Definition and Classification of Epilepsies

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7 September 2019

Definition of seizure and epilepsy

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2005 and 2014

2005

Definition of seizure and epilepsy

- An epileptic **seizure** is defined conceptually as :

  a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain ✓

- **Epilepsy** *is a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures, and by the neurobiologic, cognitive, psychological, and social consequences of this condition. The definition of epilepsy requires the occurrence of at least one epileptic seizure.* ✗

Epilepsia, 46(4):470–472, 2005

2014

ILAE OFFICIAL REPORT

A practical clinical definition of epilepsy

*Robert S. Fisher, †Carlos Acevedo, ‡Alexis Arzimanoglou, §Alicia Bogacz, ¶J. Helen Cross, #Christian E. Elger, **Jerome Engel Jr, ††Lars Forsgren, ‡‡Jacqueline A. French, §§Mike Glynn, ¶¶Dale C. Hesdorffer, ##B.I. Lee, ***Gary W. Mathern, †††Solomon L. Moshé, ‡‡‡Emilio Perucca, §§§Ingrid E. Scheffer, ¶¶¶Torbjörn Tomson, ###Masako Watanabe, and ****Samuel Wiebe

Epilepsia, 55(4):475–482, 2014
doi: 10.1111/epi.12550

Epilepsy is a disease of the brain defined by any of the following conditions

1. At **least two** unprovoked (or reflex) seizures occurring > 24 h 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

Classification of epilepsies

ILAE Classifications of Epilepsy and Seizures

- 1964: Gastaut-Proposed international classification-seizures
- 1969: Gastaut- Proposals - seizures & epilepsies
- 1970: Gastaut- Classification- seizures
- 1970 : Merlis – Classification – epilepsies
- 1981: Commission – Classification – seizures
- 1985: Commission – Classification – epilepsies
- 1989: Commission – Classification – epilepsies/ syndrome
- 1993: Commission – epidermiological standards
- 2001: Blume – Glossary of ictal semiology
- 2001: Engel- Proposed diagnostic scheme/Axis 1-5
- 2005: Fisher- Definition of seizure and epilepsy
- 2006: Task Force: Report- seizures and epilepsies
- 2010: Commission: Revised terminology and concepts for organization of seizures and epilepsies…. Debate until 2013
- 2017: Classification of the epilepsies and operational classification of seizure type

ILAE 1981

Clinical seizure type | EEG sz type | EEG interictal expression

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 2^0 gen sz
- SPS → GTC
- CPS → GTC
- SPS → CPS → GTC

2. Generalized sz (convulsive and non-convulsive)

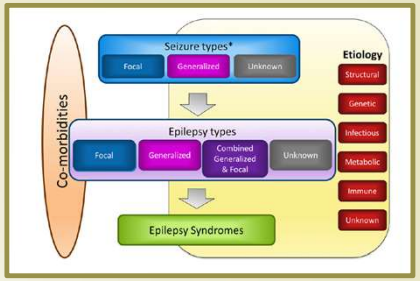
Absence, Myoclonic, Clonic, Tonic, Tonic-clonic, Atonic

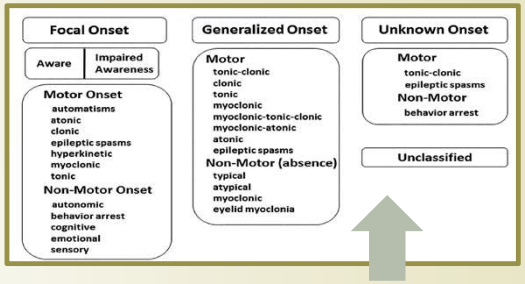
3. Unclassified epileptic sz

4. Prolonged or repetitive seizure (status epilepticus)

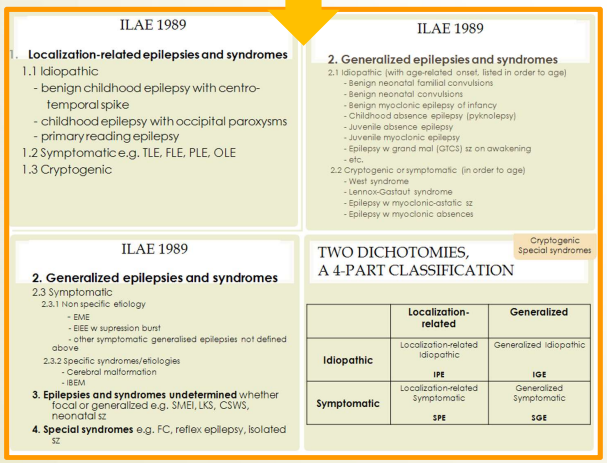
A 5 axes diagnostic scheme

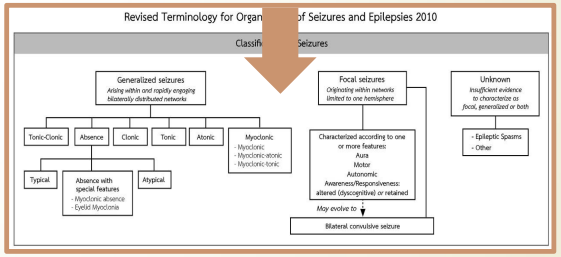
1. Ictal phenomenology
2. Seizure type
3. Epilepsy syndrome
4. Etiology
5. Impairment

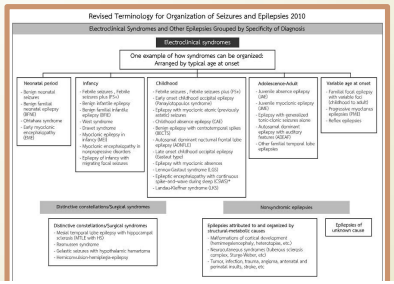




1981 → **1989** → **2001** → **2010-13** → **2017**







1981

ILAE 1981

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1989

ILAE 1989

1. **Localization-related epilepsies and syndromes**
   - 1.1 Idiopathic
     - benign childhood epilepsy 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
  - Benign myoclonic epilepsy of infancy
  - Childhood absence epilepsy (pyknolepsy)
  - Juvenile absence epilepsy
  - Juvenile myoclonic epilepsy
  - Epilepsy w grand mal (GTCS) sz on awakening
  - etc.
- 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
    - EIEE w supression burst
    - other symptomatic generalised epilepsies not defined above
  - 2.3.2 Specific syndromes/etiologies
    - Cerebral malformation
    - IBEM
- **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

Cryptogenic
Special syndromes

TWO DICHOTOMIES, A 4-PART CLASSIFICATION

	Localization-related	**Generalized**
Idiopathic	Localization-related Idiopathic **IPE**	Generalized Idiopathic **IGE**
Symptomatic	Localization-related Symptomatic **SPE**	Generalized Symptomatic **SGE**

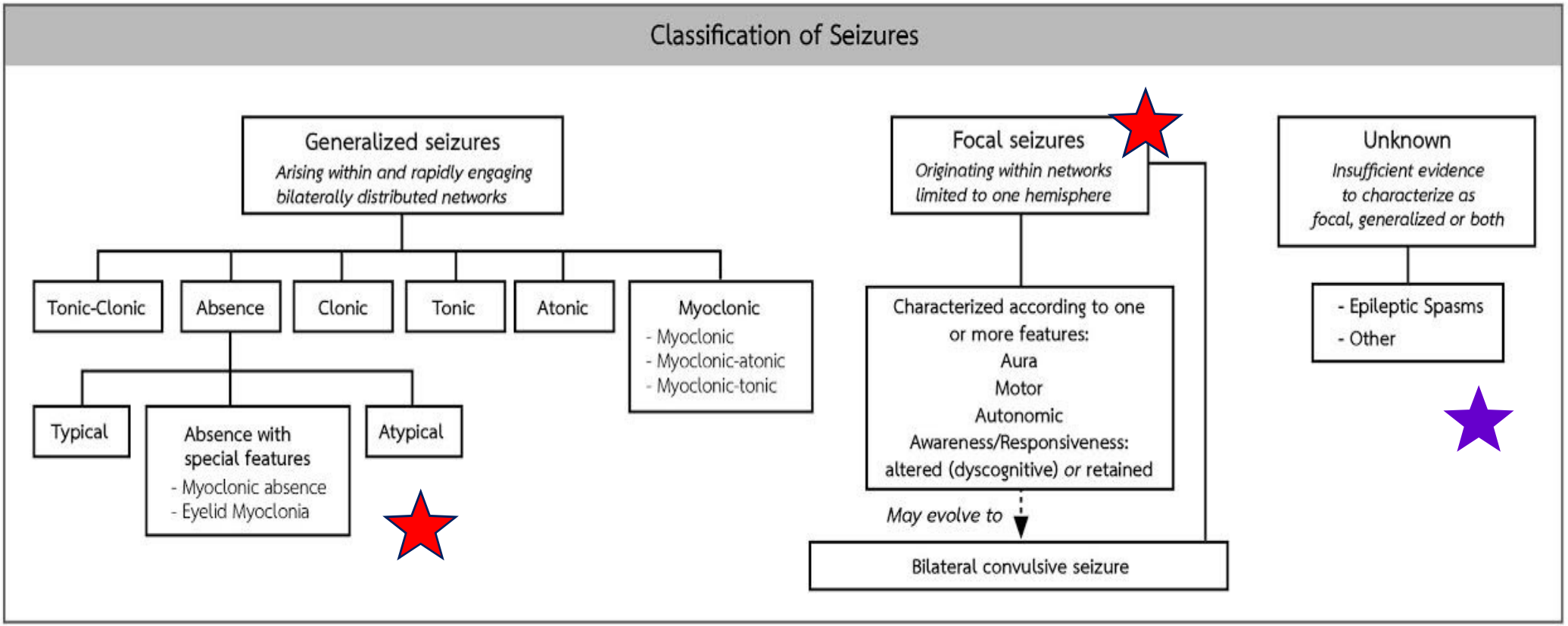
2001

A proposed diagnostic scheme for people with epileptic seizures and with epilepsy 2001	
Axis 1	Ictal phenomenology
Axis 2	Seizure type (1981+1989)
Axis 3	Syndrome (1989 + new synd)
Axis 4	Etiology
Axis 5	Impairment

Axis 1: Ictal phenomenology, from the Glossary of Descriptive Ictal Terminology, can be used to describe ictal events with any degree of detail needed.
Axis 2: Seizure type, from the List of Epileptic Seizures. Localization within the brain and precipitating stimuli for reflex seizures should be specified when appropriate.
Axis 3: Syndrome, from the List of Epilepsy Syndromes, with the understanding that a syndromic diagnosis may not always be possible.
Axis 4: Etiology, from a Classification of Diseases Frequently Associated with Epileptic Seizures or Epilepsy Syndromes when possible, genetic defects, or specific pathologic substrates for symptomatic focal epilepsies.
Axis 5: Impairment, this optional, but often useful, additional diagnostic parameter can be derived from an impairment classification adapted from the WHO ICIDH-2.

2010

Revised Terminology for Organization of Seizures and Epilepsies 2010



2010

2010

Partial sz
- Simple
- Complex

Focal sz

Describe @ features

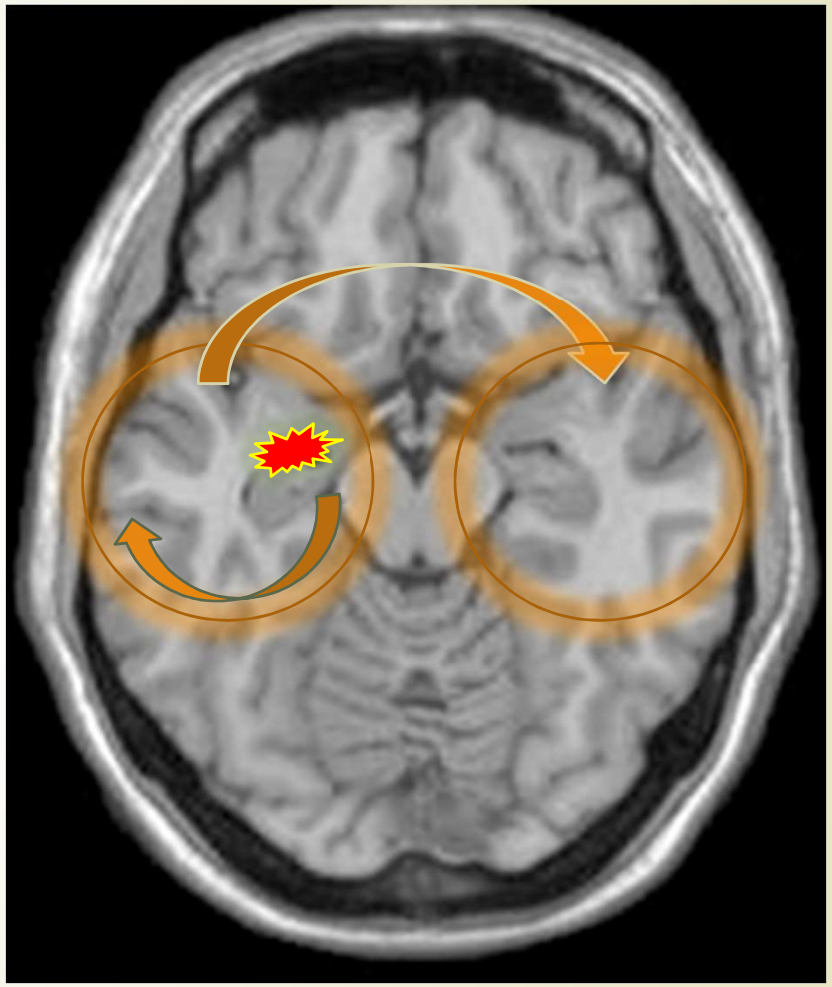
Generalized sz

Generalized sz
(add more subtypes)

2010

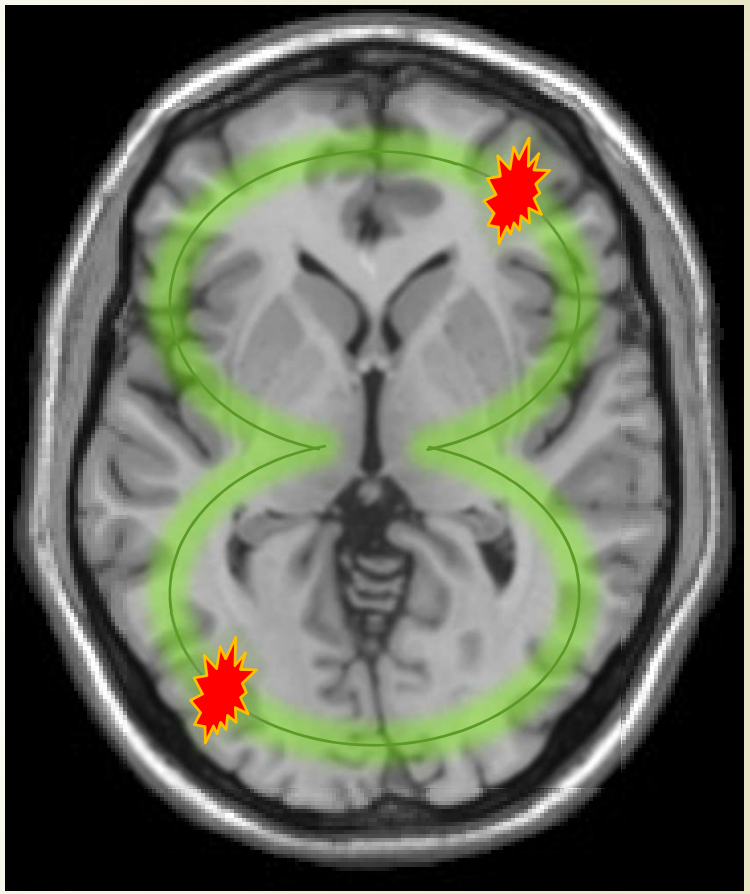
Focal seizures

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

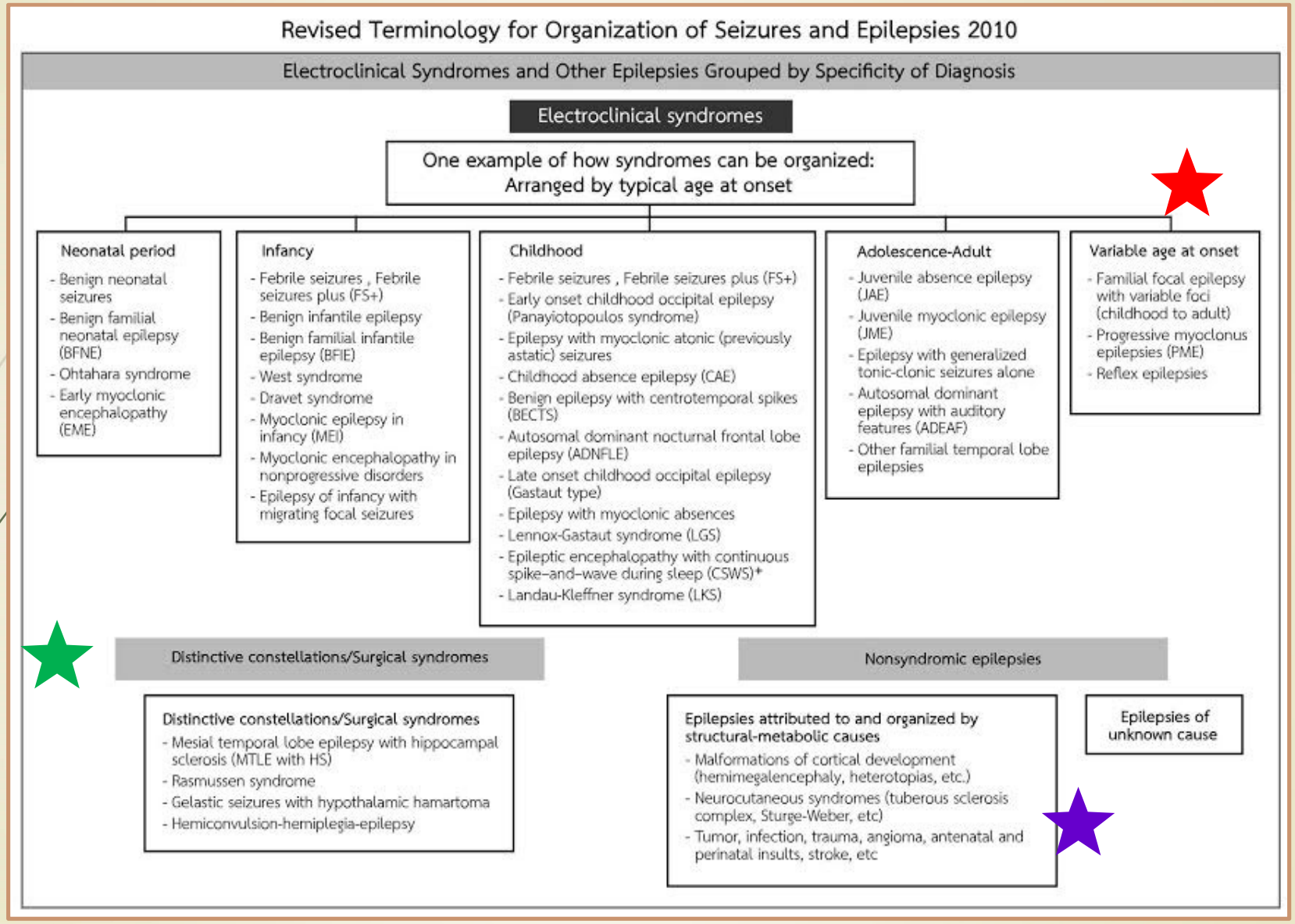


Generalized seizures

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



2010


Revised Terminology for Organization of Seizures and Epilepsies 2010
Electroclinical Syndromes and Other Epilepsies Grouped by Specificity of Diagnosis
Electroclinical syndromes
One example of how syndromes can be organized:
Arranged by typical age at onset
Neonatal period
- Benign neonatal seizures
- Benign familial neonatal epilepsy (BFNE)
- Ohtahara syndrome
- Early myoclonic encephalopathy (EME)
Infancy
- Febrile seizures , Febrile seizures plus (FS+)
- Benign infantile epilepsy
- Benign familial infantile epilepsy (BFIE)
- West syndrome
- Dravet syndrome
- Myoclonic epilepsy in infancy (MEI)
- Myoclonic encephalopathy in nonprogressive disorders
- Epilepsy of infancy with migrating focal seizures
Childhood
- Febrile seizures , Febrile seizures plus (FS+)
- Early onset childhood occipital epilepsy (Panayiotopoulos syndrome)
- Epilepsy with myoclonic atonic (previously astatic) seizures
- Childhood absence epilepsy (CAE)
- Benign epilepsy with centrotemporal spikes (BECTS)
- Autosomal dominant nocturnal frontal lobe epilepsy (ADNFLE)
- Late onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome (LGS)
- Epileptic encephalopathy with continuous spike–and–wave during sleep (CSWS)+
- Landau-Kleffner syndrome (LKS)
Adolescence-Adult
- Juvenile absence epilepsy (JAE)
- Juvenile myoclonic epilepsy (JME)
- Epilepsy with generalized tonic-clonic seizures alone
- Autosomal dominant epilepsy with auditory features (ADEAF)
- Other familial temporal lobe epilepsies
Variable age at onset
- Familial focal epilepsy with variable foci (childhood to adult)
- Progressive myoclonus epilepsies (PME)
- Reflex epilepsies
Distinctive constellations/Surgical syndromes
Nonsyndromic epilepsies
Distinctive constellations/Surgical syndromes
- Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
- Rasmussen syndrome
- Gelastic seizures with hypothalamic hamartoma
- Hemiconvulsion-hemiplegia-epilepsy
Epilepsies attributed to and organized by structural-metabolic causes
- Malformations of cortical development (hemimegalencephaly, heterotopias, etc.)
- Neurocutaneous syndromes (tuberous sclerosis complex, Sturge-Weber, etc)
- Tumor, infection, trauma, angioma, antenatal and perinatal insults, stroke, etc
Epilepsies of unknown cause

The year 2017…what happened ?

Motivation for revision of 2017

- Some seizure types can have either focal or generalized onset e.g. epileptic spasms, tonic sz, atonic sz, clonic sz, myoclonic sz
- Some important seizures types are not included
- Some terms do not have high level of acceptance or public understanding e.g. partial, psychic , SPS, CPS, dyscognitive(2010)

Fisher et al. Instruction manual for the ILAE, Epilepsia 2017

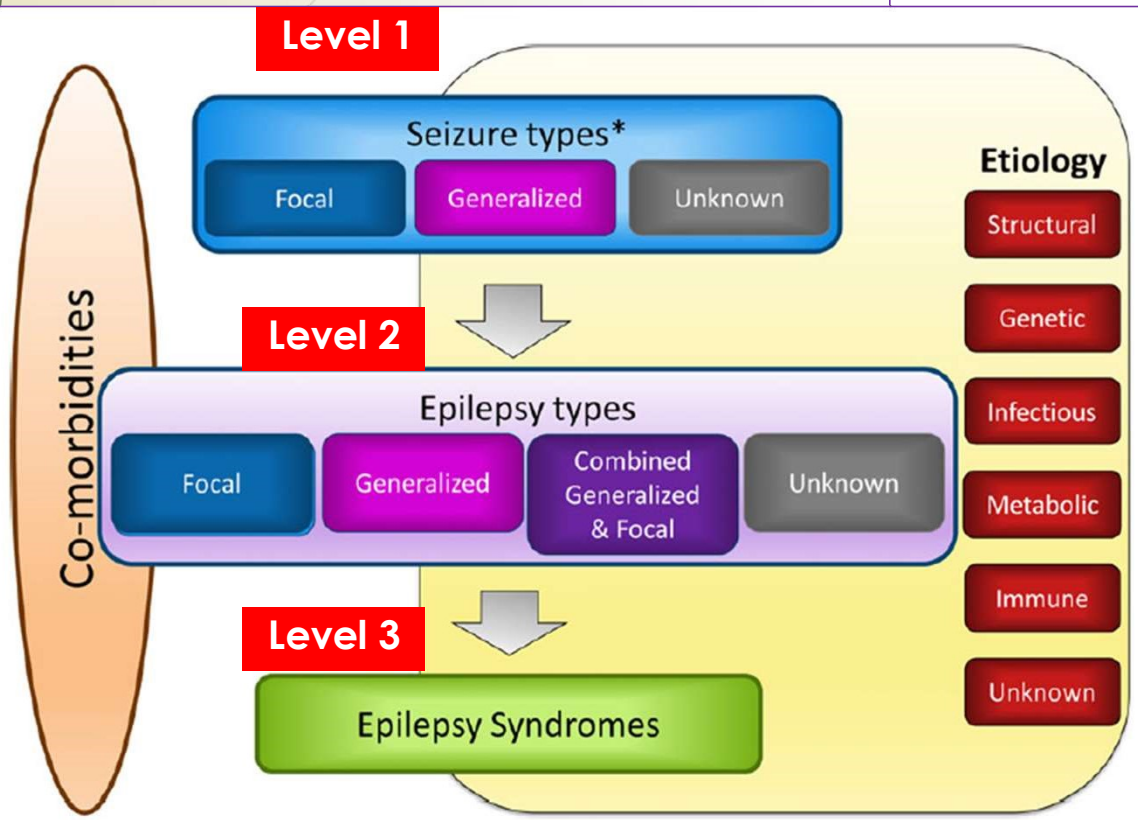
2017

ILAE POSITION PAPER

ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology

[1,2,3]Ingrid E. Scheffer, [1]Samuel Berkovic, [4]Giuseppe Capovilla, [5]Mary B. Connolly, [6]Jacqueline French, [7]Laura Guilhoto, [8,9]Edouard Hirsch, [10]Satish Jain, [11]Gary W. Mathern, [12]Solomon L. Moshé, [13]Douglas R. Nordli, [14]Emilio Perucca, [15]Torbjörn Tomson, [16]Samuel Wiebe, [17]Yue-Hua Zhang, and [18,19]Sameer M. Zuberi

Epilepsia, **(*):1–10, 2017
doi: 10.1111/epi.13709



2017

ILAE COMMISSION REPORT

Instruction manual for the ILAE 2017 operational classification of seizure types

[1]Robert S. Fisher, [2]J. Helen Cross, [3]Carol D'Souza, [4]Jacqueline A. French, [5]Sheryl R. Haut, [6]Norimichi Higurashi, [7]Edouard Hirsch, [8]Floor E. Jansen, [9]Lieven Lagae, [10]Solomon L. Moshé, [11]Jukka Peltola, [12]Eliane Roulet Perez, [13]Ingrid E. Scheffer, [14]Andreas Schulze-Bonhage, [15]Ernest Somerville, [16]Michael Sperling, [17]Elza Márcia Yacubian, and [18,19]Sameer M. Zuberi on behalf of the ILAE Commission for Classification and Terminology

Epilepsia, **(*):1–12, 2017
doi: 10.1111/epi.13671

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]

ILAE 2017 Classification of Seizure Types Basic Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
Nonmotor Onset

focal to bilateral tonic–clonic

Generalized Onset

Motor
- Tonic–clonic
- Other motor

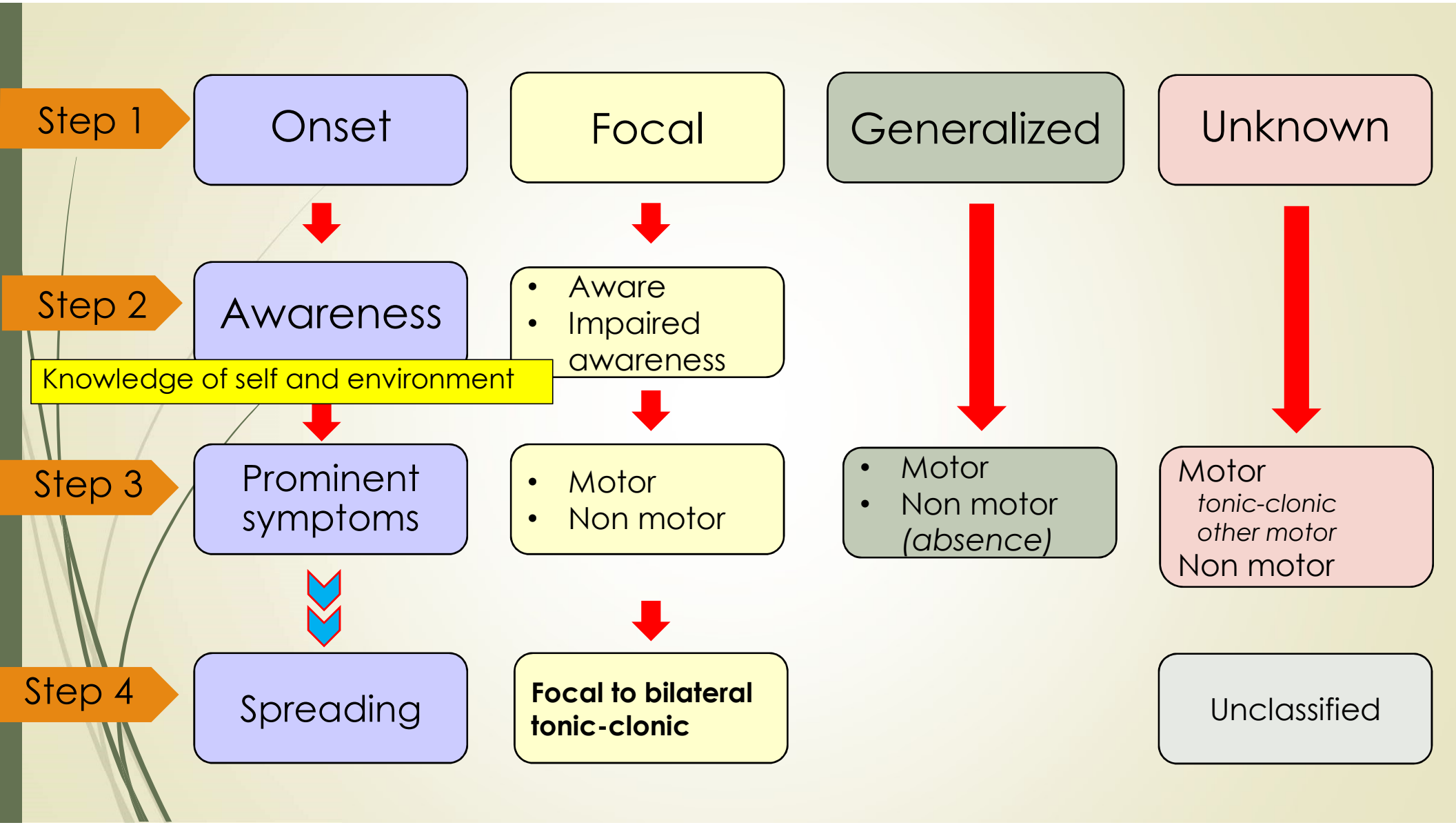
Nonmotor (Absence)

Unknown Onset

Motor
- Tonic–clonic
- Other motor

Nonmotor

Unclassified [2]


Step 1
Onset
Focal
Generalized
Unknown
Step 2
Awareness
Aware
Impaired awareness
Knowledge of self and environment
Step 3
Prominent symptoms
Motor
Non motor
Motor
Non motor (absence)
Motor
tonic-clonic
other motor
Non motor
Step 4
Spreading
Focal to bilateral tonic-clonic
Unclassified

80% level of certainty

Step 1 | Onset | Focal | Generalized | Unknown

Step 2 | Awareness

Knowledge of self or environment

Ongoing of activities

Step 3

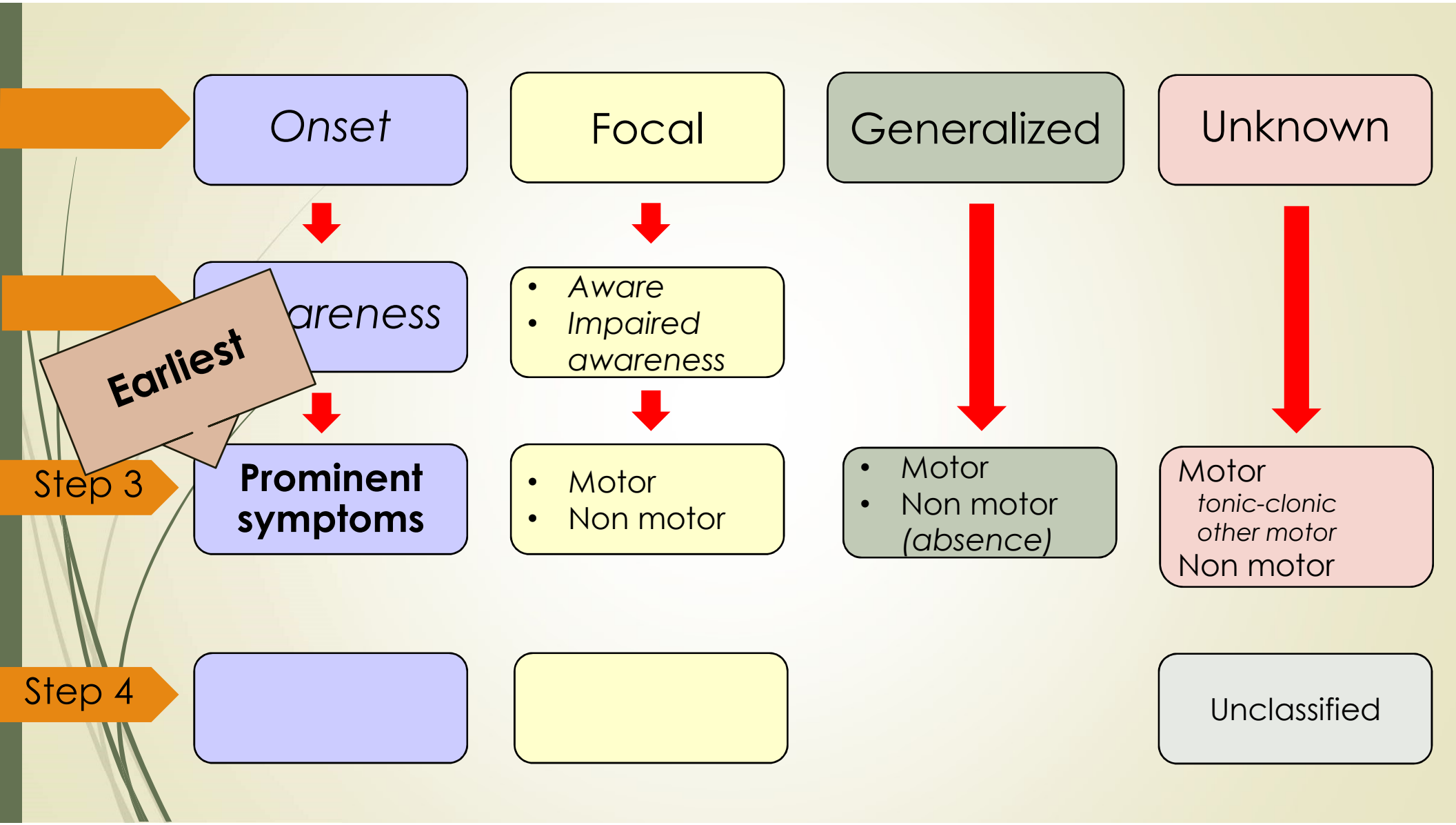
Step 4

Step 1. Onset: semiology

Step 2. Awareness: impaired awareness= (during <u>any segment</u> of the seizure)

Evaluation of **awareness** for **ictal** and not postictal

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


Onset
Focal
Generalized
Unknown
areness
Earliest
• Aware
• Impaired awareness
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• Motor
• Non motor
• Motor
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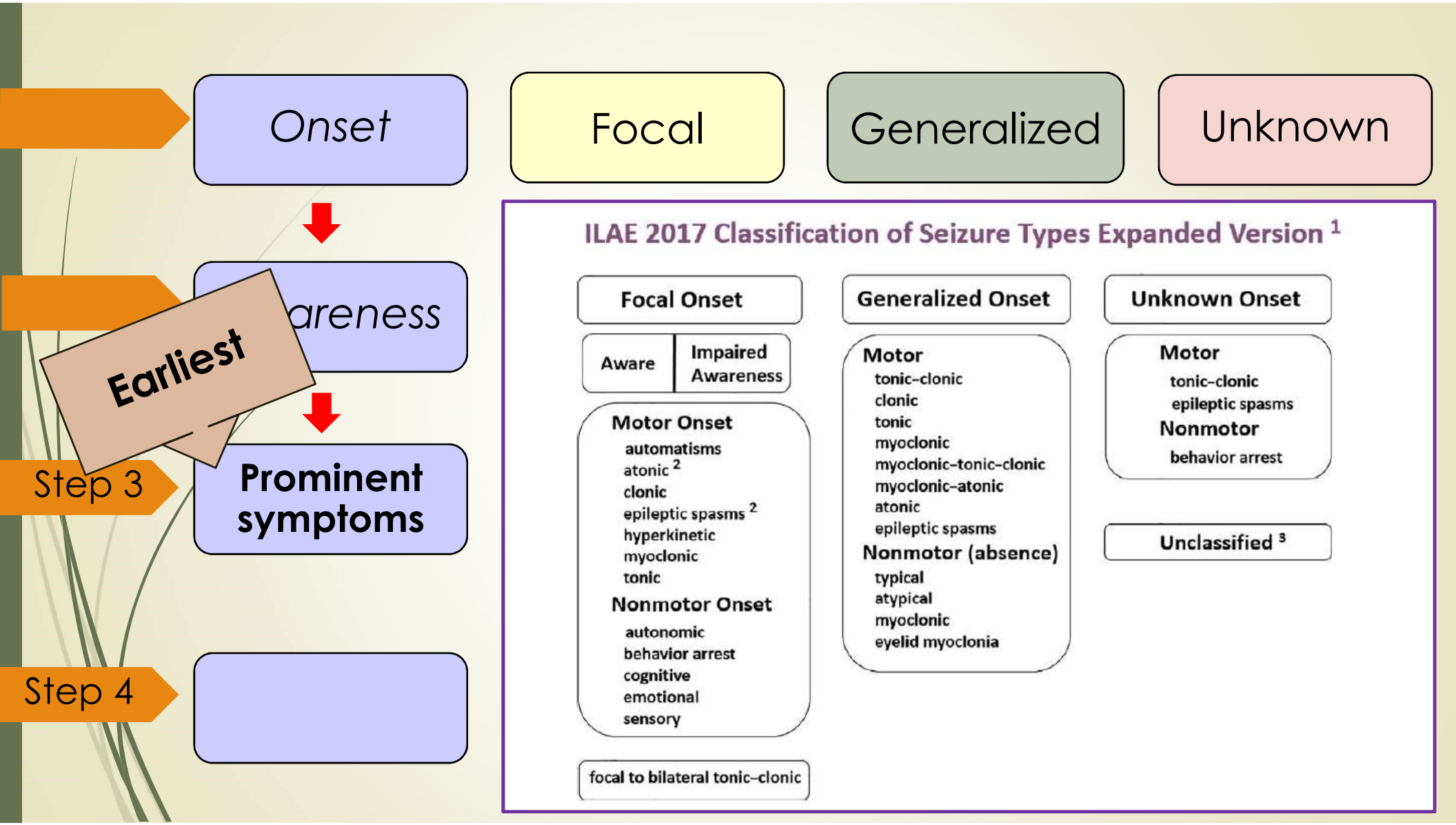
tonic-clonic
other motor
Non motor
Step 4
Unclassified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tonic
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Nonmotor (absence)
typical
atypical
myoclonic
eyelid myoclonia
Unknown Onset
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Nonmotor
behavior arrest
Unclassified [3]

Table 2. Glossary of terms

Word	Definition	Source
Absence, typical	A sudden onset, interruption of ongoing activities, a blank stare, possibly a brief upward deviation of the eyes. Usually the patient will be unresponsive when spoken to. Duration is a few seconds to half a minute with very rapid recovery. Although not always available, an EEG would show generalized epileptiform discharges during the event. An absence seizure is by definition a seizure of generalized onset. The word is not synonymous with a blank stare, which also can be encountered with focal onset seizures	Adapted from Ref. 12
Absence, atypical	An absence seizure with changes in tone that are more pronounced than in typical absence or the onset and/or cessation is not abrupt, often associated with slow, irregular, generalized spike-wave activity	Adapted from Ref. 11
Arrest	See behavior arrest	New
Atonic	Sudden loss or diminution of muscle tone without apparent preceding myoclonic or tonic event lasting ~1–2 s, involving head, trunk, jaw, or limb musculature	12
Automatism	A more or less coordinated motor activity usually occurring when cognition is impaired and for which the subject is usually (but not always) amnesic afterward. This often resembles a voluntary movement and may consist of an inappropriate continuation of preictal motor	12
	sustained as a tonic seizure. Limited forms may occur: Grimacing, head nodding, or subtle eye movements. Epileptic spasms frequently occur in clusters. Infantile spasms are the best known form, but spasms can occur at all ages	
Epilepsy	A disease of the brain defined by any of the following conditions: (1) At least two unprovoked (or reflex) seizures occurring >24 h 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. Epilepsy is considered to be resolved for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure free for the last 10 years, with no antiseizure medicines for the last 5 years	3
Eyelid myoclonia	Jerking of the eyelids at frequencies of at least 3 per second, commonly with upward eye deviation, usually lasting < 10 s, often precipitated by eye closure. There may or may not be associated brief loss of awareness	New
Fencer's posture seizure	A focal motor seizure type with extension of one arm and flexion at the contralateral elbow and wrist, giving an imitation of swordplay with a foil. This has also been called a supplementary motor area seizure	New
Figure-of-4 seizure	Upper limbs with extension of the arm (usually contralateral to the epileptogenic zone) with elbow flexion of the other arm, forming a figure-of-4	New
Focal	Originating within networks limited to one hemisphere. They may be discretely localized or more widely distributed. Focal seizures may originate in subcortical structures	5
Focal onset bilateral tonic–clonic seizure	A seizure type with focal onset, with awareness or impaired awareness, either motor or non-motor, progressing to bilateral tonic–clonic activity. The prior term was seizure with partial onset with secondary generalization	New

Table 2. Continued.

Word	Definition	Source
Gelastic	Bursts of laughter or giggling, usually without an appropriate affective tone	12
Generalized	Originating at some point within, and rapidly engaging, bilaterally distributed networks	5
Generalized tonic–clonic	Bilateral symmetric or sometimes asymmetric tonic contraction and then bilateral clonic contraction of somatic muscles, usually associated with autonomic phenomena and loss of awareness. These seizures engage networks in both hemispheres at the start of the seizure	Adapted from Refs 5, 12
Hallucination	A creation of composite perceptions without corresponding external stimuli involving visual, auditory, somatosensory, olfactory, and/or gustatory phenomena. Example: "Hearing" and "seeing" people talking	12
Behavior arrest	Arrest (pause) of activities, freezing, immobilization, as in behavior arrest seizure	New
Immobility	See activity arrest	New
Impaired awareness	See awareness. Impaired or lost awareness is a feature of focal impaired awareness seizures,	New
Seizure	A transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain	4
Sensory seizure	A perceptual experience not caused by appropriate stimuli in the external world	12
Spasm	See epileptic spasm	
Tonic	A sustained increase in muscle contraction lasting a few seconds to minutes	12
Tonic–clonic	A sequence consisting of a tonic followed by a clonic phase	12
Unaware	The term unaware can be used as shorthand for impaired awareness	New
Unclassified	Referring to a seizure type that cannot be described by the ILAE 2017 classification either because of inadequate information or unusual clinical features. If the seizure is unclassified because the type of onset is unknown, a limited classification may still derive from observed features	New
Unresponsive	Not able to react appropriately by movement or speech when presented with stimulation	New
Versive	A sustained, forced conjugate ocular, cephalic, and/or truncal rotation or lateral deviation from the midline	12

Automatism : A more or less coordinated motor activity usually occurring when cognition is impaired and for which the subject is usually (but not always) amnesic afterward. This often resembles a voluntary movement and may consists of an inappropriate continuation of preictal motor activity.

Epilepsia 2017

Common descriptors

Table 1. Common descriptors of behaviors during and after seizures (alphabetically)

Cognitive	Automatisms
Acalculia	Aggression
Aphasia	Eye-blinking
Attention impairment	Head-nodding
Déjà vu or jamais vu	Manual
Dissociation	Oral-facial
Dysphasia	Pedaling
Hallucinations	Pelvic thrusting
Illusions	Perseveration
Memory impairment	Running (cursive)
Neglect	Sexual
Forced thinking	Undressing
Responsiveness impairment	Vocalization/speech
	Walking
Emotional or affective	Motor
Agitation	Dysarthria
Anger	Dystonic
Anxiety	Fencer's posture (figure-of-4)
Crying (dacrystic)	Incoordination
Fear	Jacksonian
Laughing (gelastic)	Paralysis
Paranoia	Paresis
Pleasure	Versive
Autonomic	Sensory
Asystole	Auditory
Bradycardia	Gustatory
Erection	Hot-cold sensations
Flushing	Olfactory
Gastrointestinal	Somatosensory
Hyper/hypoventilation	Vestibular
Nausea or vomiting	Visual
Pallor	
Palpitations	Laterality
Piloerection	Left
Respiratory changes	Right
Tachycardia	Bilateral

Epilepsia 2017

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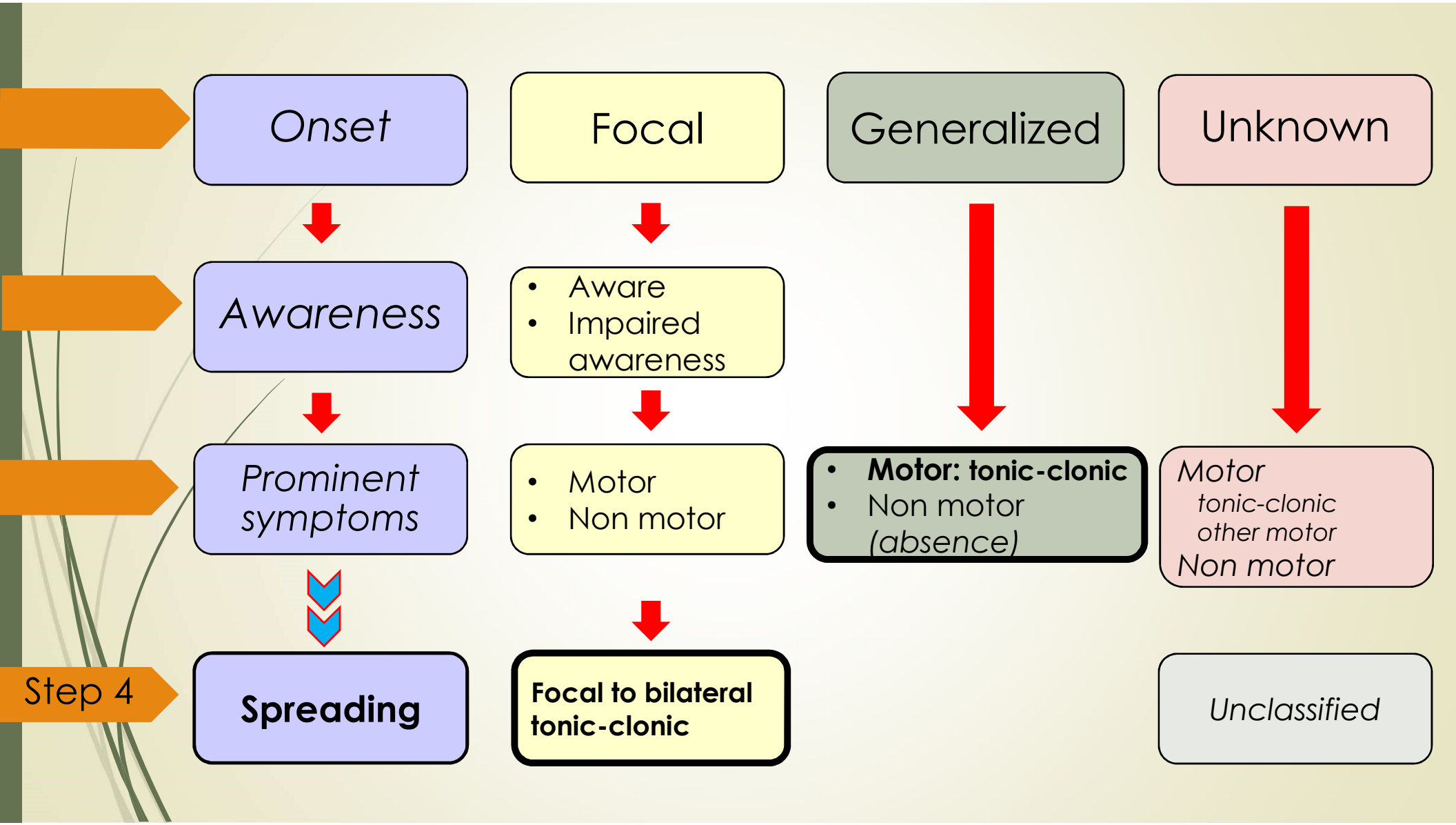
Automatism	A more or less coordinated motor activity usually occurring when cognition is impaired and for which the subject is usually (but not always) amnesic afterward. This often resembles a voluntary movement and may consist of an inappropriate continuation of preictal motor	12
Autonomic seizure		
Aura		
Awareness		
Bilateral		
Clonic		
Cognitive	memory, and praxis. The previous term for similar usage as a seizure type was psychic	
Consciousness	A state of mind with both subjective and objective aspects, comprising a sense of self as a unique entity, awareness, responsiveness, and memory	New
Dacrystic	Bursts of crying, which may or may not be associated with sadness	12
Dystonic	Sustained contractions of both agonist and antagonist muscles producing athetoid or twisting movements, which may produce abnormal postures	Adapted from Ref. 12
Emotional seizures	Seizures presenting with an emotion or the appearance of having an emotion as an early prominent feature, such as fear, spontaneous joy or euphoria, laughing (gelastic), or crying (dacrystic)	New
Epileptic spasms	A sudden flexion, extension, or mixed extension–flexion of predominantly proximal and truncal muscles that is usually more sustained than a myoclonic movement but not as sustained as a tonic seizure. Limited forms may occur: Grimacing, head nodding, or subtle eye movements. Epileptic spasms frequently occur in clusters. Infantile spasms are the best known form, but spasms can occur at all ages	Adapted from Ref. 12
Epilepsy	A disease of the brain defined by any of the following conditions: (1) At least two unprovoked (or reflex) seizures occurring >24 h 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. Epilepsy is considered to be resolved for individuals who had an age-dependent epilepsy syndrome but are now past the applicable age or those who have remained seizure free for the last 10 years, with no antiseizure medicines for the last 5 years	3
Eyelid myodonia	Jerking of the eyelids at frequencies of at least 3 per second, commonly with upward eye deviation, usually lasting < 10 s, often precipitated by eye closure. There may or may not be associated brief loss of awareness	New
Fencer's posture seizure	A focal motor seizure type with extension of one arm and flexion at the contralateral elbow and wrist, giving an imitation of swordplay with a foil. This has also been called a supplementary motor area seizure	New
Figure-of-4 seizure	Upper limbs with extension of the arm (usually contralateral to the epileptogenic zone) with elbow flexion of the other arm, forming a figure-of-4	New
Focal	Originating within networks limited to one hemisphere. They may be discretely localized or more widely distributed. Focal seizures may originate in subcortical structures	5
Focal onset bilateral tonic–clonic seizure	A seizure type with focal onset, with awareness or impaired awareness, either motor or non-motor, progressing to bilateral tonic–clonic activity. The prior term was seizure with partial onset with secondary generalization	New

Table 2. Continued.

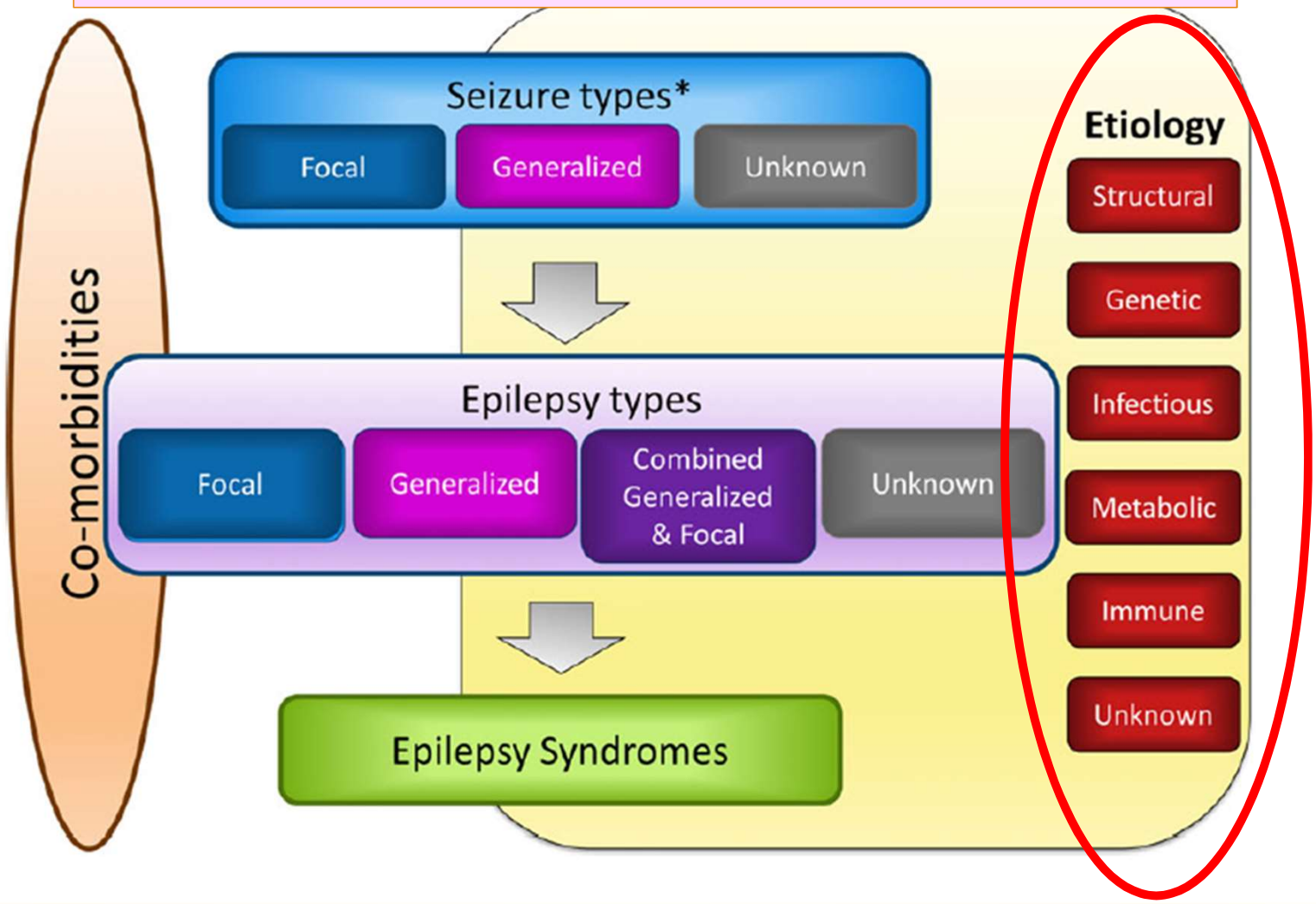
Word	Definition	Source
Gelastic	Bursts of laughter or giggling, usually without an appropriate affective tone	12
Generalized	Originating at some point within, and rapidly engaging, bilaterally distributed networks	5
Generalized tonic–clonic	Bilateral symmetric or sometimes asymmetric tonic contraction and then bilateral clonic contraction of somatic muscles, usually associated with autonomic phenomena and loss of awareness. These seizures engage networks in both hemispheres at the start of the seizure	Adapted from Refs 5, 12
Hallucination	A creation of composite perceptions without corresponding external stimuli involving visual, auditory, somatosensory, olfactory, and/or gustatory phenomena. Example: "Hearing" and "seeing" people talking	12
Behavior arrest	Arrest (pause) of activities, freezing, immobilization, as in behavior arrest seizure	New
Immobility	See activity arrest	New
Impaired awareness	See awareness. Impaired or lost awareness is a feature of focal impaired awareness seizures,	New
Myoclonic–atonic	A generalized seizure type with a myoclonic jerk leading to an atonic motor component. This type was previously called myoclonic–astatic	New
Myoclonic–tonic–clonic	One or a few jerks of limbs bilaterally, followed by a tonic–clonic seizure. The initial jerks can be considered to be either a brief period of clonus or myoclonus. Seizures with this characteristic are common in juvenile myoclonic epilepsy	Derived from Ref. 1
Nonmotor	Focal or generalized seizure types in which motor activity is not prominent	New
Propagation	Spread of seizure activity from one place in the brain to another, or engaging of additional brain networks	New
Responsiveness	Ability to appropriately react by movement or speech when presented with a stimulus	New
Seizure	A transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain	4
Sensory seizure	A perceptual experience not caused by appropriate stimuli in the external world	12
Spasm	See epileptic spasm	
Tonic	A sustained increase in muscle contraction lasting a few seconds to minutes	12
Tonic–clonic	A sequence consisting of a tonic followed by a clonic phase	12
Unaware	The term unaware can be used as shorthand for impaired awareness	New
Unclassified	Referring to a seizure type that cannot be described by the ILAE 2017 classification either because of inadequate information or unusual clinical features. If the seizure is unclassified because the type of onset is unknown, a limited classification may still derive from observed features	New
Unresponsive	Not able to react appropriately by movement or speech when presented with stimulation	New
Versive	A sustained, forced conjugate ocular, cephalic, and/or truncal rotation or lateral deviation from the midline	12

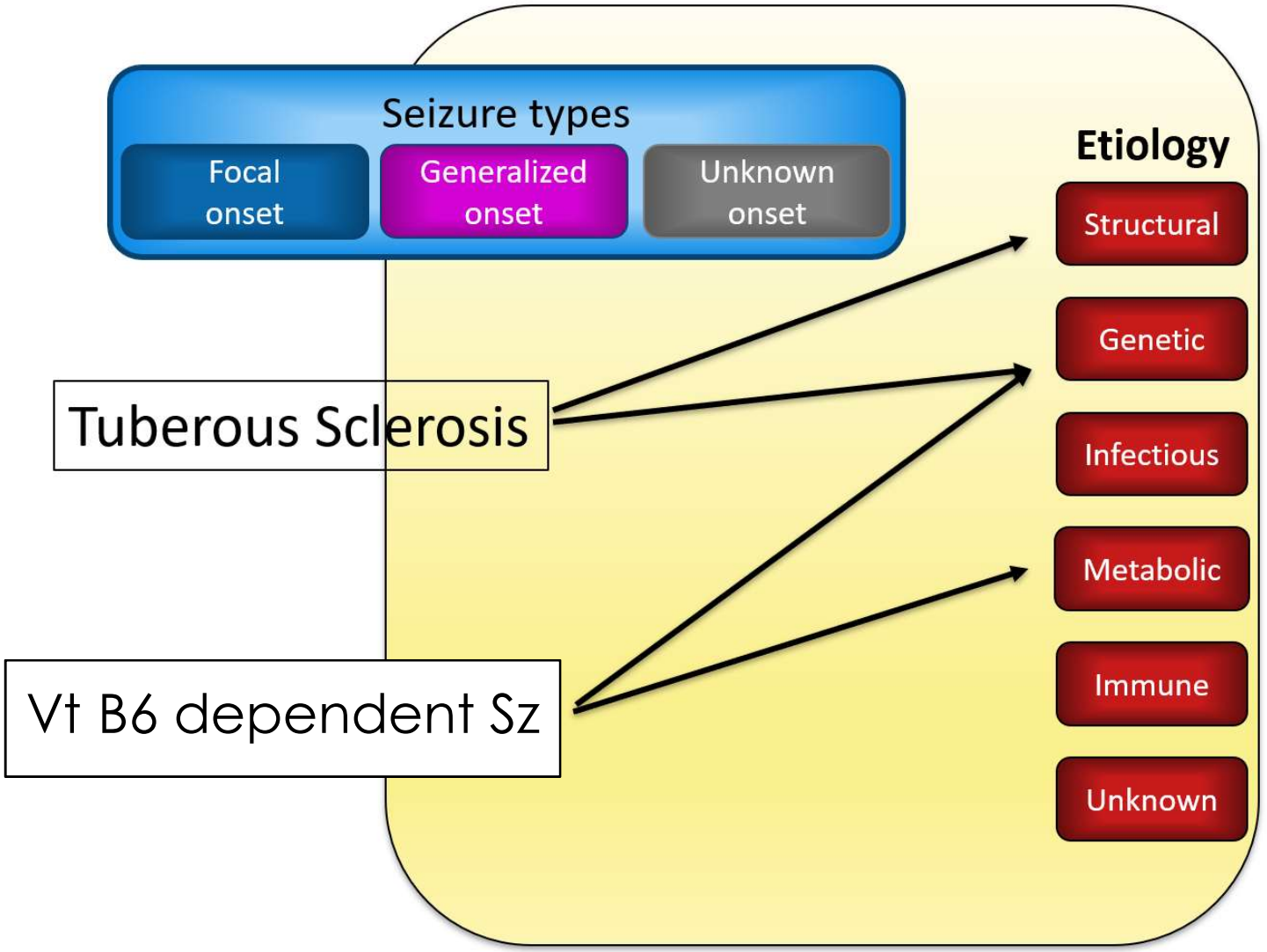
Behavior arrest : Arrest (pause) of activity, freezing, immobilization, as in behavior arrest seizure

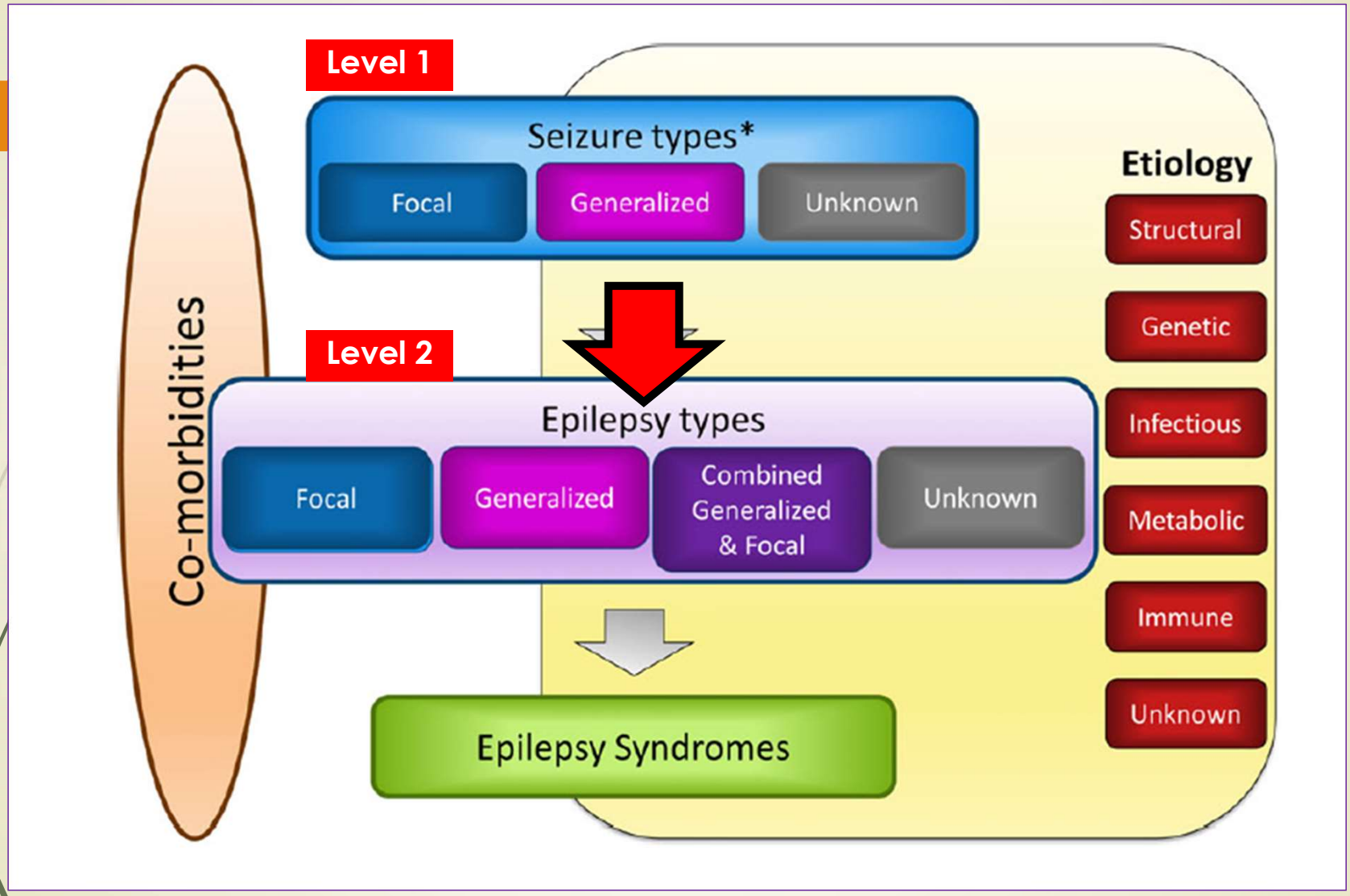
Epilepsia 2017

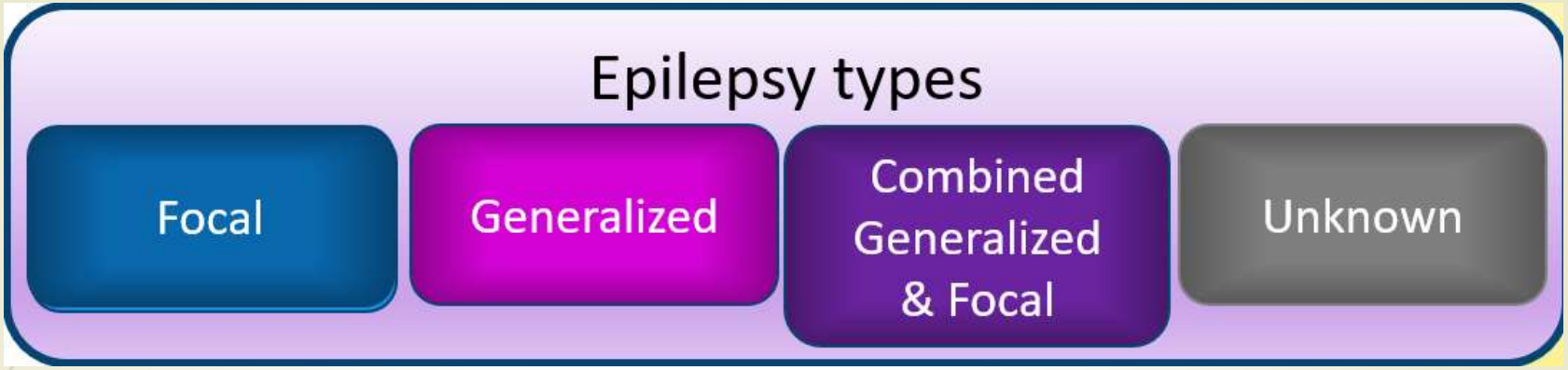

Onset
Focal
Generalized
Unknown
Awareness
• Aware
• Impaired awareness
Prominent symptoms
• Motor
• Non motor
• Motor: tonic-clonic
• Non motor (absence)
Motor
tonic-clonic
other motor
Non motor
Step 4
Spreading
Focal to bilateral tonic-clonic
Unclassified

Search for etiology in any level: seizure type, epilepsy type




Seizure types
Focal onset
Generalized onset
Unknown onset
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown
Tuberous Sclerosis
Vt B6 dependent Sz


Level 1
Seizure types*
Focal
Generalized
Unknown
Level 2
Epilepsy types
Focal
Generalized
Combined Generalized & Focal
Unknown
Epilepsy Syndromes
Co-morbidities
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown



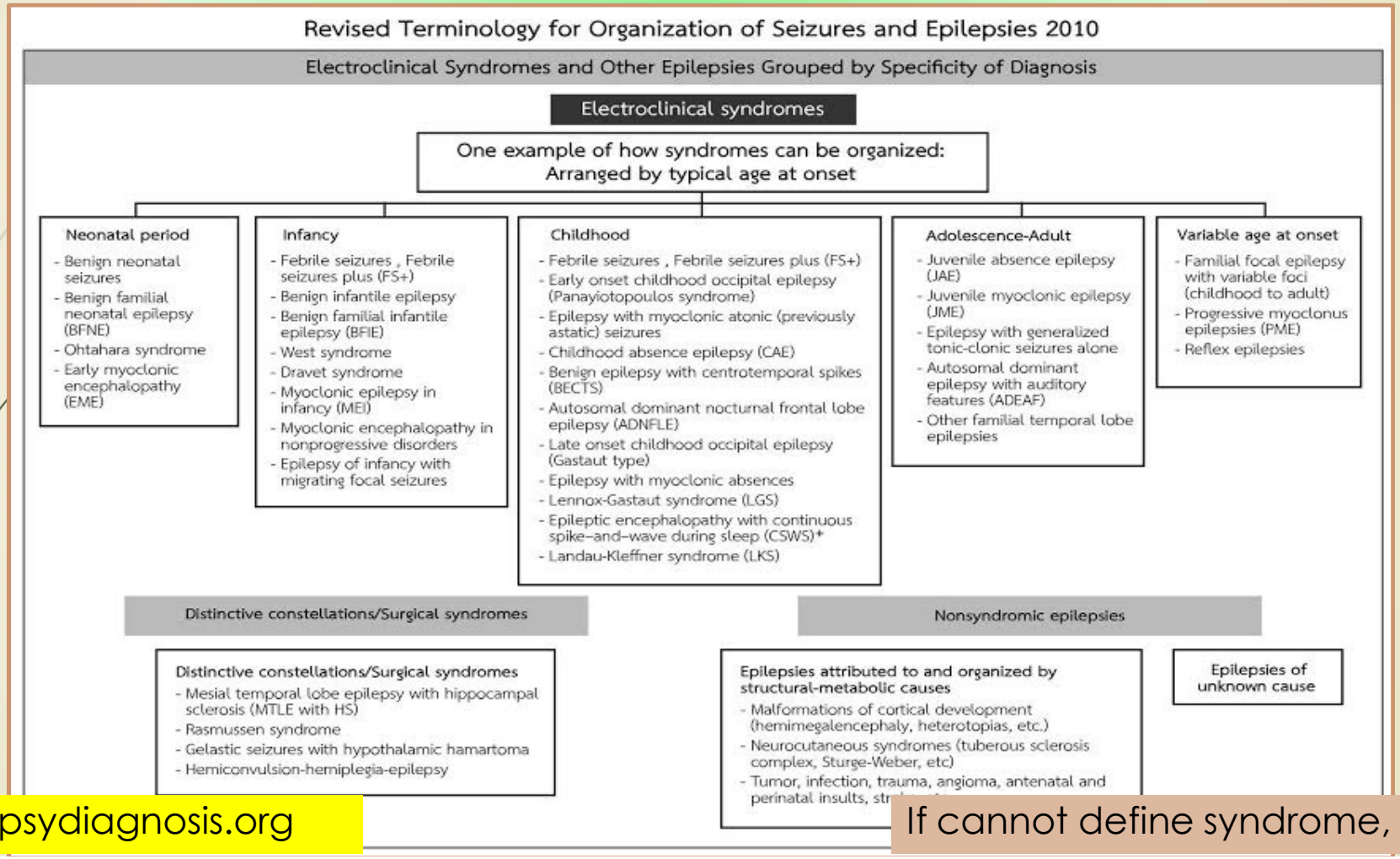
Level 2

Combined generalized and focal epilepsy types

- **Dravet syndrome**
- **Lennox Gastaut syndrome**
- ***KCNT1* gene mutation, EIEE, EME**

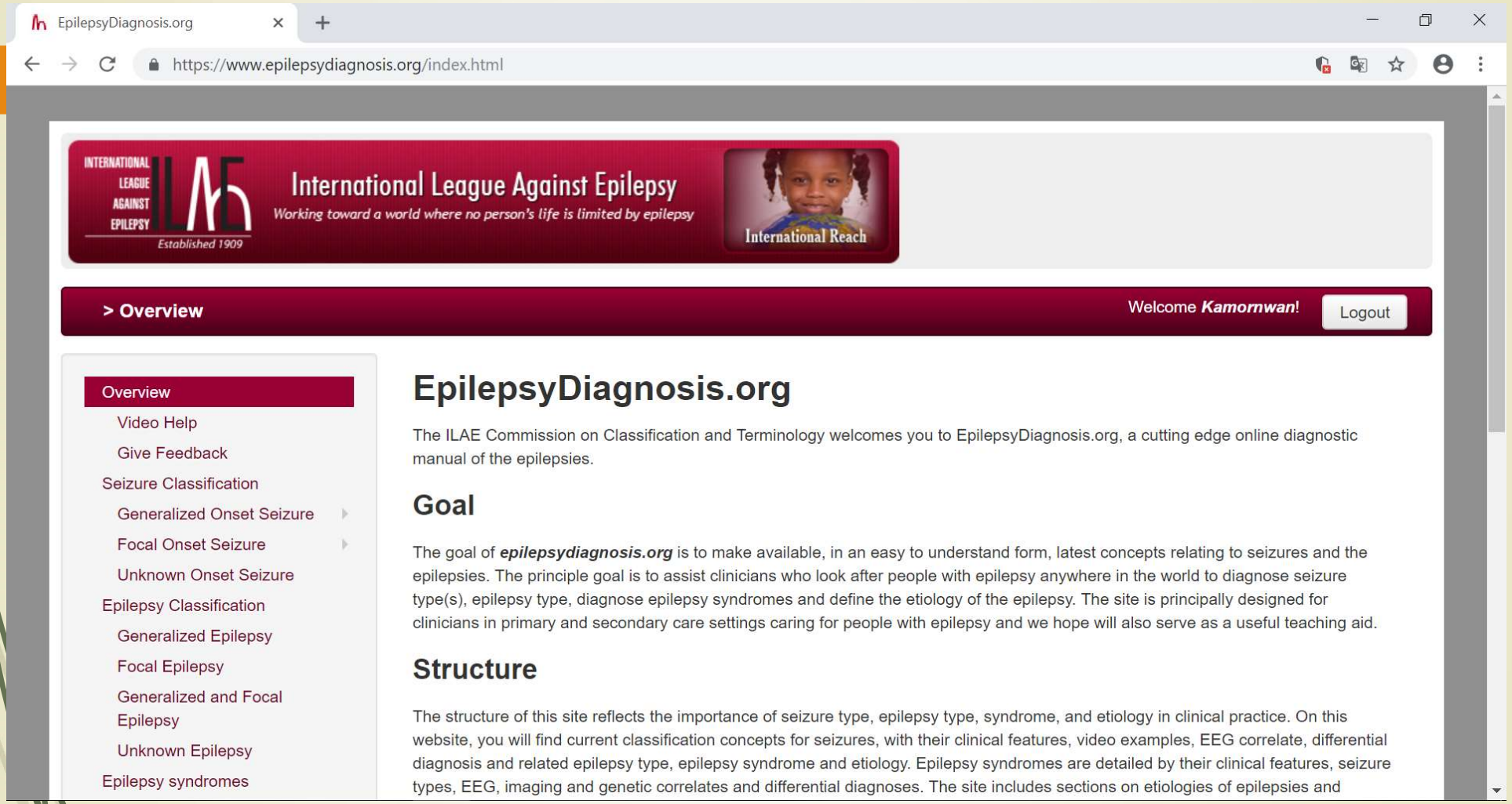
Level 3

Epilepsy syndrome

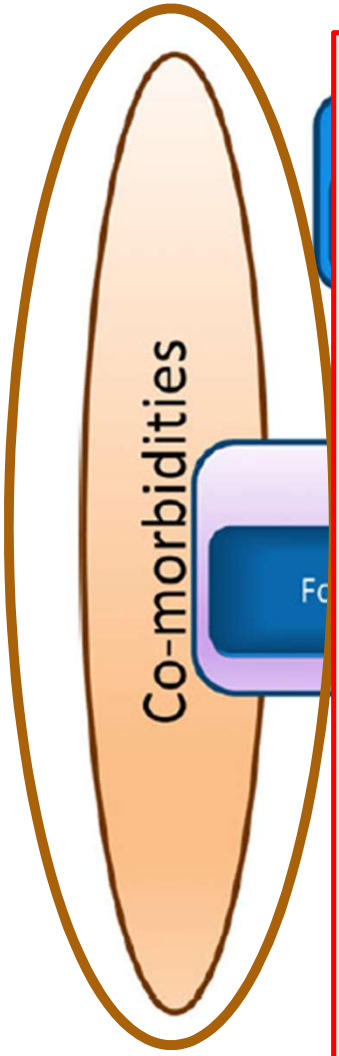


www.epilepsydiagnosis.org

If cannot define syndrome, it's ok



www.epilepsydiagnosis.org



Co-morbidities

- Neuropsychiatric
  - cognitive impairment
  - attention deficit
  - adjustment disorder
  - aggression
- Physical problem: hemiparesis, gait, movement problem
- Eyes and ears problem
- Eating problem
- etc

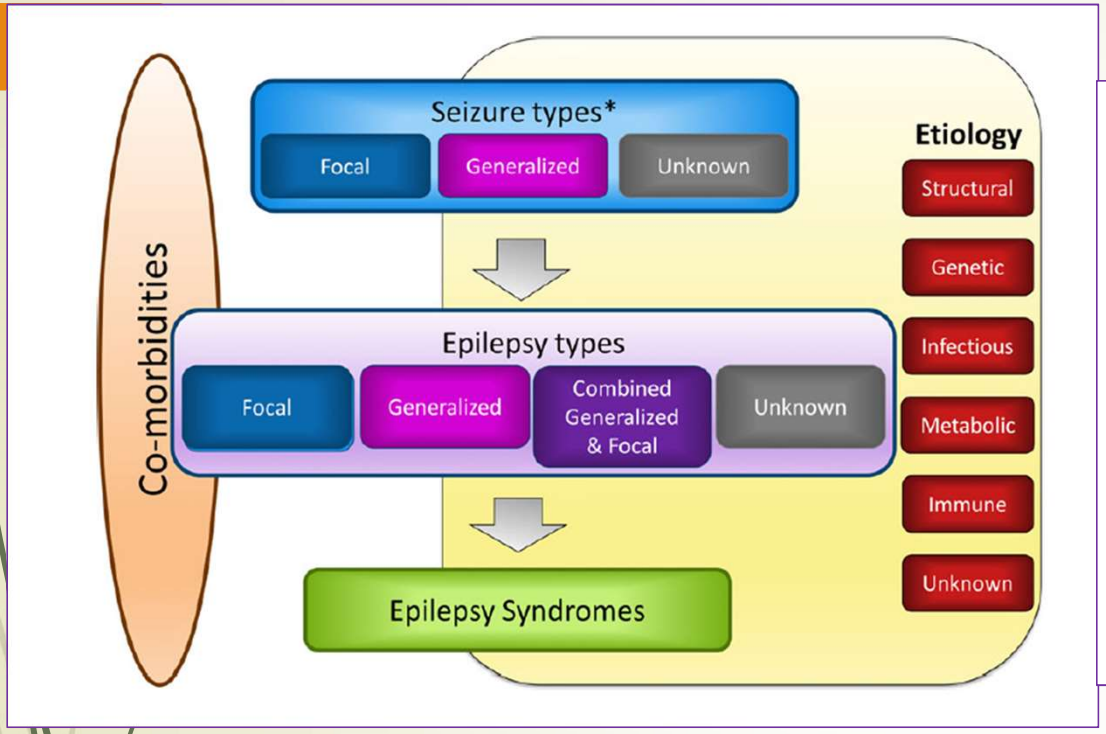
ILAE

Some terminology in 1989 ??

Idiopathic Generalized Epilepsies ✓

- Childhood Absence Epilepsy
- Juvenile Absence Epilepsy
- Juvenile Myoclonic Epilepsy
- Generalized Tonic-Clonic Seizures Alone

Genetic generalized epilepsy (GGE) ✓



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]

Time and experience will indicate whether the ILAE revisions meet their goal

Four-dimensional epilepsy classification

Classification of paroxysmal events and the four-dimensional epilepsy classification system.
Hans Luders, et al.

Table 1. Paroxysmal event classification.

Paroxysmal events (PE)	Epileptic		Ictal semiology (*ie*. automotorseizure)
			Epileptogenic zone (*ie*. left temporal)
			Etiology (*ie*. hippocampal sclerosis)
			Co-morbidities (*ie*. anxiety)
	Non-epileptic	Psychogenic	Ictal semiology (*ie*. clonic event)
			Etiology (*ie*. post-traumatic stress disorder)
			Co-morbidities (*ie*. none)
		Organic	Ictal semiology (*ie*. cataplexy event)
			Etiology (*ie*. narcolepsy)
			Co-morbidities (*ie*. none)

Epileptic Disord, Vol. 21, No. 1, February 2019

Ictal semiology

Auras *	Auditory aura*	
	Autonomic aura	Abdominal Aura
	Gustatory aura	
	Olfactory aura	
	Psychic aura	
	Somatosensory aura*	
	Vestibular aura	
	Visual aura*	
Autonomic seizure*	Bradycardic seizure	
	Emetic seizure	
	Sialorrheic seizure	
	Tachycardic seizure	
	Urinary seizure	
Dyscognitive seizure	Aphasic seizure	
	Akinetic seizure	
	Dialeptic seizure	
Motor seizure*	Simple motor seizures*	Clonic seizure*
		Epileptic spasm*
		Myoclonic seizure*
		Tonic seizure*
		Tonic-clonic seizure*
		Versive seizure*
	Complex motor seizures	Automotor seizure
		Gelastic seizure
		Hypermotor seizure
Special seizures	Astatic seizure	
	Atonic seizure	
	Central apneic seizure	
	Hypnopompic seizure	
	Hypomotor seizure	
	Negative myoclonic seizure*	
Asymptomatic EEG seizure		

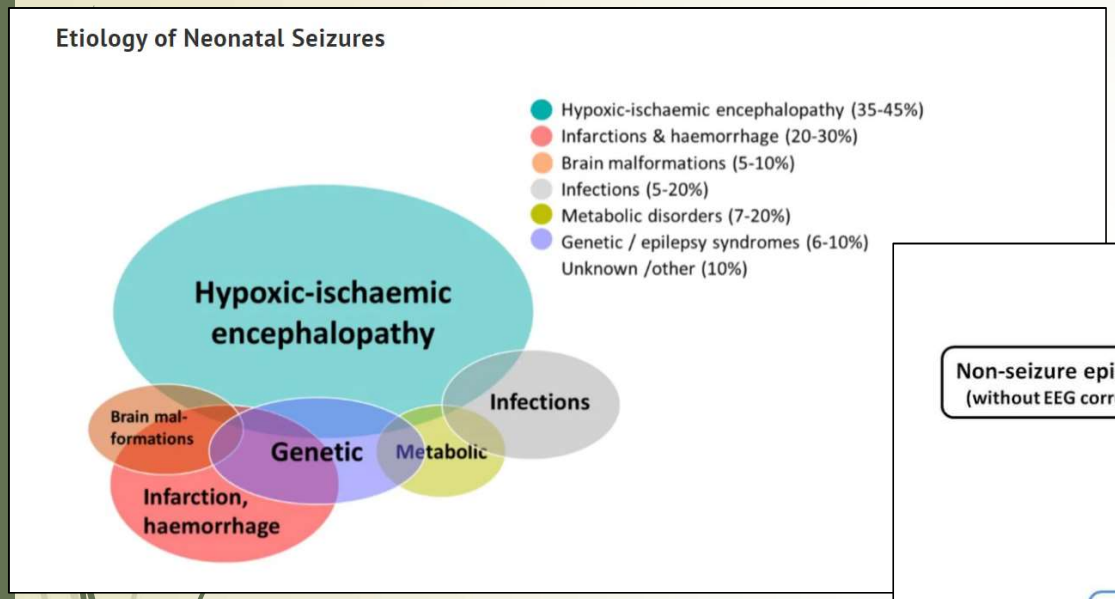
Epileptogenic Zone

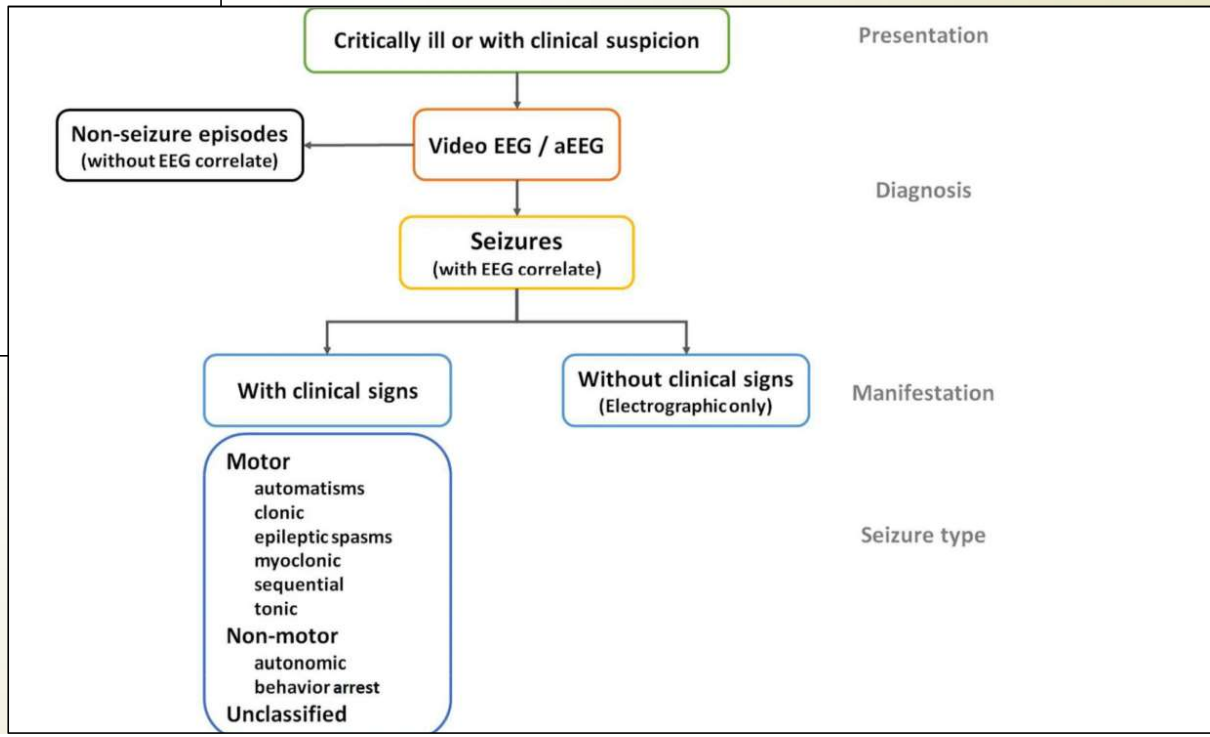
Generalized			
Focal	Hemis-phere*	Temporal*	Lateral temporal*
			Mesial temporal*
			Temporal pole*
			Basal temporal*
		Frontal*	Prefrontal lateral*
			Prefrontal mesial*
			Basal frontal*
			Premotor lateral*
			Premotor mesial*
		Central*	Centro-temporal*
			Mesial central*
		Parietal*	Mesial parietal*
			Lateral parietal*
		Occipital*	Lateral occipital*
			Mesial occipital*
		Cingulate*	Anterior cingulate*
			Mid cingulate*
			Posterior cingulate*
		Insula*	Anterior insula*
			Posterior insula*
Multifocal			
Unknown			

Etiology

Structural*
Genetic
Inflammatory
Infectious
Unknown

Neonatal seizure classification (ongoing)





ILAE website

2015

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

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