
Definition and Classification of Epilepsies



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Definition of epilepsy

A practical clinical definition of epilepsy

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 Glynn, ¶¶Dale C. Hesdorffer, ##B.I. Lee, ***Gary W. Mathern, †††Solomon L. Moshé,
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 ****Samuel Wiebe

Definition of epilepsy

: A disease of the brain defined by any of the following conditions

1. At least **two unprovoked** (or reflex) **seizures** occurring **> 24 hr** apart
2. **One unprovoked** (or reflex) **seizure** and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome

Diagnosis ≠ Treatment

Terminology: Provoked seizure

- **Provoked seizure**: acute symptomatic seizure;
 - Electrolyte disturbances
 - Acute toxic effects (antidepressants, sympathomimetics, etc.)
 - Withdrawal syndromes (BZP, ethanol, etc.)
 - Sepsis, infection, HIE, traumatic brain injury, stroke, tumor
 - Fever, sleep deprivation, inflammatory
- **An unprovoked seizure** does not have an immediate cause.

Terminology: Reflex seizure

- อาการชักที่เกิดขึ้นจากตัวกระตุ้นที่เฉพาะเจาะจง (certain trigger/stimulus)
- Simple reflex seizure = light/photic, touch
- Complex reflex seizure = hot bath, read, chew, write, tooth brushing, doing arithmetic

Scenario 1

- เด็กชายอายุ 10 ปี ญาตินำส่ง รพ เนื่องจากขณะที่นอนอยู่กลางคืน นอนกำมือกับแม่ (ปกติ เวลานอนเด็กมักกำมือกับแม่) และแม่รู้สึกว่ามีมือเกร็งแน่นกว่าปกติ ไม่มีอาการกระตุก ไม่มีอาการตาเหลือก ระยะเวลาไม่ชัดเจน แม่ปลุกนำส่ง รพ ตอนจะขึ้นรถ เด็กพูดว่า “ไป รพ กัน”
- เมื่อถึง รพ ตรวจร่างกายเป็นปกติ
- วินิจฉัย ??
- ให้การรักษา อย่างไร ??

Comment:

1. Seizure vs non-seizure
2. Basic investigations
3. EEG

Resolved Epilepsy (หาย)

- Epilepsy → treatment → seizure free period (2 yrs in kid vs 4 yrs in adult) → tapering AED
- Task Force ILAE: 1. individuals who had an age-dependent epilepsy syndrome but are now past the applicable age
 2. who have remained seizure-free for the last 10 years, with no medicines for the last 5 years
- Resolved epilepsy ≠ Remission ≠ Cure

Scenario 2

- ผู้ป่วยชาย เมื่อมีอายุ 16 ปี มีอาการชักเกร็งกระตุกทั้งตัว ระยะเวลา 1 นาที และ มีอาการ 2 ครั้งใน 6 เดือน นอกจากนั้น ในอดีตยังมีอาการสะดุ้งบ่อย ๆ
- EEG: Generalized fast 4.5 Hz spike/wave discharges
- วินิจฉัยเป็น โรคลมชัก รับประทานยากันชัก อยู่จาก 16 ปี ถึง 20 ปี ผู้ป่วยถามว่าจะหายจากโรคลมชักเมื่อไหร่

Comment:

1. Epileptic syndrome: JME
2. Advise to continue ASM

Scenario 3

A 6 year-old boy has had 2 seizures 3 days apart while playing a videogame involving flashing lights. There have been no other seizures. EEG shows an abnormal photoparoxysmal response

Comment

- : Epilepsy, even provoked by light but this is reflex epilepsy
- : Predisposition to have seizure with light flashes

Epilepsy classification

ILAE 1981

Clinical seizure type

1. Partial (focal, local) seizures

Simple partial sz

- with motor signs
- with somatosensory symptoms
- with autonomic symptoms and signs
- with psychic symptoms

Complex partial sz

- start with SPS followed by impairment of consciousness
- with impairment of consciousness at onset

Partial sz evolving to 2^o gen sz

- SPS → GTC
- CPS → GTC
- SPS → CPS → GTC

2. Generalized sz (convulsive and non-convulsive)

Absence, Myoclonic, Clonic, Tonic, Tonic-clonic, Atonic

3. Unclassified epileptic sz

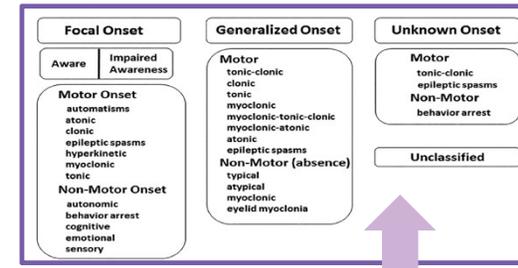
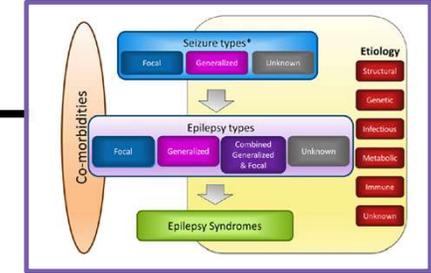
4. Prolonged or repetitive seizure (status epilepticus)

EEG sz type

EEG interictal expression

A 5 axes diagnostic scheme

1. Ictal phenomenology
2. Seizure type
3. Epilepsy syndrome
4. Etiology
5. Impairment



1981, 1985

1989

2001

2010-13

2017

ILAE 1989

1. Localization-related epilepsies and syndromes

- 1.1 Idiopathic
 - benign childhood epilepsies with centro-temporal spike
 - childhood epilepsy with occipital paroxysms
 - primary reading epilepsy
- 1.2 Symptomatic e.g. TLE, FLE, PLE, OLE
- 1.3 Cryptogenic

ILAE 1989

2. Generalized epilepsies and syndromes

- 2.1 Idiopathic (with age-related onset, listed in order to age)
 - Benign neonatal familial convulsions
 - Benign neonatal convulsions
 - Childhood absence epilepsy (pyknolepsy)
 - Juvenile absence epilepsy
 - Juvenile myoclonic epilepsy
 - Epilepsy w grand mal (GTCS) sz on awakening
- 2.2 Cryptogenic or symptomatic (in order to age)
 - West syndrome
 - Lennox-Gastaut syndrome
 - Epilepsy w myoclonic-astatic sz
 - Epilepsy w myoclonic absences

ILAE 1989

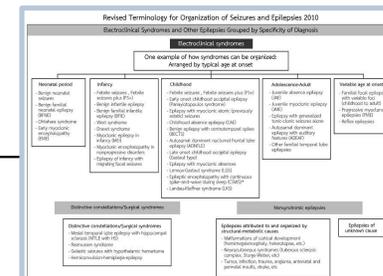
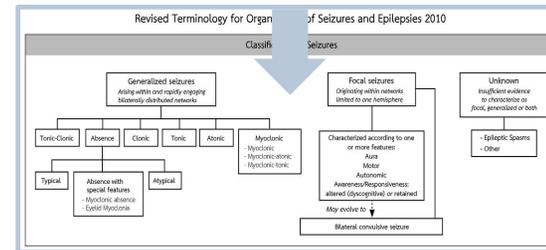
2. Generalized epilepsies and syndromes

- 2.3 Symptomatic
 - 2.3.1 Non specific etiology
 - EME
 - EME w supression burst
 - other symptomatic generalised epilepsies not defined above
 - 2.3.2 Specific syndromes/etiologies
 - Cerebral malformation
 - BSA
3. Epilepsies and syndromes undetermined whether focal or generalized e.g. SMEI, LKS, CSWS, neonatal sz
4. Special syndromes e.g. FC, reflex epilepsy, isolated sz

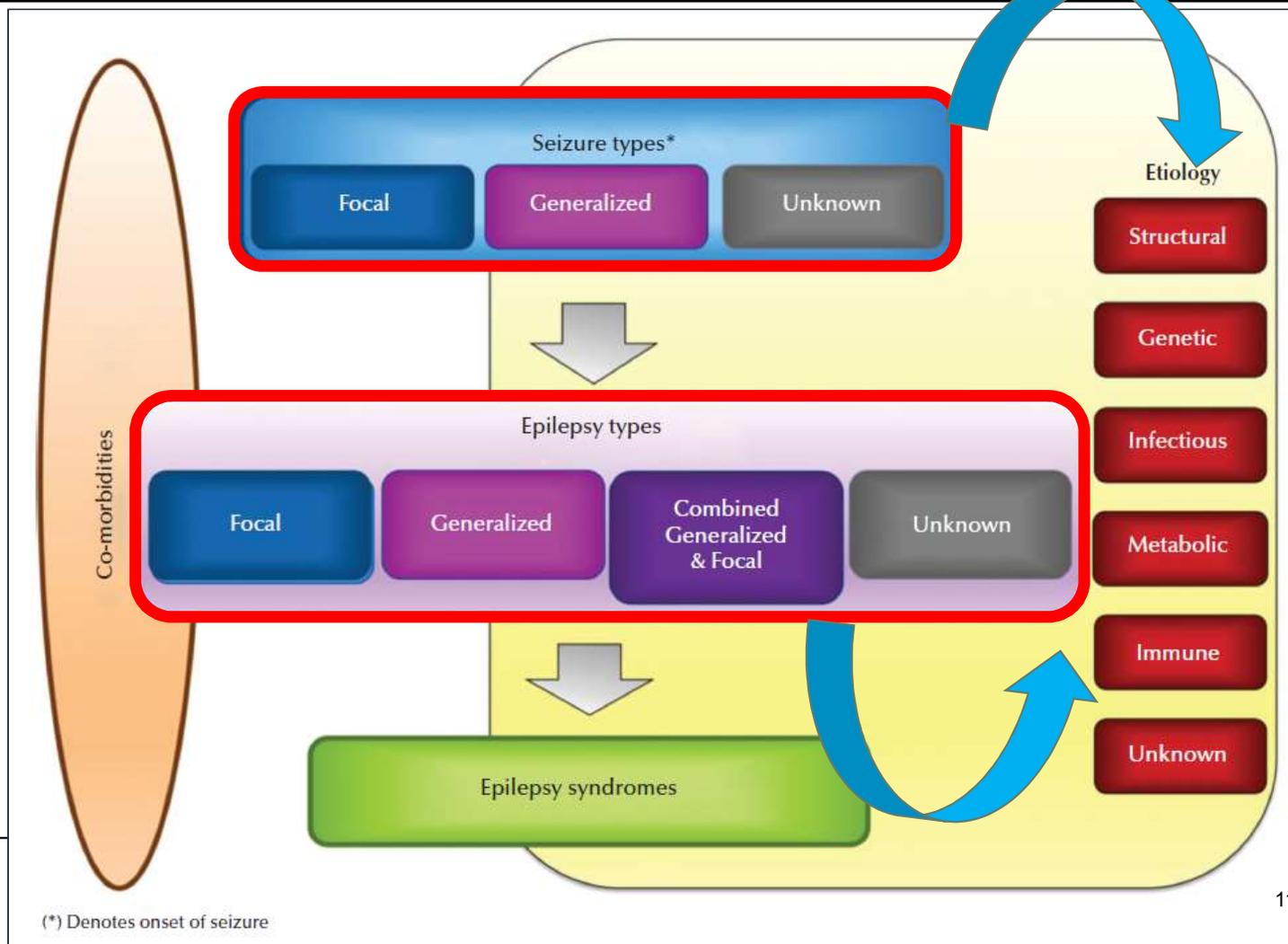
TWO DICHOTOMIES, A 4-PART CLASSIFICATION

	Localization-related	Generalized
Idiopathic	Localization-related idiopathic	Generalized idiopathic
Symptomatic	Localization-related symptomatic	Generalized symptomatic
	IPE	IGE
	SPE	SGE

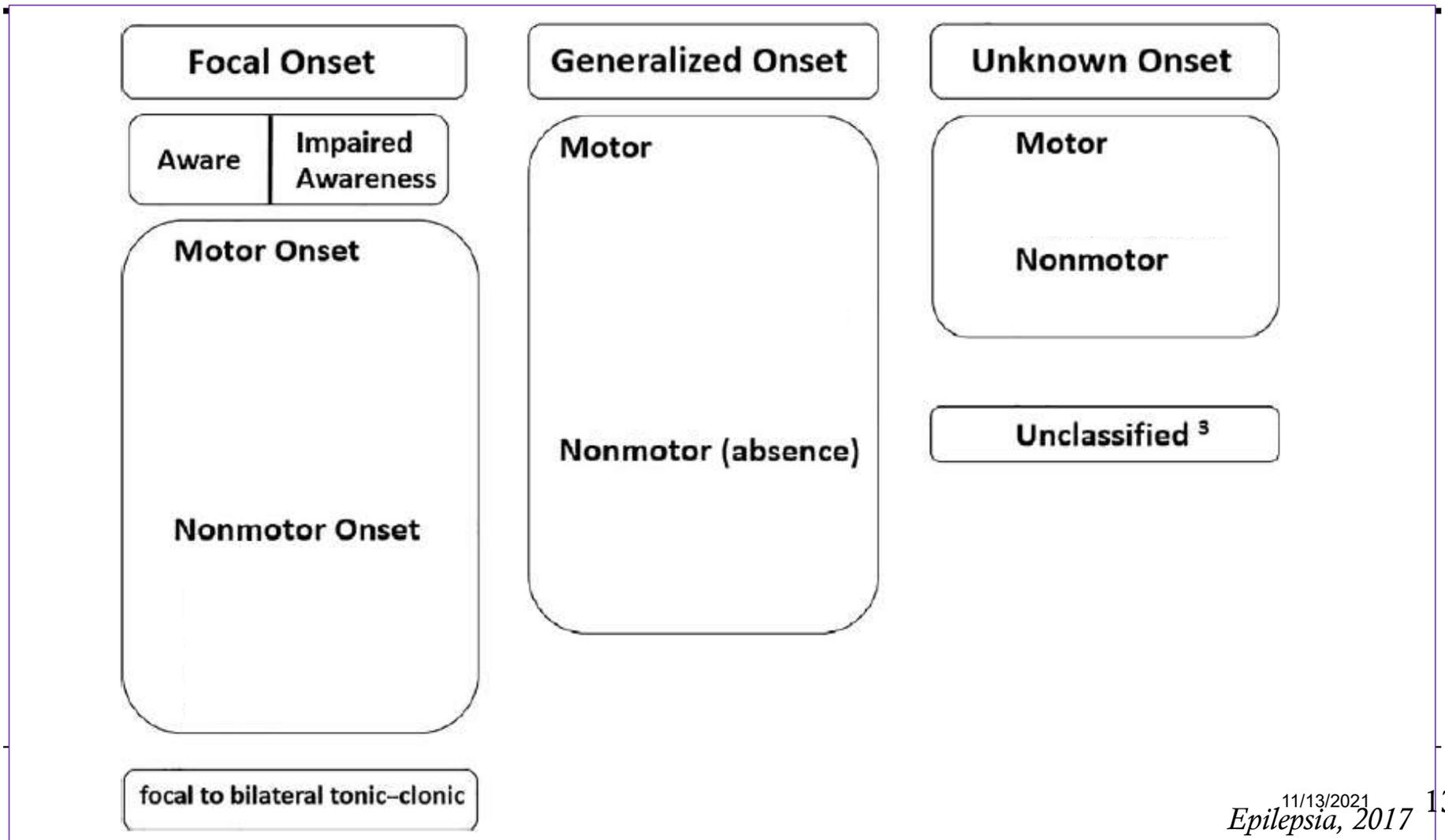
Cryptogenic Special syndromes



2017: Framework for epilepsy classification



ILAE 2017 Classification of seizure type: **basic version**



ILAE 2017 Classification of Seizure Types Expanded Version ¹

Focal Onset

Aware

Impaired
Awareness

Motor Onset

automatisms
atonic ²
clonic
epileptic spasms ²
hyperkinetic
myoclonic
tonic

Nonmotor Onset

autonomic
behavior arrest
cognitive
emotional
sensory

focal to bilateral tonic-clonic

Generalized Onset

Motor

tonic-clonic
clonic
tonic
myoclonic
myoclonic-tonic-clonic
myoclonic-atonic
atonic
epileptic spasms

Nonmotor (absence)

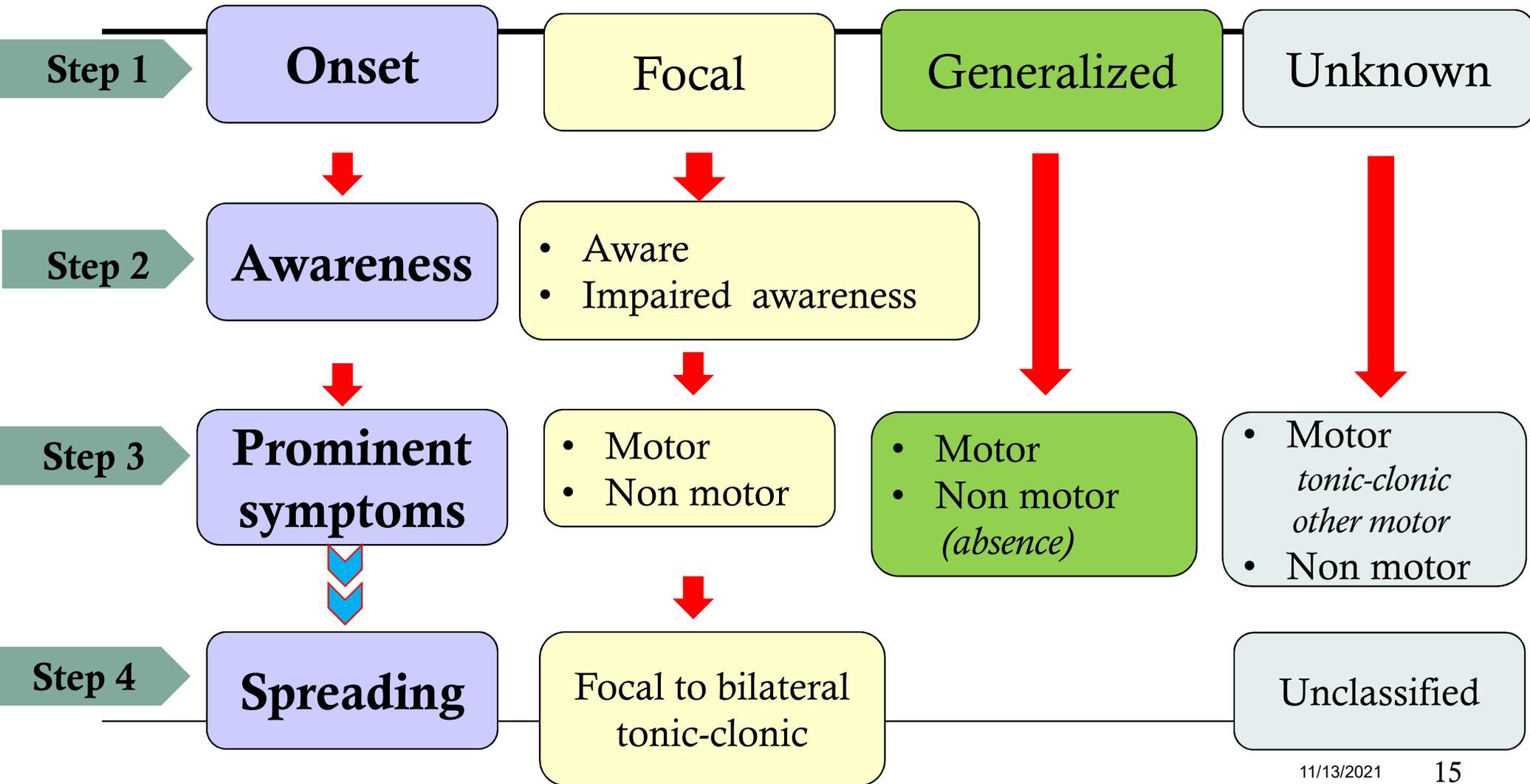
typical
atypical
myoclonic
eyelid myoclonia

Unknown Onset

Motor

tonic-clonic
epileptic spasms
Nonmotor
behavior arrest

Unclassified ³



Step 1

Onset

Focal

Generalized

Unknown



If you have further information (VDO, EEG, MRI, details) in any level, seizure type can be changed



Step 4

Spreading

Focal to bilateral tonic-clonic

• Non motor

Unclassified

Step 1

Onset

Focal

Generalized

Unknown

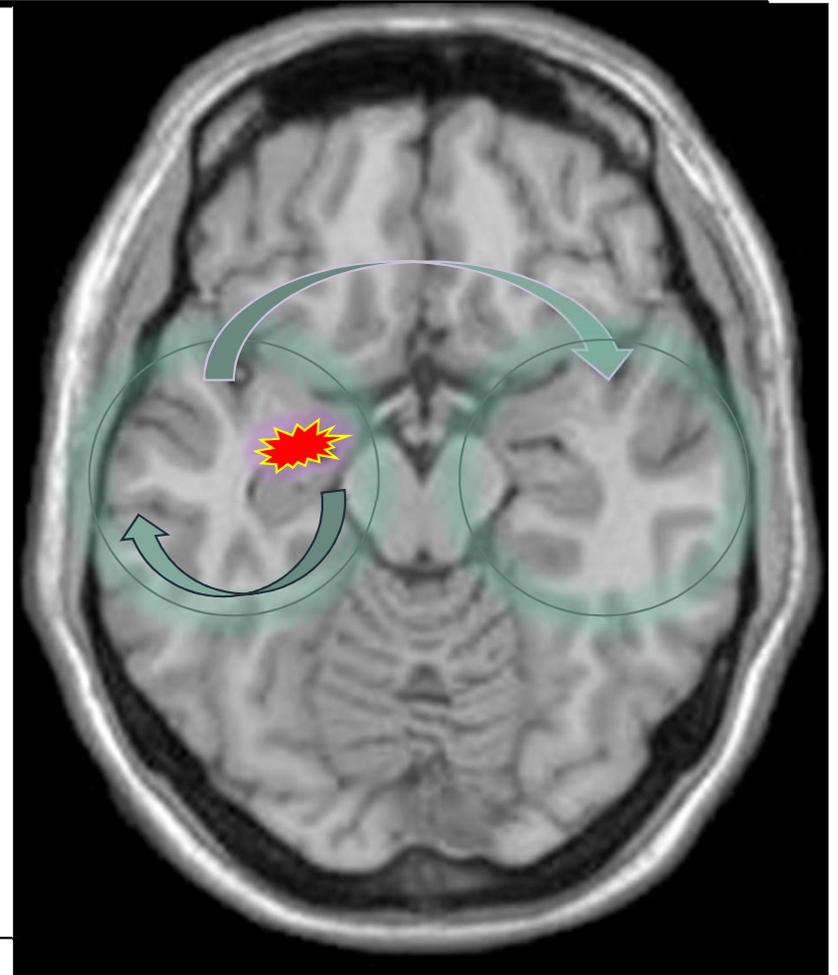


- 80% Confidence level
- Try to look for lateralizing signs
- Concept of focal vs generalized seizure (2010)

2010

Focal seizures

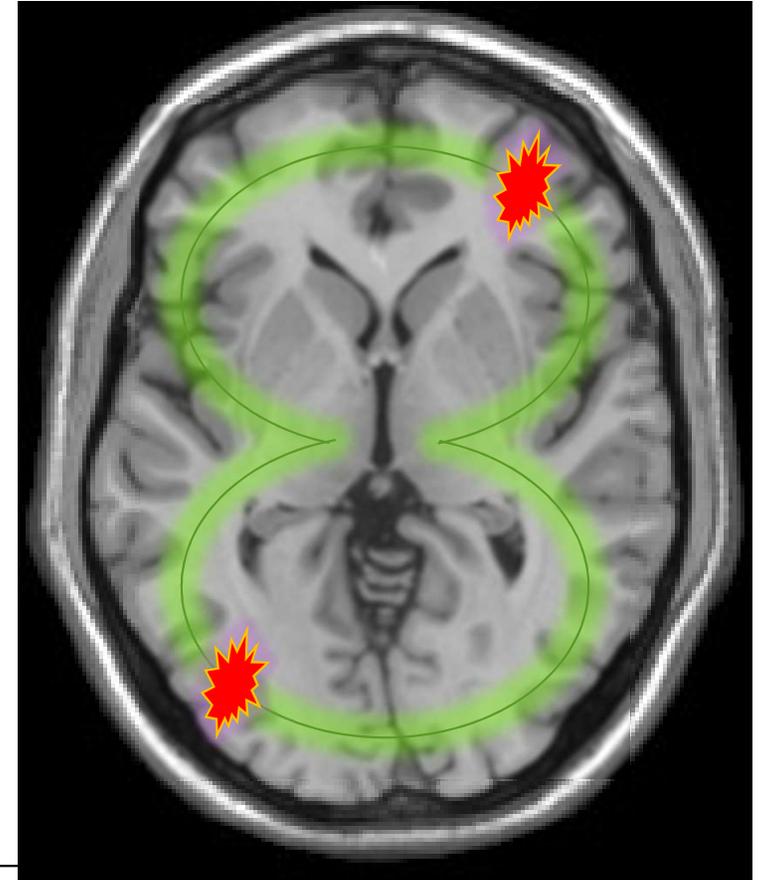
- Originate within networks limited to one hemisphere
- May be discretely localized or more widely distributed



2010

Generalized seizures

- Originate at some point within and **rapidly engage** bilaterally distributed networks
- Can include cortical and subcortical structures but not necessarily the entire cortex



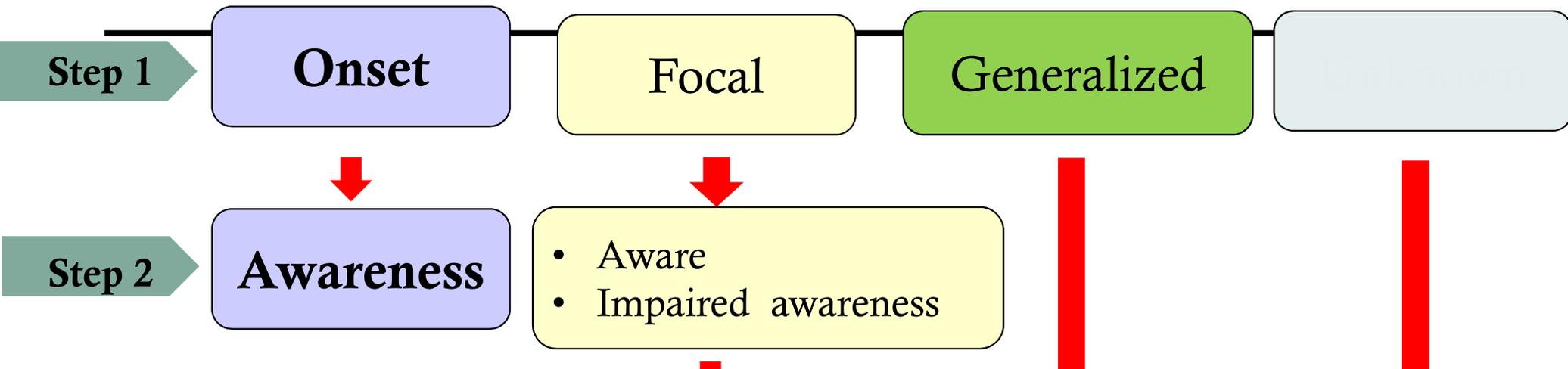
Step 1: Onset

Focal

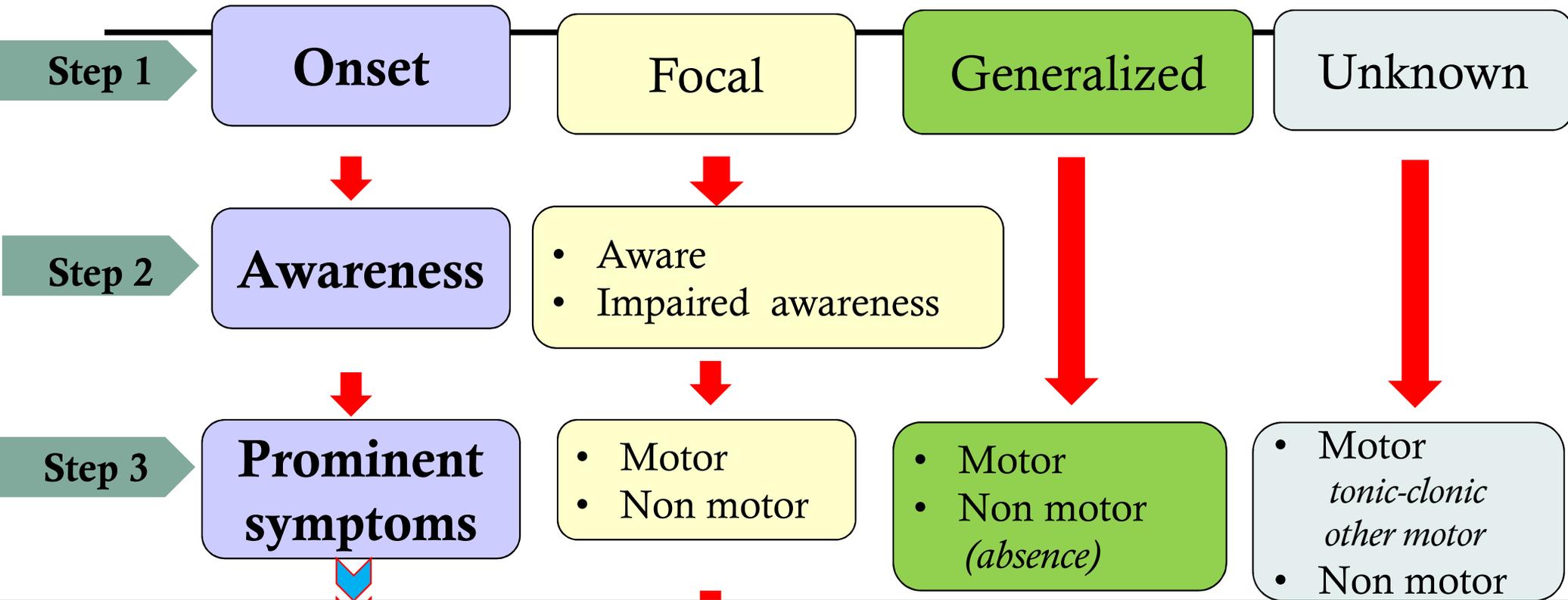


Unknown



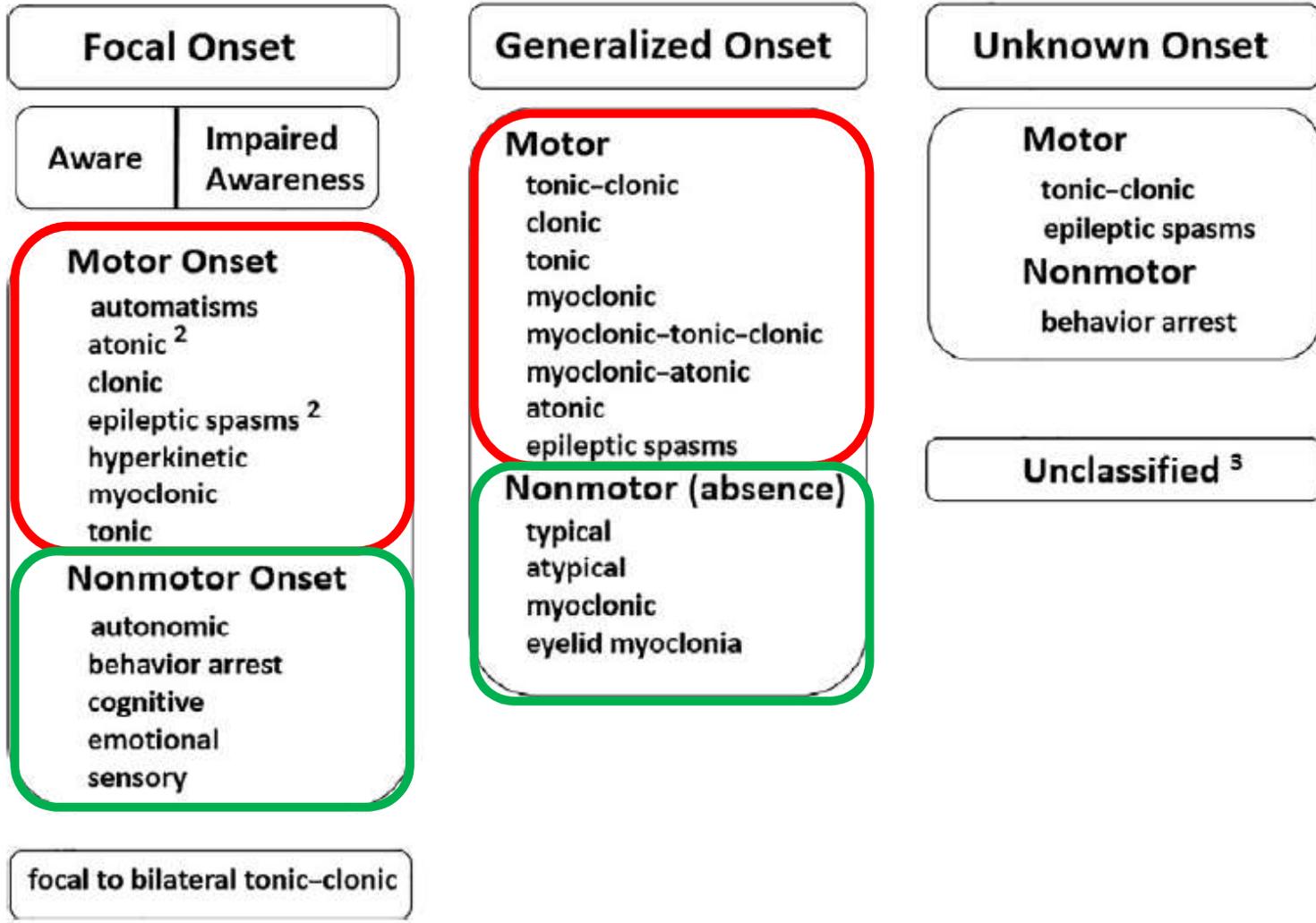


- Awareness = knowledge of self and environment
- Knowledge of events occurring during seizure
(not to knowledge of whether a seizure has occurred)
- Evaluate awareness during ictal period, not post-ictal period
- Can skip this step if we do not have enough information

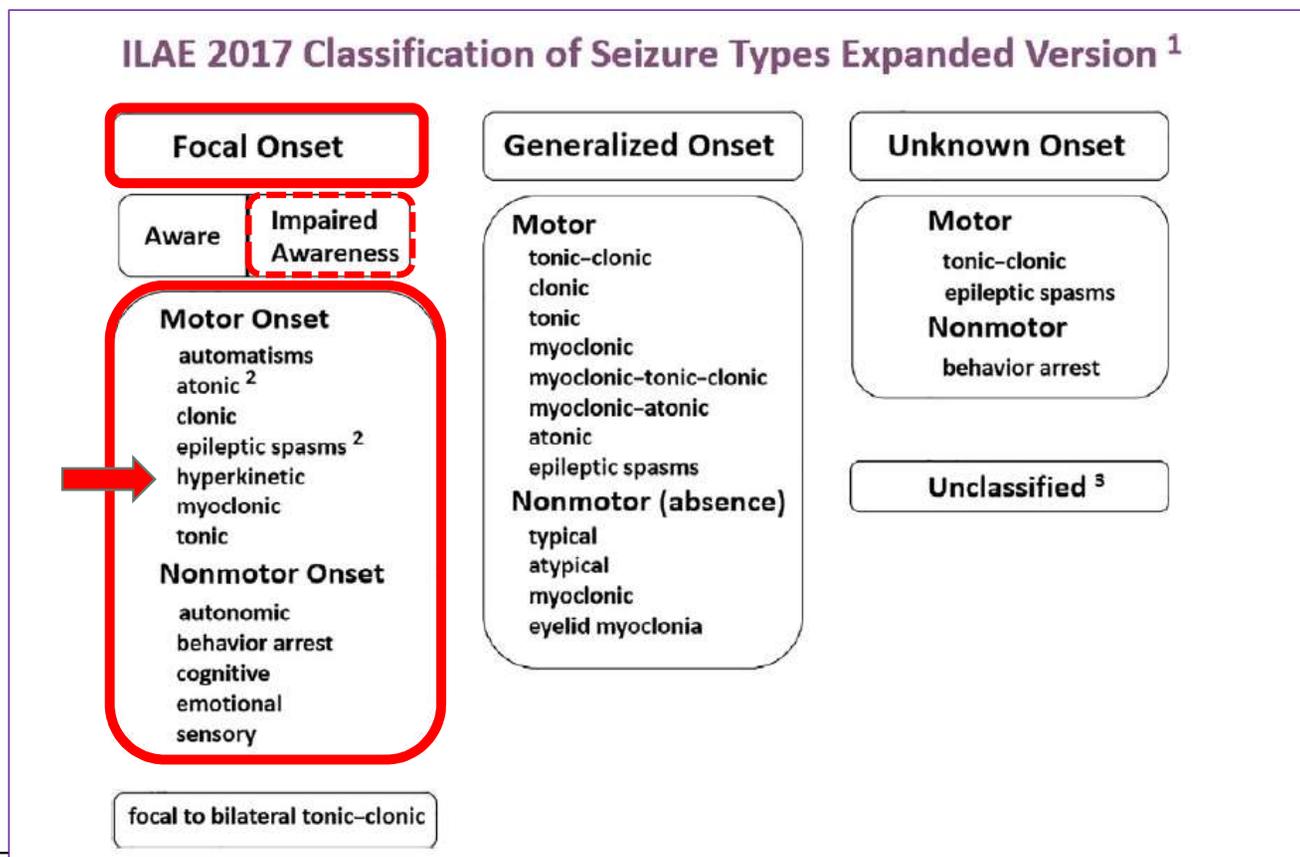


- Earliest symptom
- Motor **vs** Non motor

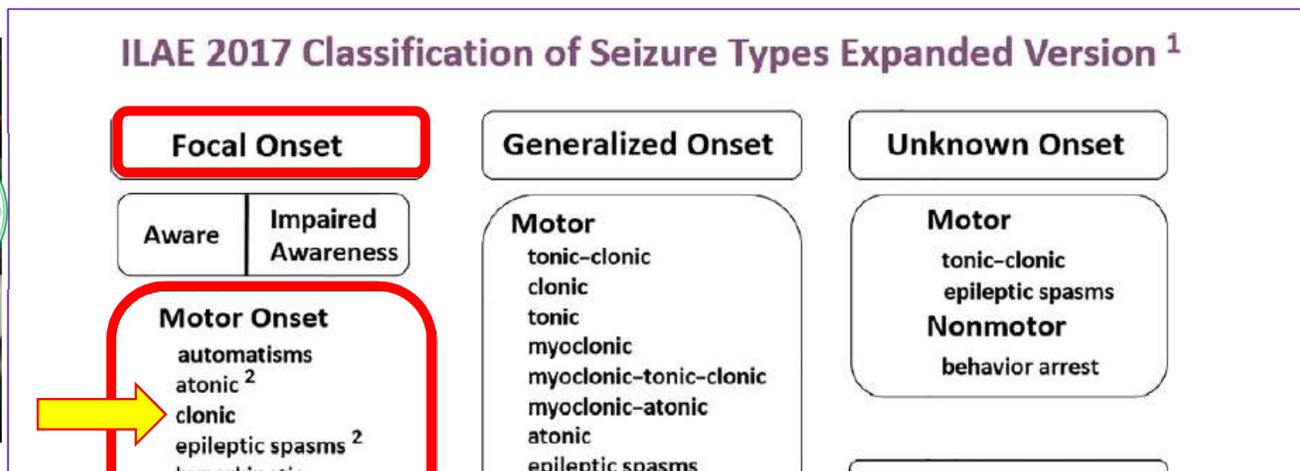
ILAE 2017 Classification of Seizure Types Expanded Version ¹



Prominent symptoms: **motor** vs non motor onset



Prominent symptoms: motor vs non motor onset



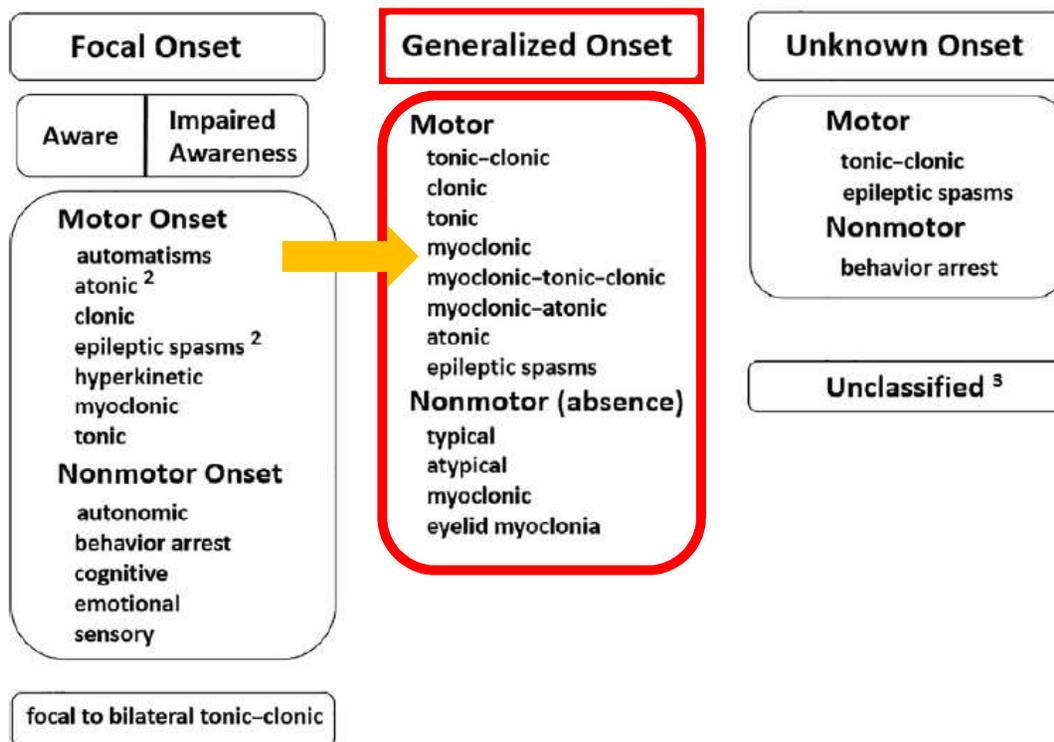
- Focal seizure
- Focal impaired awareness seizure
- Focal impaired awareness motor seizure
- Focal impaired awareness clonic seizure
(Focal impaired awareness right arm clonic seizure)
- Focal motor onset clonic seizure
- Focal clonic seizure

Prominent symptoms: **motor** vs non motor onset

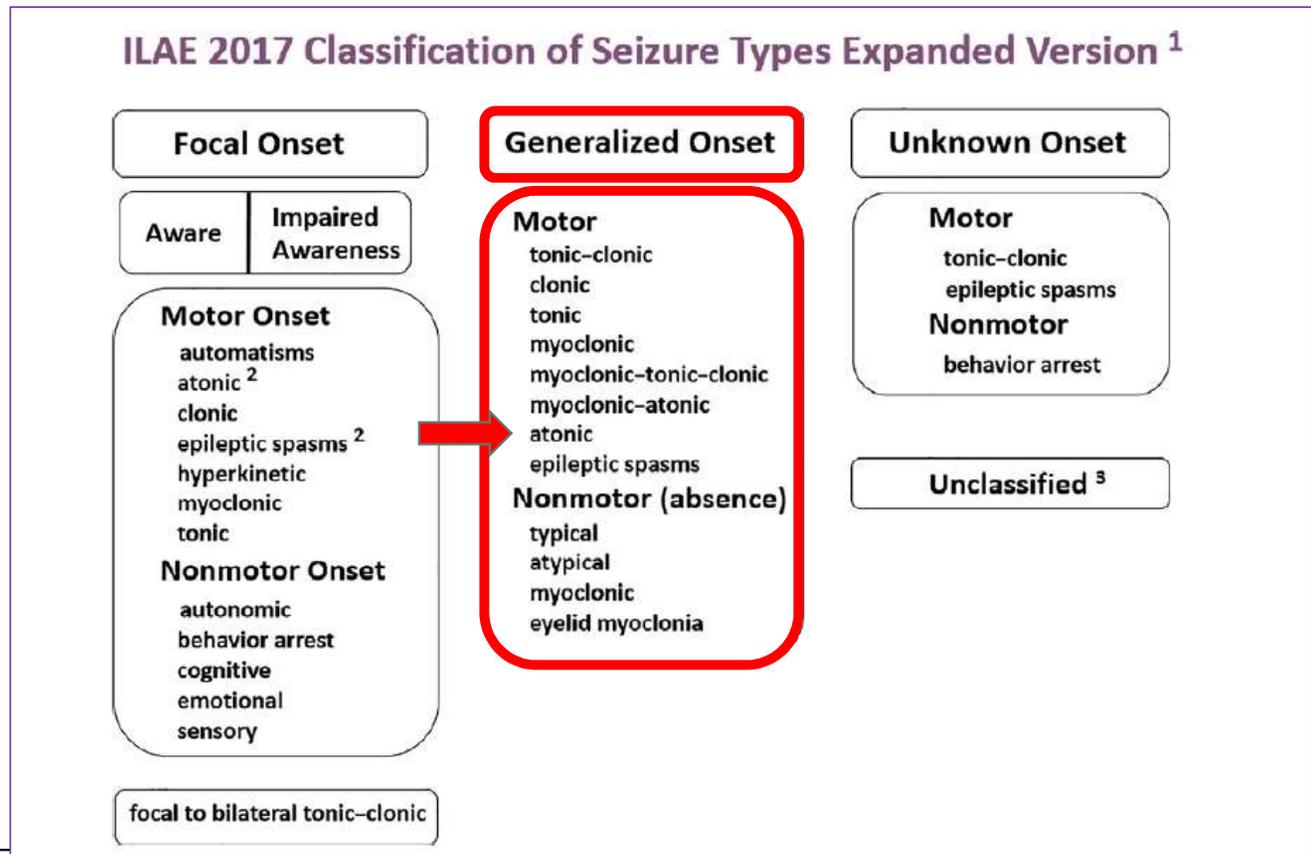
Epilepsy.org



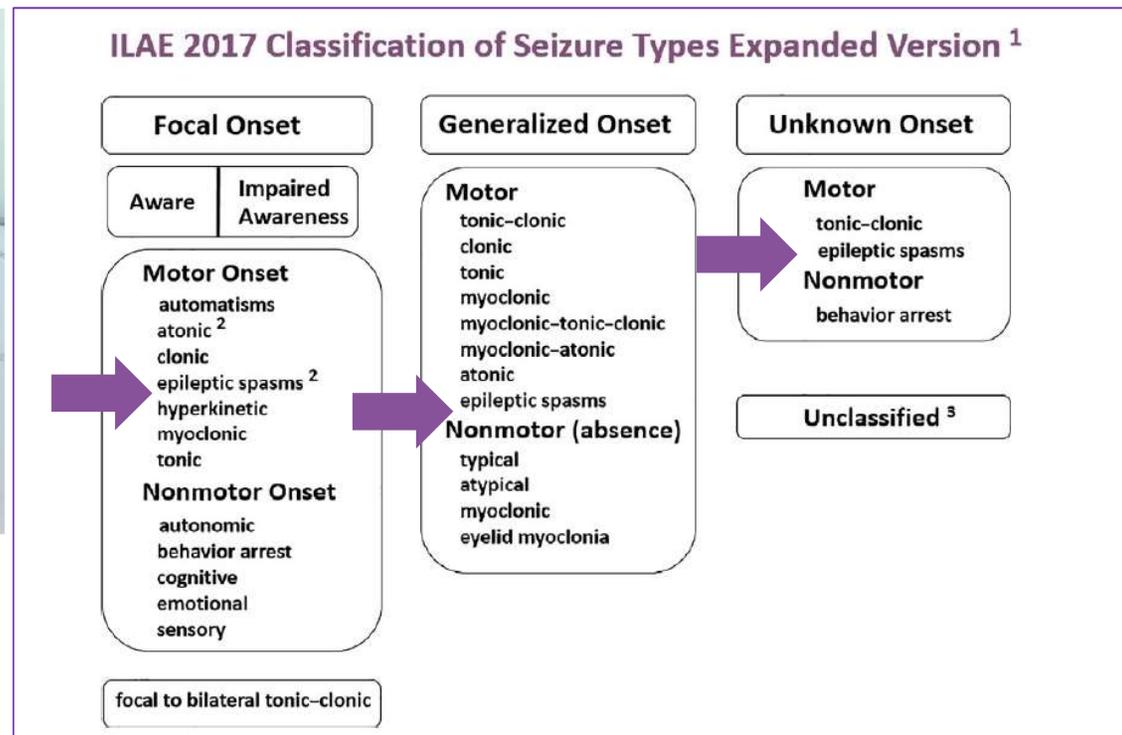
ILAE 2017 Classification of Seizure Types Expanded Version ¹



Prominent symptoms: **motor** vs non motor onset

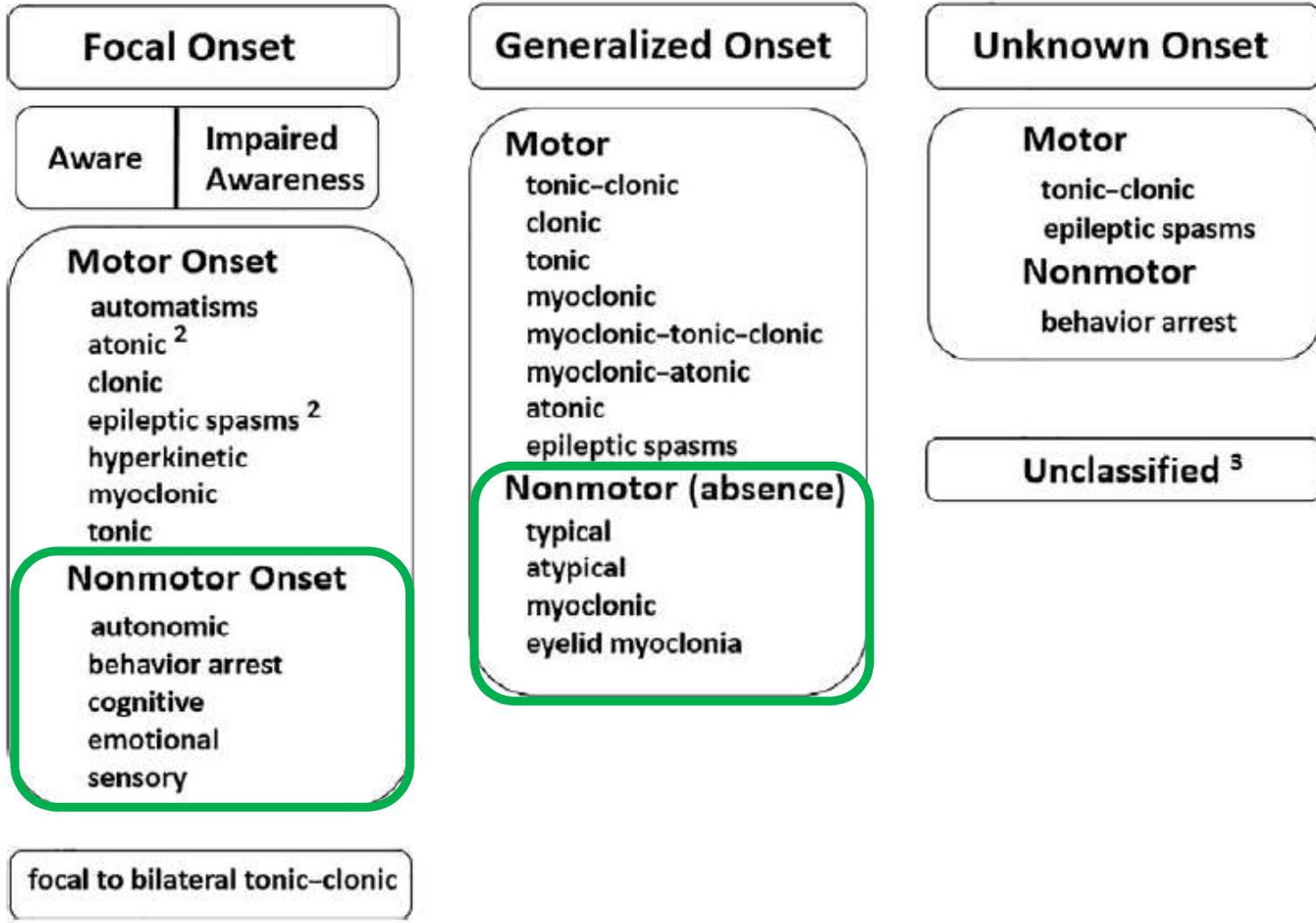


Prominent symptoms: motor vs non motor onset



EEG will help clarify focal or generalized onset of epileptic spasms ²⁸

ILAE 2017 Classification of Seizure Types Expanded Version ¹



Focal **non-motor** seizure

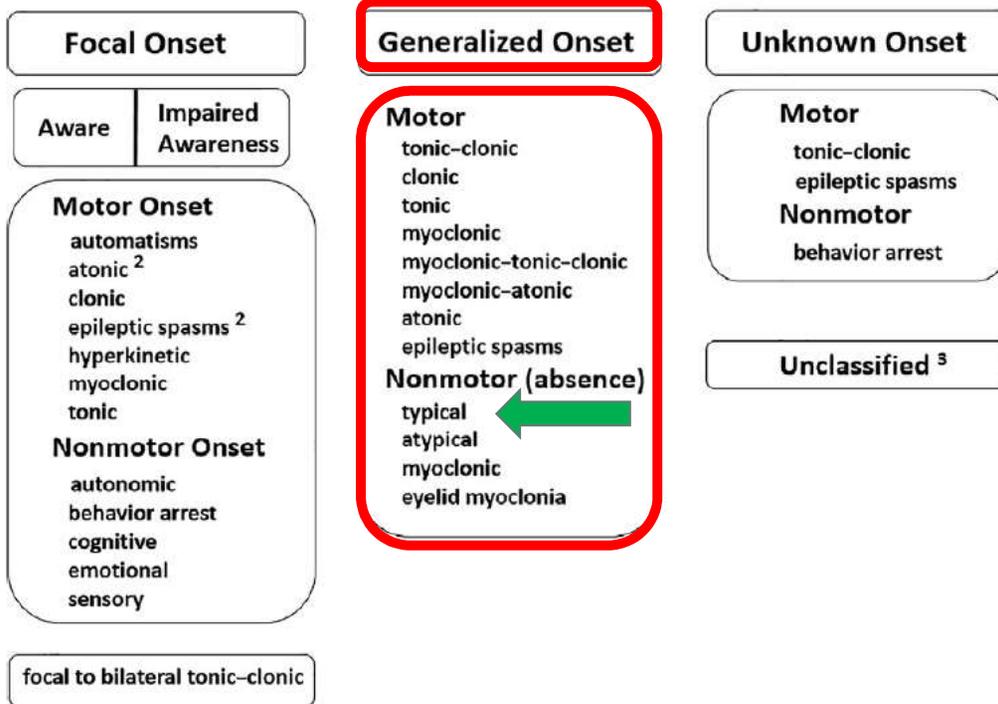
(Hx taking+ VDO + EEG+ Imaging)

Autonomic	Behavior arrest	Cognitive	Emotional	Sensory
<ul style="list-style-type: none"> : Asystole : Bradycardia : Tachycardia : Flushing : Nausea : Vomiting : Palpitation : Piloerection : etc 		<ul style="list-style-type: none"> : Acalculia : Aphasia : Attention impairment : Déjà vu : Dysphasia : Hallucination : Memory impairment : etc 	<ul style="list-style-type: none"> : Agitation : Anger : Anxiety : Fear : Laughing : etc 	<ul style="list-style-type: none"> : Auditory : Gustatory : Hot-cold sensation : Olfactory : Somatosensory : Vestibular : Visual : etc

What would you describe this video?

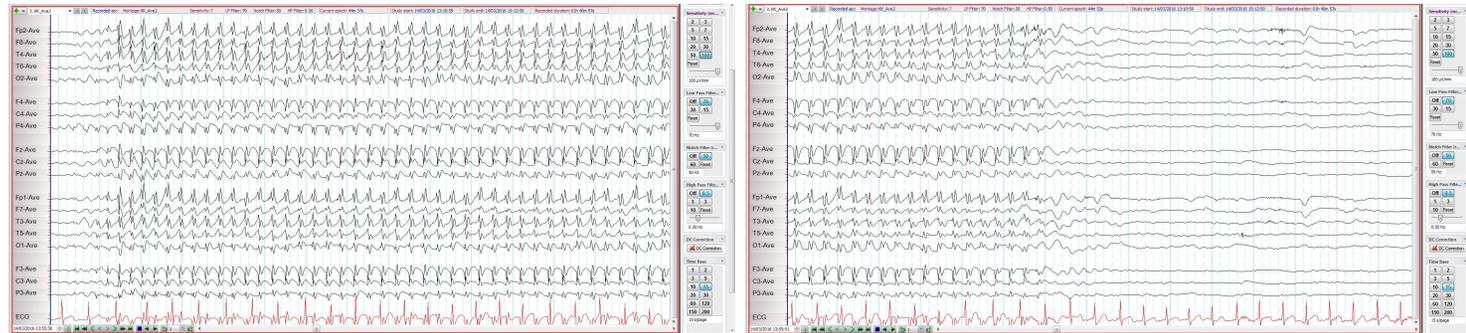


ILAE 2017 Classification of Seizure Types Expanded Version ¹



Generalized **non-motor** seizure (absence)

Do you need EEG ?



Gen non-motor **atypical absence** : need Hx + EEG



Unknown onset



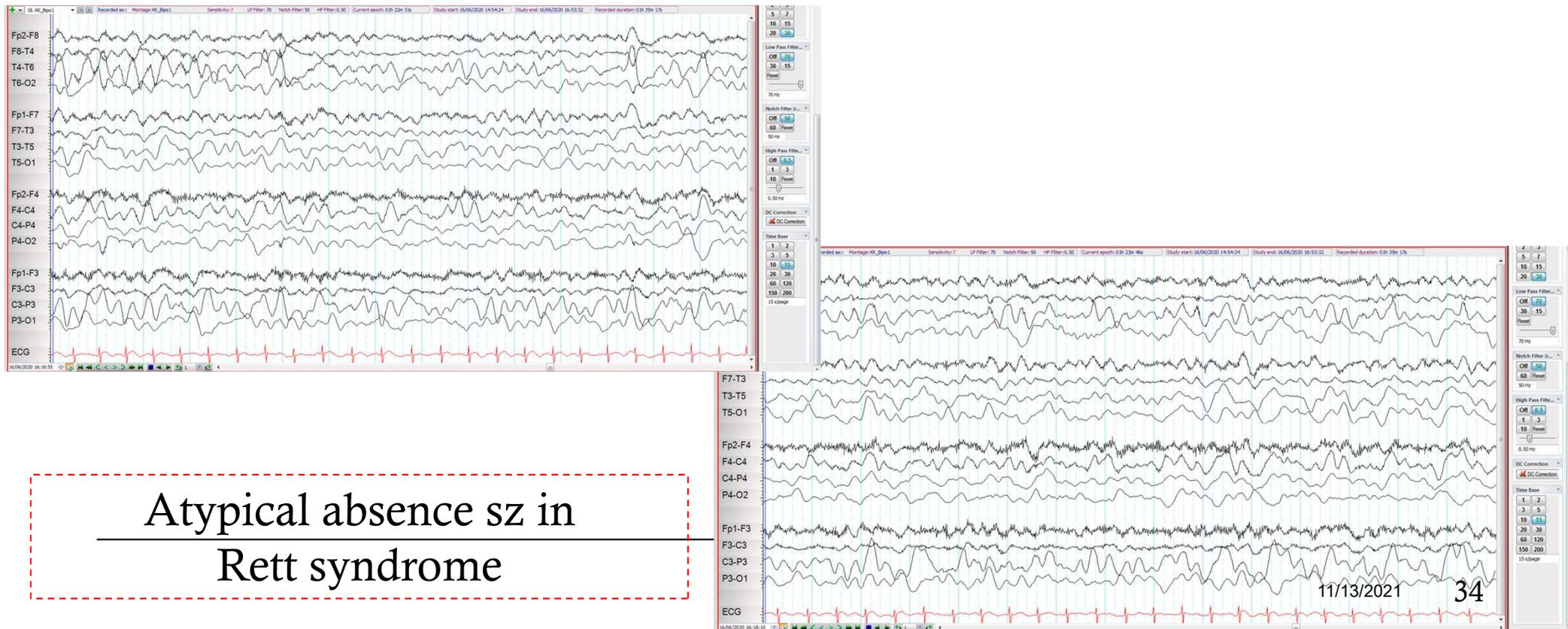
Stereotyped mvt



Gen non-motor sz

EEG during non-ictal period

Baseline EEG of atypical absence @ underlying epilepsy syndrome/clinical syndrome



Atypical absence sz in
Rett syndrome

From **previous** evaluation



ILAE 2017 Classification of Seizure Types Expanded Version ¹

Focal Onset

Aware

Impaired
Awareness

Motor Onset

automatisms
atonic ²
clonic
epileptic spasms ²
hyperkinetic
myoclonic
tonic

Nonmotor Onset

autonomic
behavior arrest
cognitive
emotional
sensory

focal to bilateral tonic-clonic

Generalized Onset

Motor

tonic-clonic
clonic
tonic
myoclonic
myoclonic-tonic-clonic
myoclonic-atonic
atonic
epileptic spasms

Nonmotor (absence)

typical
atypical
myoclonic
eyelid myoclonia

Unknown Onset

Motor

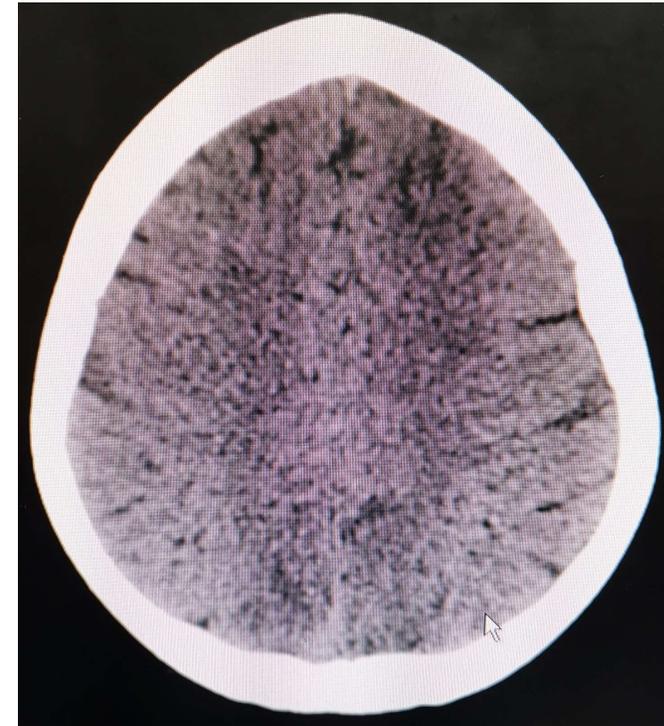
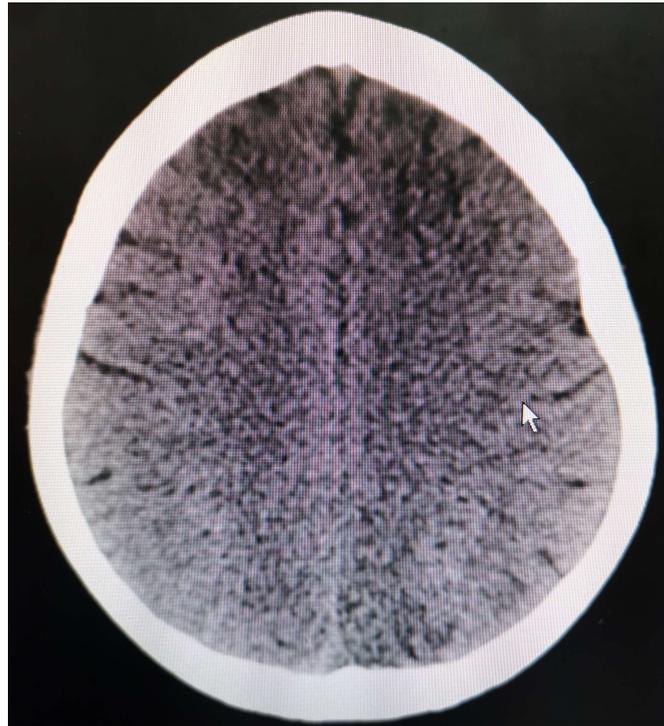
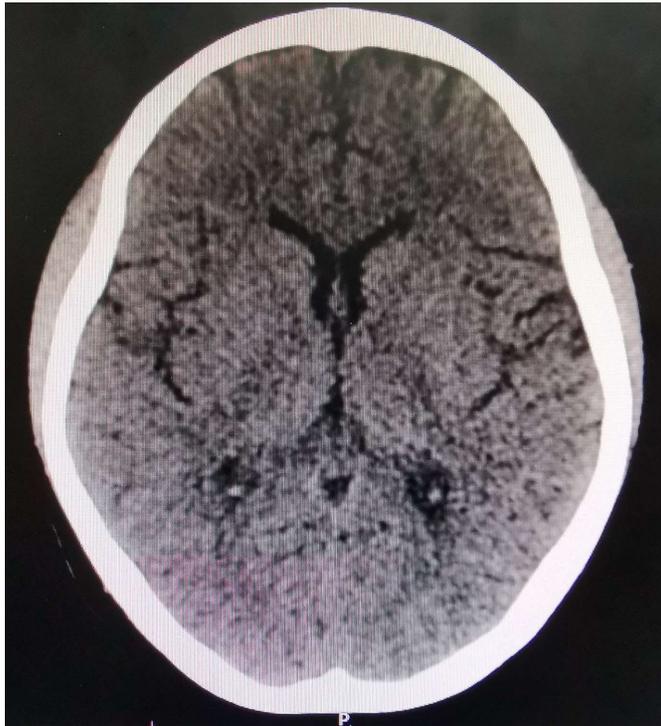
tonic-clonic
epileptic spasms

Nonmotor

behavior arrest

Unclassified ³

Imaging



From previous evaluation but now **changing to**



ILAE 2017 Classification of Seizure Types Expanded Version ¹

Focal Onset

Aware	Impaired Awareness
-------	--------------------

Motor Onset

- automatisms
- atonic ²
- clonic
- epileptic spasms ²
- hyperkinetic
- myoclonic
- tonic

Nonmotor Onset

- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic-clonic

Generalized Onset

Motor

- tonic-clonic
- clonic
- tonic
- myoclonic
- myoclonic-tonic-clonic
- myoclonic-atonic
- atonic
- epileptic spasms

Nonmotor (absence)

- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

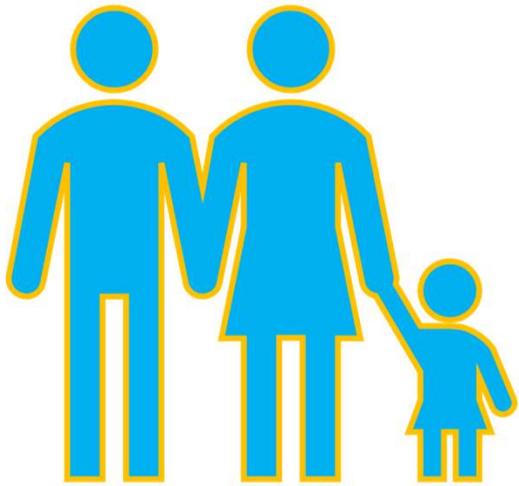
Motor

- tonic-clonic
- epileptic spasms

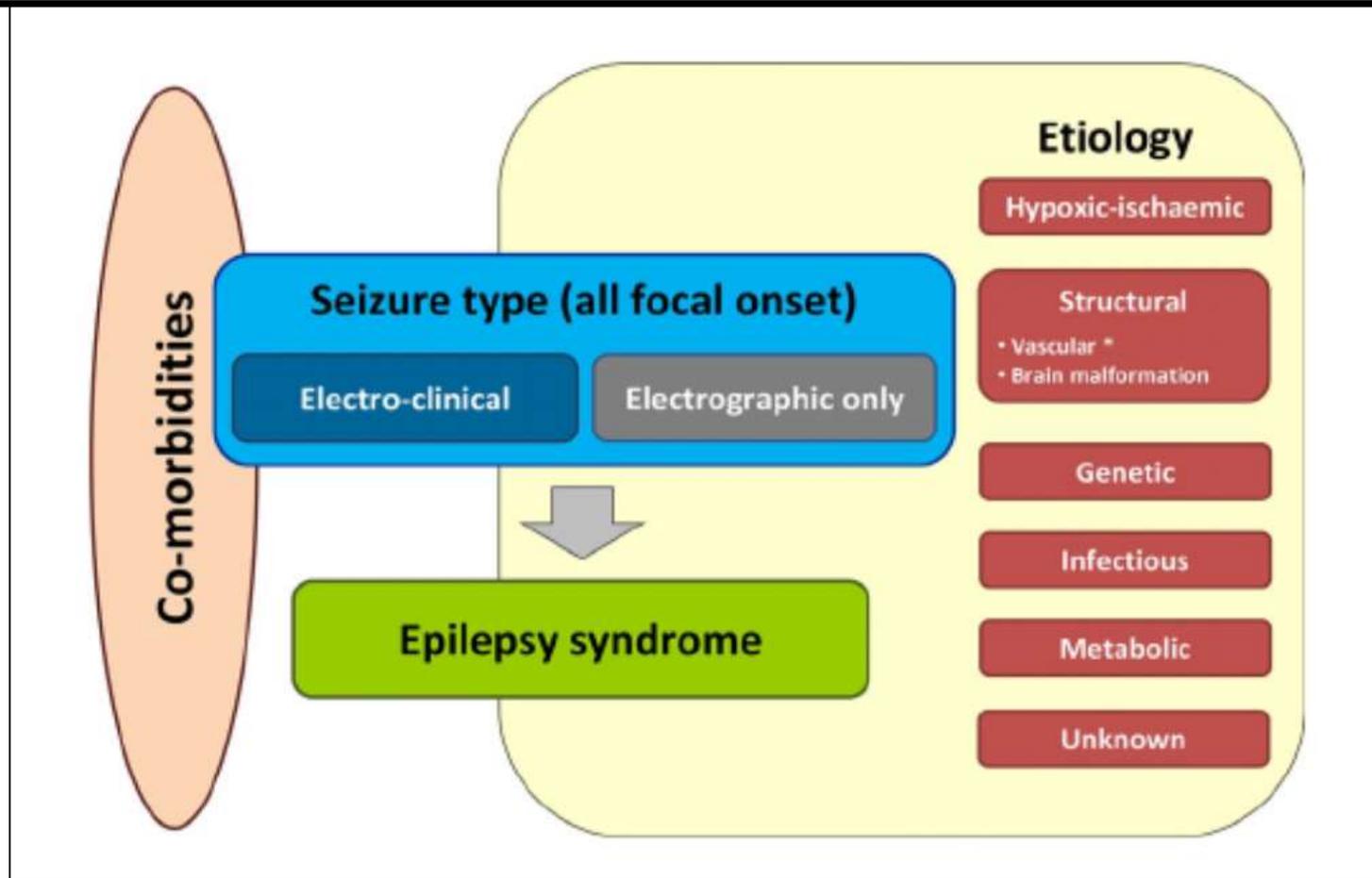
Nonmotor

- behavior arrest

Unclassified ³

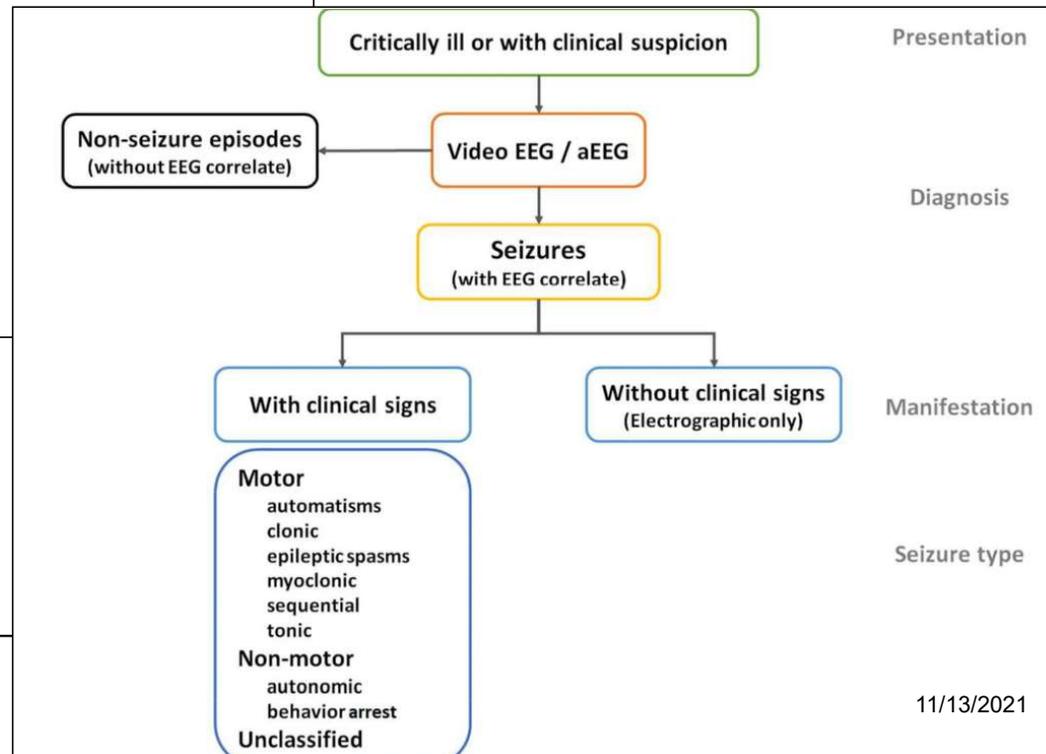
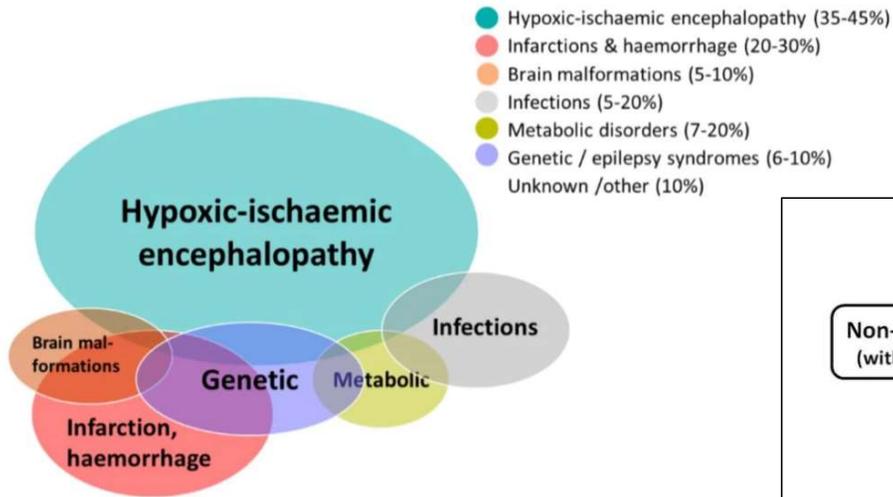


Framework for neonatal seizures and syndromes



Framework for neonatal seizures and syndromes

Etiology of Neonatal Seizures



2015

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



Eugen Trinka is professor and chairman of Department of Neurology, Paracelsus Medical University Salzburg Austria.

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

^aEvidence for the time frame is currently limited and future data may lead to modifications.

4 Axes: 1. Semiology 2. Etiology
3. EEG correlates 4. Age

Thaiepilepsysociety.com



สมาคมโรคลมชักแห่งประเทศไทย
Epilepsy Society of Thailand

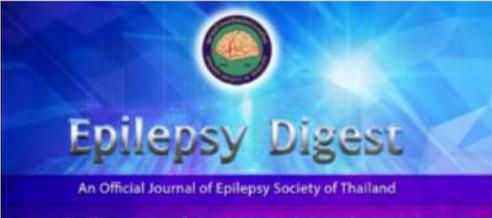


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Meta

11/13/2021