polisi [a,b], Laura Mirandola [a,b], Andrea Marudi [c], Giovanni Pinelli [d], Franco Valzania [b], Massimo Girardis [e], Paolo F. Nichelli [a,b], Stefano Meletti [a,b]

Distribuzione degli SE per anno



Media: 88 casi/anno = 1-2 casi/settimana

Distribuzione degli SE in base all'eta'



*"Condizione risultante sia dal fallimento dei meccanismi responsabili della terminazione delle crisi sia dall'attivazione di meccanismi che conducono alla comparsa di crisi abnormemente prolungate (in seguito ad un dato intervallo temporale **t1**). Si tratta di una condizione che può avere conseguenze a lungo termine (in seguito all'intervallo **t2**), incluse la morte neuronale, il danno neuronale e l'alterazione delle reti neuronali, che variano in base alla tipologia ed alla durata delle crisi".*

Table 1. Operational dimensions with t_1 indicating the time that emergency treatment of SE should be started and t_2 indicating the time at which long-term consequences may be expected

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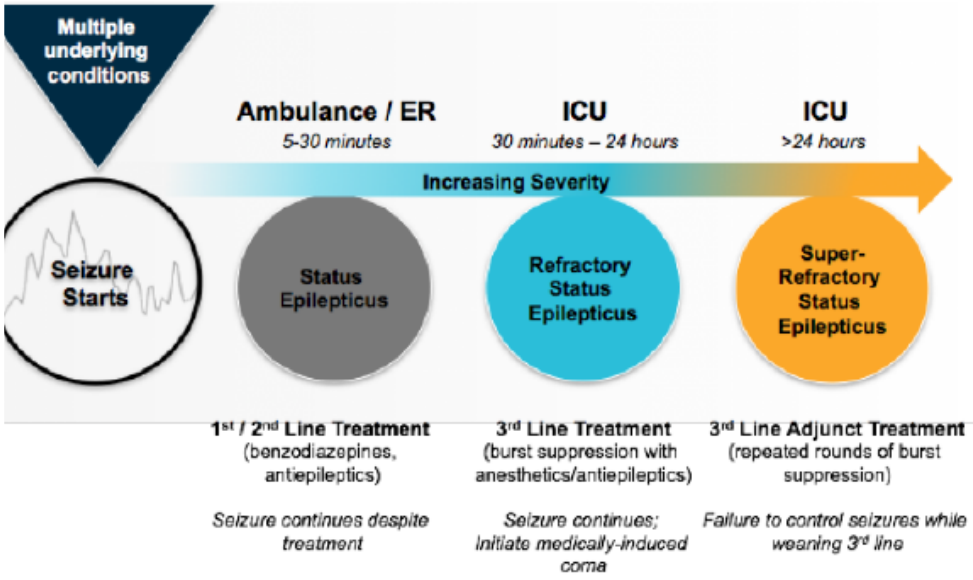
[a]Evidence for the time frame is currently limited and future data may lead to modifications.

ILAE Task force on Classification of Status Epilepticus, Trinka et al, Epilepsia 2015

dalla terapia BDZ

- **SE definito:** SE che necessita di terapia con AEDs per poter essere controllato
- **SE refrattario (RSE):** SE che necessita di ammissione in terapia intensiva e di terapia anestesiologica
- **SE Super-refrattario (SRSE):** SE che persiste dopo 24 h o ricorre al tentativo di sospensione della terapia anestesiologica.



BDZ: Benzodiazepine; AEDs: Anti-epileptic drugs

J Neurol (2008) 255:1561–1566
DOI 10.1007/s00415-008-0989-1

ORIGINAL COMMUNICATION

Andrea O. Rossetti
Giancarlo Logroscino
Tracey A. Milligan
Costas Michaelides
Christiane Ruffieux
Edward B. Bromfield

Status Epilepticus Severity Score (STESS)

A tool to orient early treatment strategy

	Features	STESS
Consciousness	Alert or somnolent/confused	0
	Stuporous or comatose	1
Worst seizure type	Simple-partial, complex-partial, absence, myoclonic*	0
	Generalized-convulsive	1
	Nonconvulsive status epilepticus in coma	2
Age	< 65 years	0
	≥ 65 years	2
History of previous seizures	Yes	0
	No or unknown	1
Total		0–6

* complicating idiopathic generalized epilepsy

Score favorevole: 0-2

All

- Advisory Sub Committee
- Fundraising Resource Development
- Global Outreach Task Force
- Guidelines Task Force
- Industry Liaison Task Force
- Joint ILAE-IFCN EEG Database Task Force
- Leadership Development Task Force
- Next Generation Task Force
- Nosology and Definitions Task Force
- Publications Task Force
- Research Advocacy Task Force

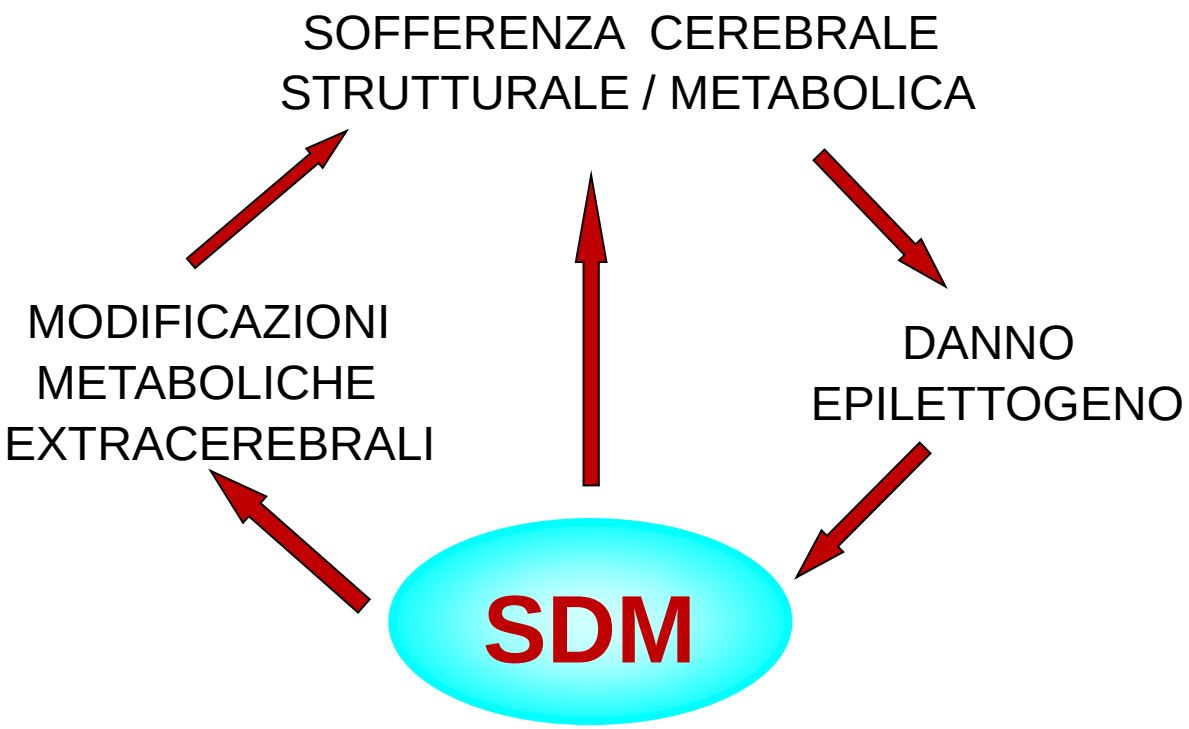
quattro definizioni per la nuova classificazione dello SE

DEF. 1: SEMEIOLOGICA

DEF. 2: EZIOLOGICA

DEF. 3: CARATTERISTICHE EEG

DEF. 4: ETA' DEL PAZIENTE



MORTALITA': 6-30% dei casi

Evaluation of a clinical tool for early etiology identification in status epilepticus

*†‡#Vincent Alvarez, †M. Brandon Westover, ‡Frank W. Drislane, *Barbara A. Dworetzky, §David Curley, *|Jong Woo Lee, and ¶|Andrea O. Rossetti

Epilepsia, **(*):1–10, 2014
doi: 10.1111/epi.12852

Table 1. List of diagnostic categories and their frequencies as definitive SE etiology

Underlying etiology after complete workup (n = 212)	n	%
Total, n = 212		
ASD-related (nonadherence, recent change or low levels)	34	16.04
Brain tumor without acute change (no change or increase in tumor load)	28	13.21
Acute hemorrhagic cerebrovascular event	21	9.91
Known epilepsy (non structural) without provocative factors (breakthrough seizures)	16	7.55
Remote ischemic cerebrovascular event	14	6.6
Unclassified[a]	13	6.13
CNS infection (meningitis or encephalitis)	12	5.66
Unknown origin	11	5.19
Toxic-metabolic	10	4.72
Systemic infection/sepsis	10	4.72
Remote hemorrhagic cerebrovascular event	8	3.77
Acute TBI	7	3.3
Acute ischemic cerebrovascular event	5	2.36
Remote TBI	6	2.83
Alcohol related (withdrawal or intoxication)	6	2.83
Brain tumor with acute change (bleeding, recent biopsy/surgery or rapid increase in edema)	5	2.36
Benzodiazepine withdrawal	4	1.89
Neurodegenerative disease	2	0.94
Other drugs known to reduce seizure threshold	0	0

ASD, antiseizure drug; CNS, central nervous system; TBI, traumatic brain injury.

[a]Unclassified includes: three multiple sclerosis, two confirmed and one possible posterior reversible encephalopathy syndrome (PRES), two tumoral meningitis, one NMDA encephalitis, one neurosarcoidosis, one eclampsia, one arteriovenous malformation without bleeding, and one case of microangiopathic hemolytic anemia.

Figure 1.

The Status Epilepticus Etiology Identification Tool (SEEIT).

The SEEIT tool has been designed to guide SE etiology assessment. It has to be used along with antiseizure drug protocol. Each point has to be assessed.

Epilepsia © ILAE

1. Is it really a Status Epilepticus (SE) episode?	⇩	
A. Any signs for Psychogenic Non-Epileptic Seizure (opposition to eyes opening, « waxing and waning » movements, pelvic thrusting, stopped or induced by suggestion)	Yes → avoid AED escalation	No
B. Seizures lasting more than 5 min or repeated seizures without regain of consciousness	Yes ⇨ Point 2	No →Review dx
2. A) Previous seizures, known epilepsy, or B) De novo seizures but known structural brain damage (stroke, trauma, old meningo-encephalitis…) or C) De novo seizure but known brain progressive condition (dementia, tumor…)? • *Specify:*	Yes ⇩	No ⇨ Point 3
A. AED non-compliance / recent decrease dosage / low level: *Specify:*.....	Yes	No
B. Systemic infection: *Specify:*.....	Yes	No
C. Alcohol or drug (incl. benzodiazepine and illicit drugs) withdrawal or acute intoxication • Consider toxicology screen: *Specify:*.....	Yes	No
D. Significant metabolic disturbances (i.e. blood glucose and natremia). *Specify:*.....	Yes	No
E. Progression or change in previous neurological condition clinically or radiology (e.g., new symptoms, tumor progression, bleeding, biopsy, recent surgery…): *Specify:*	Yes	No
F. Refractory SE without clear explanation, new neurological abnormality, neck stiffness or fever without systemic explanation. *Specify:*.....	Yes ⇨ point 4	No
G. None of these.	Yes → no acute etiology→ adapt AED ttt	
3. De novo SE (first ever seizure) without known brain disease?	Yes ⇩	No ⇨ Point 2
A. Newly diagnosed, previously asymptomatic structural brain damage or EEG suggesting Idiopathic Generalized Epilepsy (IGE) / Genetic Generalized Epilepsy (GGE): *Specify:*.....	Yes	No
B. Acute brain lesion (ischemic or hemorrhagic stroke, cerebral venous thrombosis, SAH, SDH, traumatic brain injury, encephalitis…): *Specify:*.....	Yes	No
C. Alcohol or drug (incl. benzodiazepine and illicit drugs) withdrawal or acute intoxication • Consider toxicology screening: *Specify:*.....	Yes	No
D. Significant metabolic disturbance (i.e. blood glucose and natremia). *Specify:*.....	Yes	No
E. Severe systemic infection (sepsis): *Specify:*.....	Yes	No
F. Refractory SE without clear explanation, neck stiffness or fever without systemic explanation. *Specify:*.....	Yes ⇨ point 4	No
G. None of these with favorable evolution under AED	Yes →SE possibly cryptogenic, but consider MRI if clinical/EEG focal sign and normal CT	
4. Fever or systemic inflammatory response without extra-neurological infectious process, meningeal sign, unusual headache, recent behavior change, or refractory SE without clear etiology • *Specify:*..... • Think about IV empirical antimicrobial therapy for meningo-encephalitis and blood culture, then lumbar puncture if no contraindication	Yes ⇩	No ⇨ Stop
A. Normal CSF (pay attention to normal CSF in early phase of encephalitis)? • CSF details:.....	Normal CSF→ May be cryptogenic. Consider autoimmune disease	Abnormal CSF→ Empirical antimicrobial therapy. Consider autoimmune causes
➢ **Suspected etiology after the first evaluation:**		

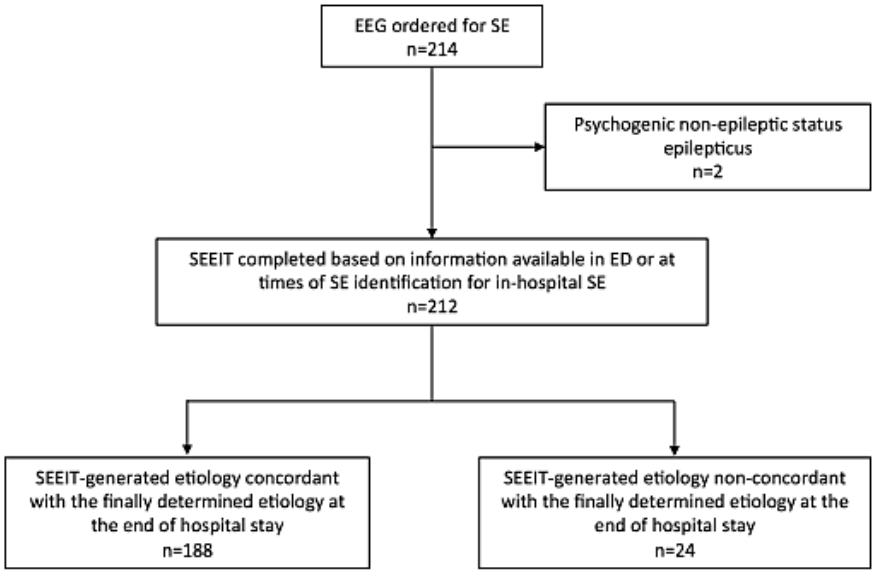


Figure 2.
Study profile. EEG, electroencephalography; SE, status epilepticus; SEEIT, Status Epilepticus Etiology Identification Tool.

A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus

*†‡Eugen Trinka, §Hannah Cock, ¶Dale Hesdorffer, #Andrea O. Rossetti, **Ingrid E. Scheffer, ††Shlomo Shinnar, ‡‡Simon Shorvon, and §§Daniel H. Lowenstein

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Table 1. Operational dimensions with t_1 indicating the time that emergency treatment of SE should be started and t_2 indicating the time at which long-term consequences may be expected

Type of SE	Operational dimension 1 Time (t_1), when a seizure is likely to be prolonged leading to continuous seizure activity	Operational dimension 2 Time (t_2), when a seizure may cause long term consequences (including neuronal injury, neuronal death, alteration of neuronal networks and functional deficits)
Tonic–clonic SE	5 min	30 min
Focal SE with impaired consciousness	10 min	>60 min
Absence status epilepticus	10–15 min[a]	Unknown

[a]Evidence for the time frame is currently limited and future data may lead to modifications.

Table 2. Axis 1: Classification of status epilepticus (SE)

(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. Oculoclonic 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

Table 2. Cohort description

Patients (n = 212)		
Demographics		
Age (median, range)	60	18–93
Male (n,%)	106	50
History of previous seizures (n,%)	104	49.1
Center (n,%)		
CHUV	104	49.1
BWH	65	30.7
MGH	30	14.2
BIDMC	13	6.1
SE characteristics		
Worst seizure type (n,%)		
Focal without consciousness impairment	32	15.1
Focal with consciousness impairment	57	28.9
Absence	3	1.42
Myoclonic	1	0.5
Generalized convulsive	102	48.1
Nonconvulsive SE in coma	17	8
Level of consciousness before treatment (n,%)		
Alert	24	11.3
Confused	51	24.1
Somnolent	13	6.1
Stuporous	88	41.5
Comatose	36	17
STESS (mean, SD)	2.64	1.63
Refractory SE (n,%)	119	56.12
Number of different ASD used (median, range)	3	0–13
Coma induction for SE control (n,%)	24	11.3
Outcome at discharge (n,%)		
Return to clinical premorbid baseline	96	45.3
New morbidity	89	42
Death	27	12.8

ASD, antiseizure drug; BWH, Brigham and Women's Hospital; BIDMC, Beth Israel Deaconess Medical Center; CHUV, Lausanne University Hospital; MGH, Massachusetts General Hospital; STESS, Status Epilepticus Severity Score.

Table 3. Currently indeterminate conditions (or "boundary syndromes")

Epileptic encephalopathies
Coma with non evolving epileptiform EEG pattern[a]
Behavioral disturbance (e.g., psychosis) in patients with epilepsy
Acute confusional states, (e.g., delirium) with epileptiform EEG patterns

[a]Lateralized and generalized periodic discharges with monotonous appearance are not considered as evolving EEG patterns.[26,27]

Table 4. 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)

Table 5. SE in selected electroclinical syndromes according to age

SE occurring in neonatal and infantile-onset epilepsy syndromes
Tonic status (e.g., in Ohtahara syndrome or West syndrome)
Myoclonic status in Dravet syndrome
Focal status
Febrile SE
SE occurring mainly in childhood and adolescence
Autonomic SE in early-onset benign childhood occipital epilepsy (Panayiotopoulos syndrome)
NCSE in specific childhood epilepsy syndromes and etiologies (e.g., Ring chromosome 20 and other karyotype abnormalities, Angelman syndrome, epilepsy with myoclonic-atonic seizures, other childhood myoclonic encephalopathies; see Appendices 1–3)
Tonic status in Lennox-Gastaut syndrome
Myoclonic status in progressive myoclonus epilepsies
Electrical status epilepticus in slow wave sleep (ESES)
Aphasic status in Landau-Kleffner syndrome
SE occurring mainly in adolescence and adulthood
Myoclonic status in juvenile myoclonic epilepsy
Absence status in juvenile absence epilepsy
Myoclonic status in Down syndrome
SE occurring mainly in the elderly
Myoclonic status in Alzheimer's disease
Nonconvulsive status epilepticus in Creutzfeldt-Jakob disease
De novo (or relapsing) absence status of later life

These forms of SE may be encountered prevalently in some age groups, but not exclusively.