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

ILAE POSITION PAPER

Operational classification of seizure types by the International League Against Epilepsy: Position Paper of the ILAE Commission for Classification and Terminology

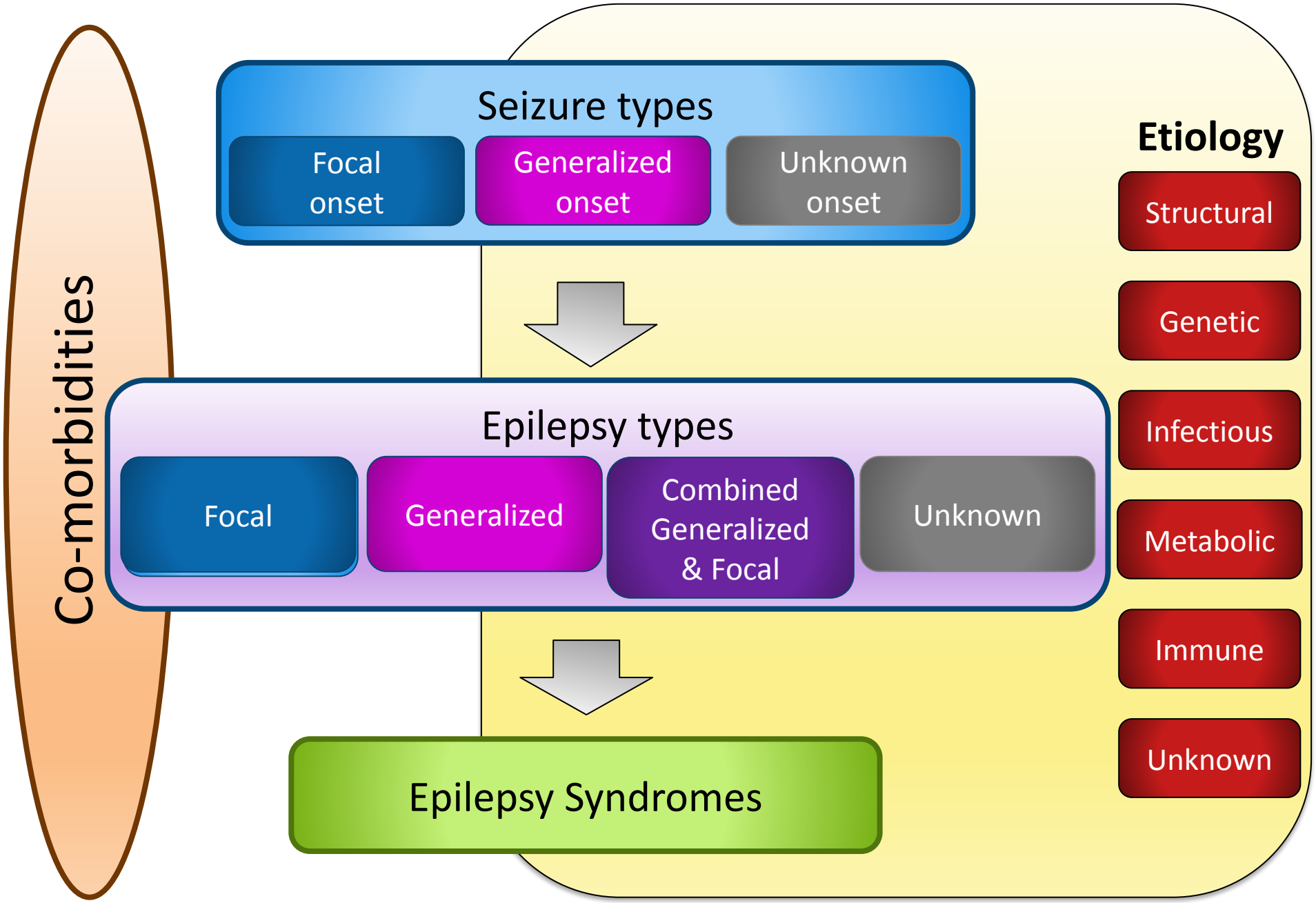
*Robert S. Fisher, †J. Helen Cross, ‡Jacqueline A. French, §Norimichi Higurashi, ¶Edouard Hirsch, #Floor E. Jansen, **Lieven Lagae, ††Solomon L. Moshé, ‡‡Jukka Peltola, §§Eliane Roulet Perez, ¶¶Ingrid E. Scheffer, and ##***Sameer M. Zuberi

Epilepsia, **(*):1–9, 2017
doi: 10.1111/epi.13670

Classification of the Epilepsies

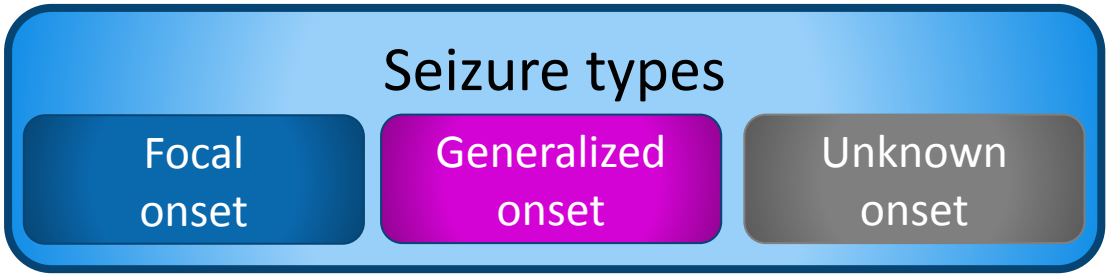
Purpose: for clinical diagnosis

Transparent language: use words that mean what they say


Co-morbidities
Seizure types
Focal onset
Generalized onset
Unknown onset
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown
Epilepsy types
Focal
Generalized
Combined Generalized & Focal
Unknown
Epilepsy Syndromes

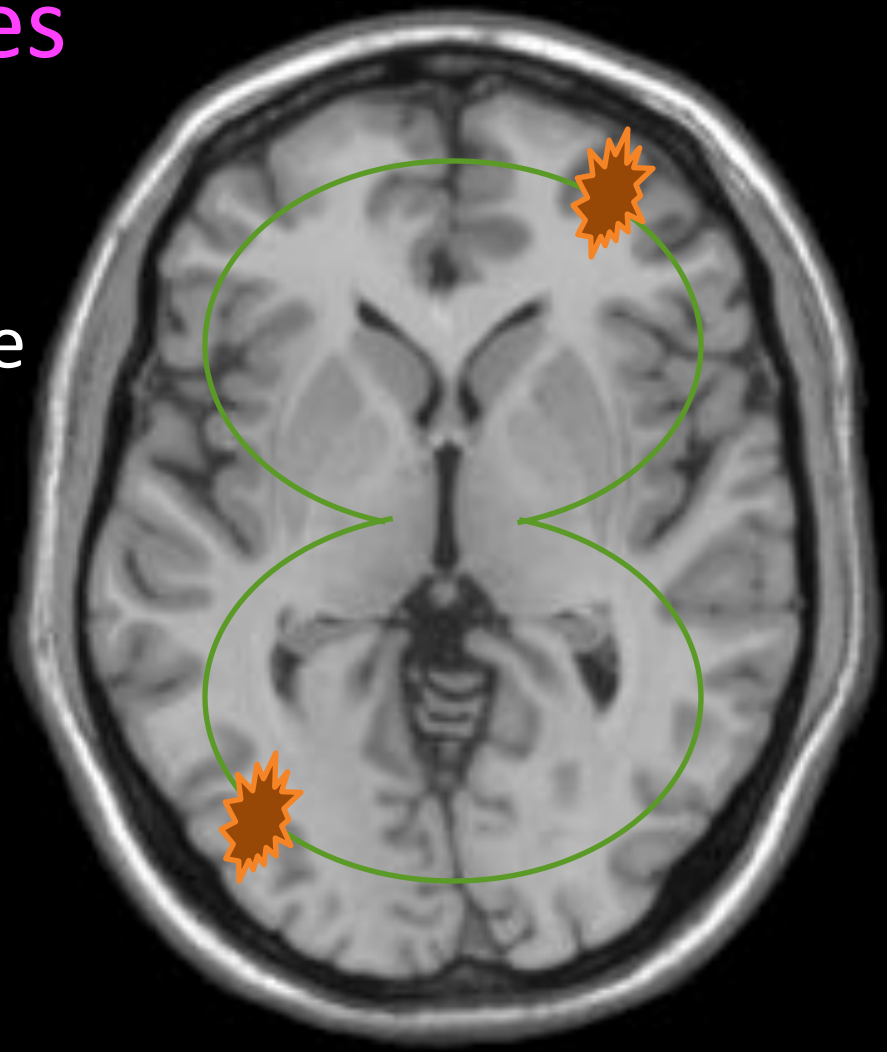
1. Seizure types

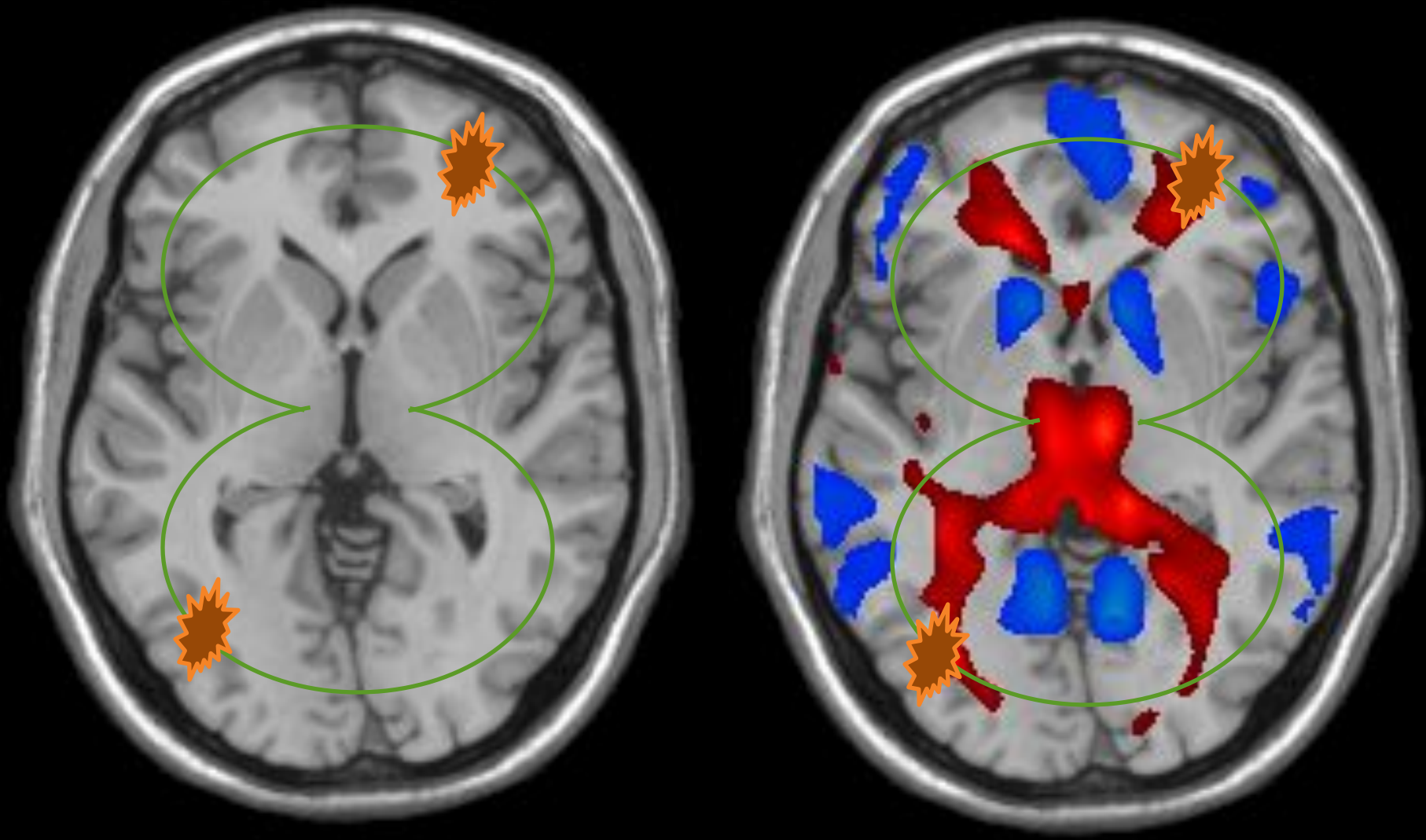
- Certain that events are epileptic seizures – **not** referring to distinguishing epileptic versus non-epileptic
- In some settings classification according to seizure type may be maximum level of diagnosis possible
- In other cases simply too little information to be able to make a higher level diagnosis
  - eg. when a patient has only had a single event


Seizure types
Focal onset
Generalized onset
Unknown onset

Generalized seizures

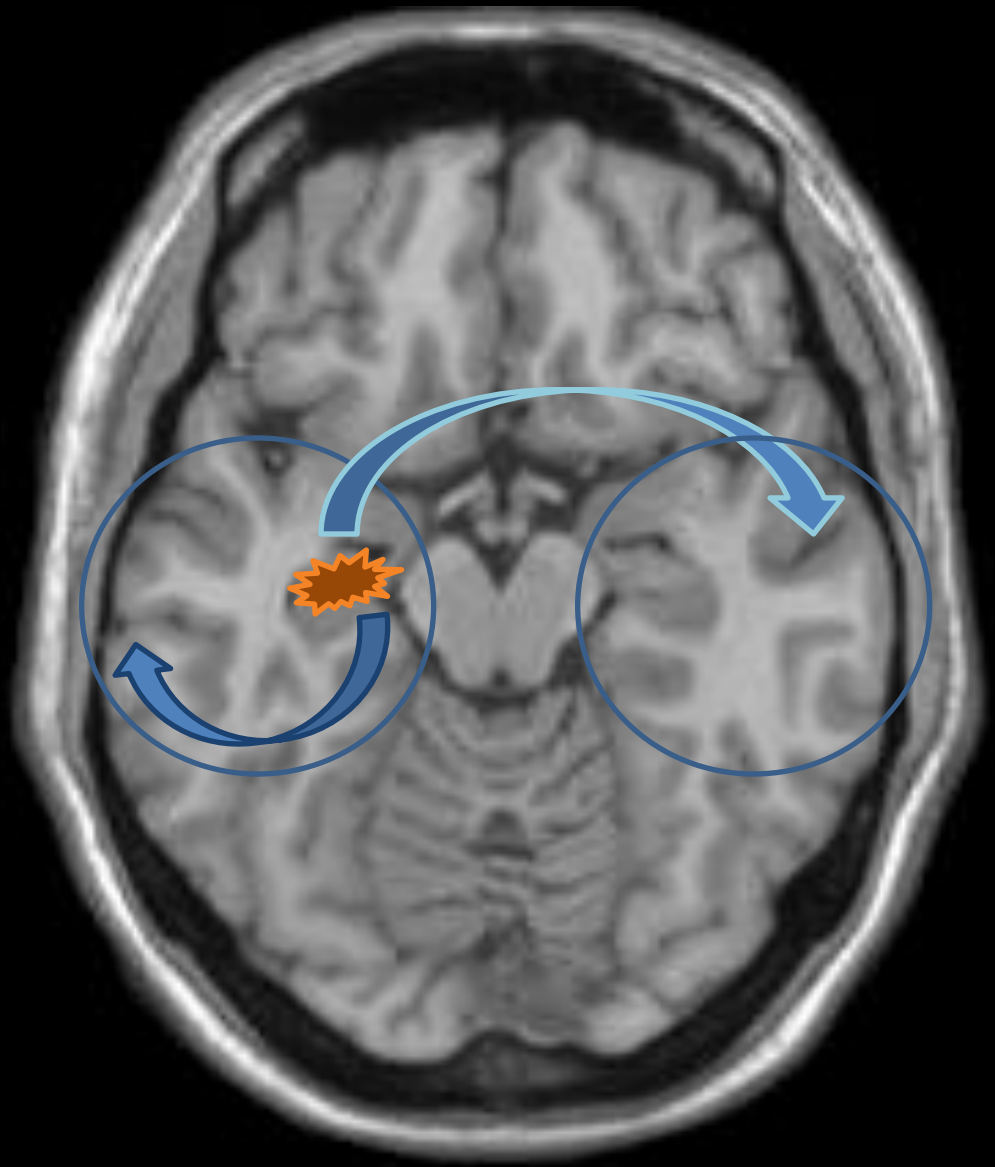
- Originate at some point within and rapidly engage bilaterally distributed networks
- Can include cortical and subcortical structures but not necessarily the entire cortex



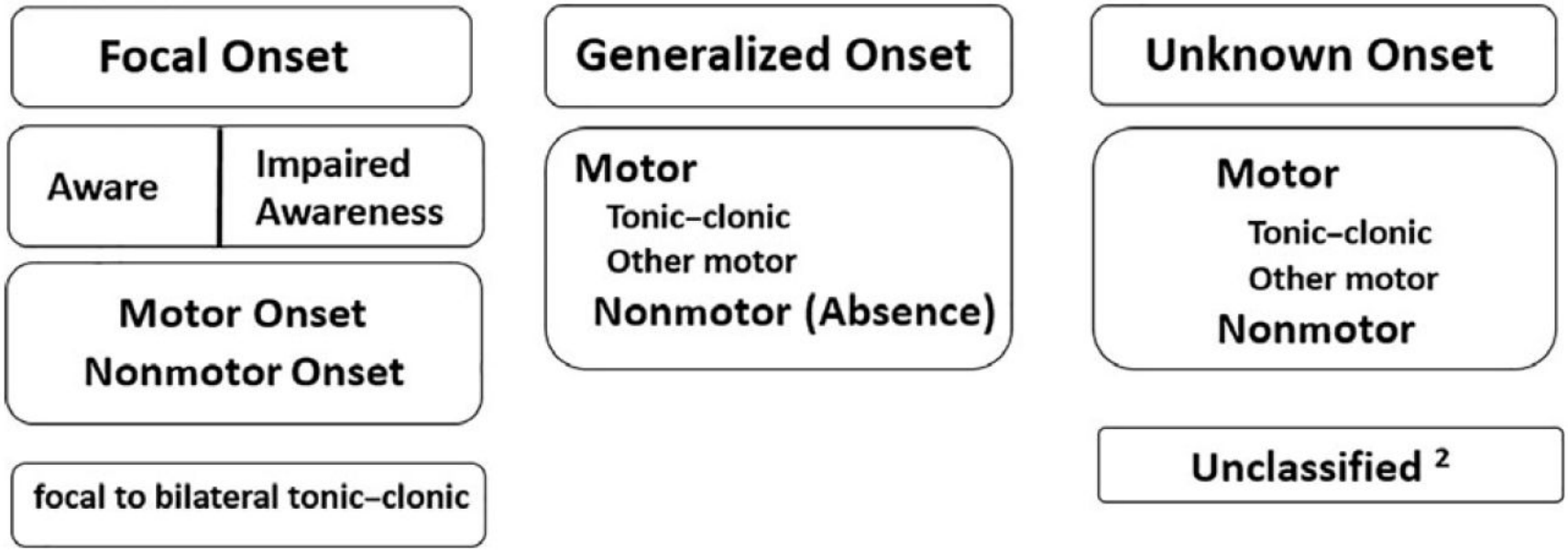


Focal seizures

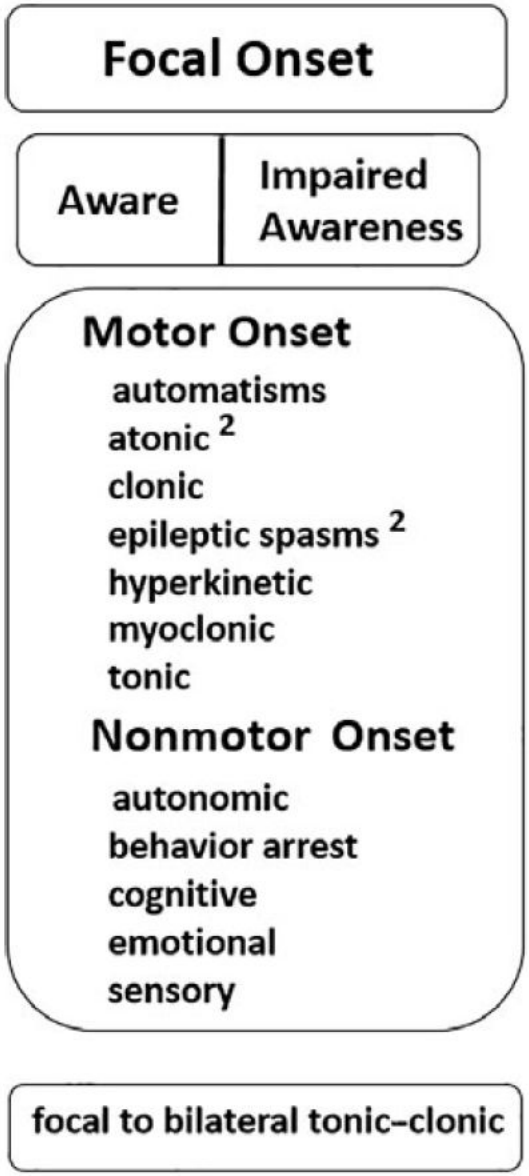
- Originate within networks limited to one hemisphere
- May be discretely localized
  or more widely distributed….



ILAE 2017 Classification of Seizure Types Basic Version [1]



ILAE 2017 Classification of Seizure Types Expanded Version [1]



Notes

- Atonic seizures and epileptic spasms would *not* have level of awareness specified
- Pedalling grouped in hyperkinetic rather than automatisms (arbitrary)
- Cognitive seizures
  - impaired language
  - other cognitive domains
  - positive features eg déjà vu, hallucinations, perceptual distortions
- Emotional seizures: anxiety, fear, joy, etc

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

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 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]

Note

When a seizure type begins with ”focal, generalized or absence” then the word “onset” can be presumed

Terms no longer in use

- Complex partial
- Simple partial
- Partial
- Psychic
- Dyscognitive
- Secondarily generalized tonic-clonic

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

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

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

Note

Clarify features of seizures but do not define unique seizure types

Free text descriptors encouraged

ILAE POSITION PAP

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,	Adapted fr Ref. 12

Ins

Summary of rules for

1 Onset: Decide whe
eralized,
onset is u
2 Awarene
classify b
as a clas
the old s
awarenes
zures.
3 Impaired
focal imp
impaired
4 Onset pr
first pron
sient beha
5 Behavior
shows an
the entire
6 Motor/no

Table 3.

Old term for	
Absence	
Absence, aty	
Absence, typ	
Akinetic	
Astatic	
Atonic	
Aura	
Clonic	
Complex p	
Convulsion	
	clonic], focal to bilateral tonic–clonic
Dacrystic	Focal [aware or impaired awareness] emotional (dacrystic)
Dialeptic	Focal impaired awareness
Drop attack	[Focal/generalized] atonic, [focal/generalized] tonic
Fencer's posture (asymmetric tonic)	Focal [aware or impaired awareness] motor tonic
Figure-of-4	Focal [aware or impaired awareness]

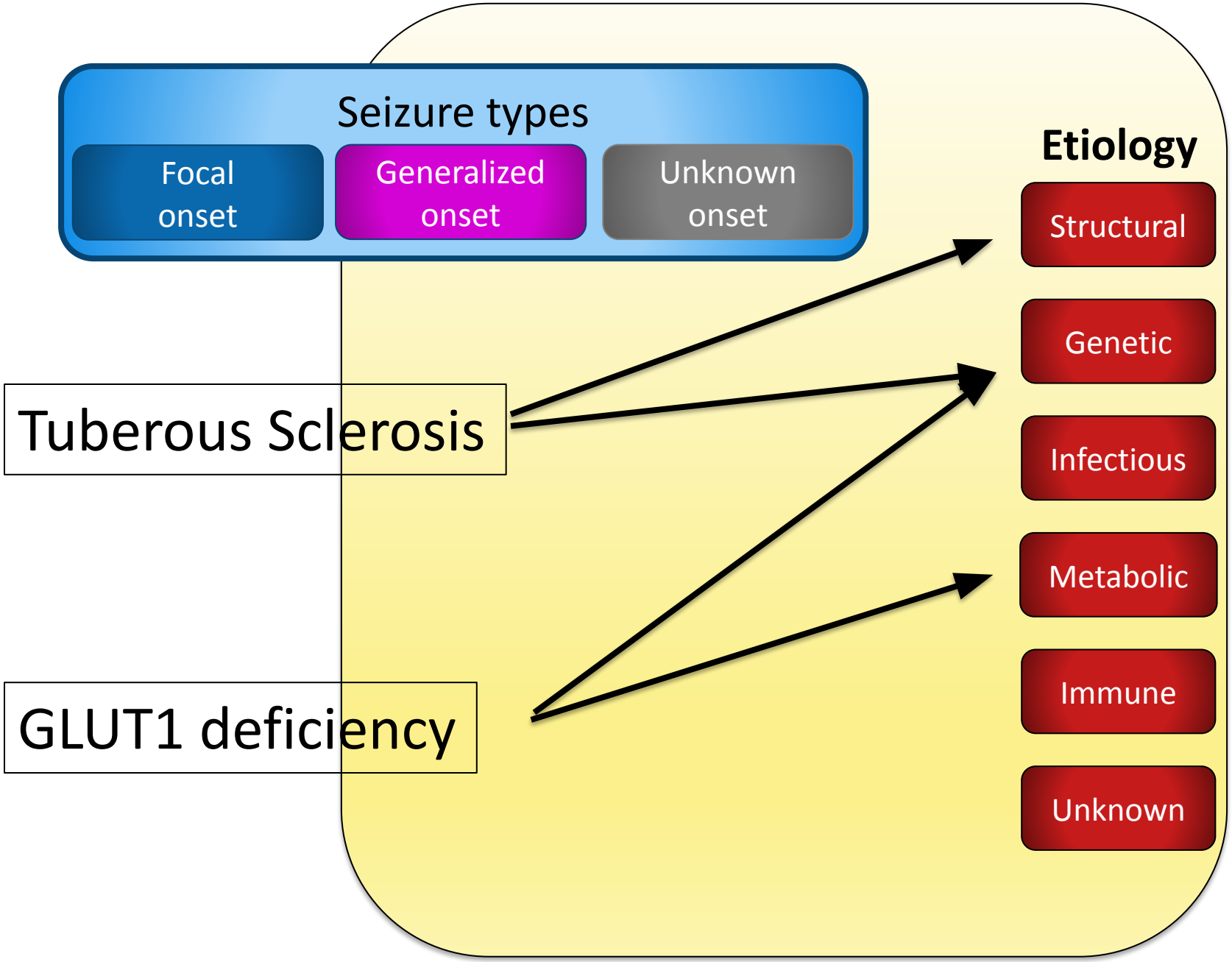
Table 4. Abbreviations for the most important seizure types

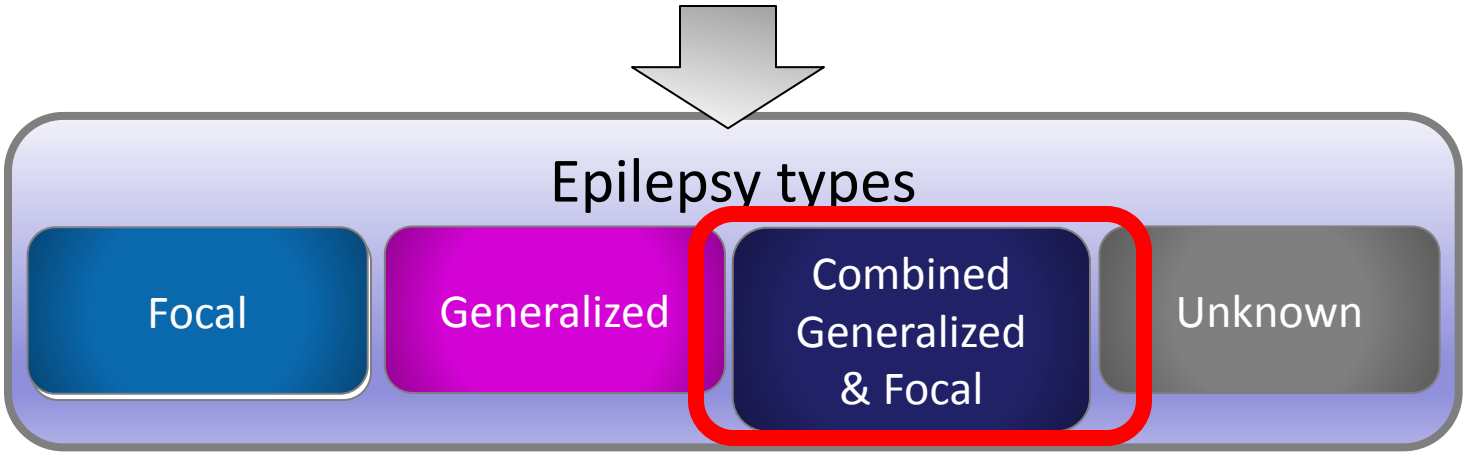
Seizure type	Abbreviations
Focal aware seizure	FAS
Focal impaired awareness seizure	FIAS
Focal motor seizure	FMS
Focal nonmotor seizure	FNMS
Focal epileptic spasm	FES
Focal to bilateral tonic–clonic seizure	FBTCS
Generalized tonic–clonic seizure	GTCS
Generalized absence seizure	GAS
Generalized motor seizure	GMS
Generalized epileptic spasm	GES
Unknown onset tonic–clonic seizure	UTCS

ce or ed — Adapted fr Ref. 11
c — New 12
and s a — 12
illary, — Adapted fr Ref. 12 12
or — New New
e — Adapted fr Ref. 12 New
New
sting — Adapted fr Ref. 12 New
ng
Adapted fr Ref. 12

rms may occur: Grimacing, head nodding, or subtle eye
ly occur in clusters. Infantile spasms are the best known

the following conditions: (1) At least two unprovoked
art; (2) one unprovoked (or reflex) seizure and a
o the general recurrence risk (at least 60%) after two
he next 10 years; (3) diagnosis of an epilepsy
resolved for individuals who had an age-dependent — 3


Seizure types
Focal onset
Generalized onset
Unknown onset
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown
Tuberous Sclerosis
GLUT1 deficiency



- Where unable to make an Epilepsy Syndrome diagnosis or a diagnosis of Etiology
- Many examples
  - Temporal lobe epilepsy
  - Generalized tonic-clonic seizures in a 5 year old with generalized spike-wave
  - Both focal impaired awareness seizures and absence seizures in a patient
  - Cannot tell if tonic-clonic seizure is focal or generalized

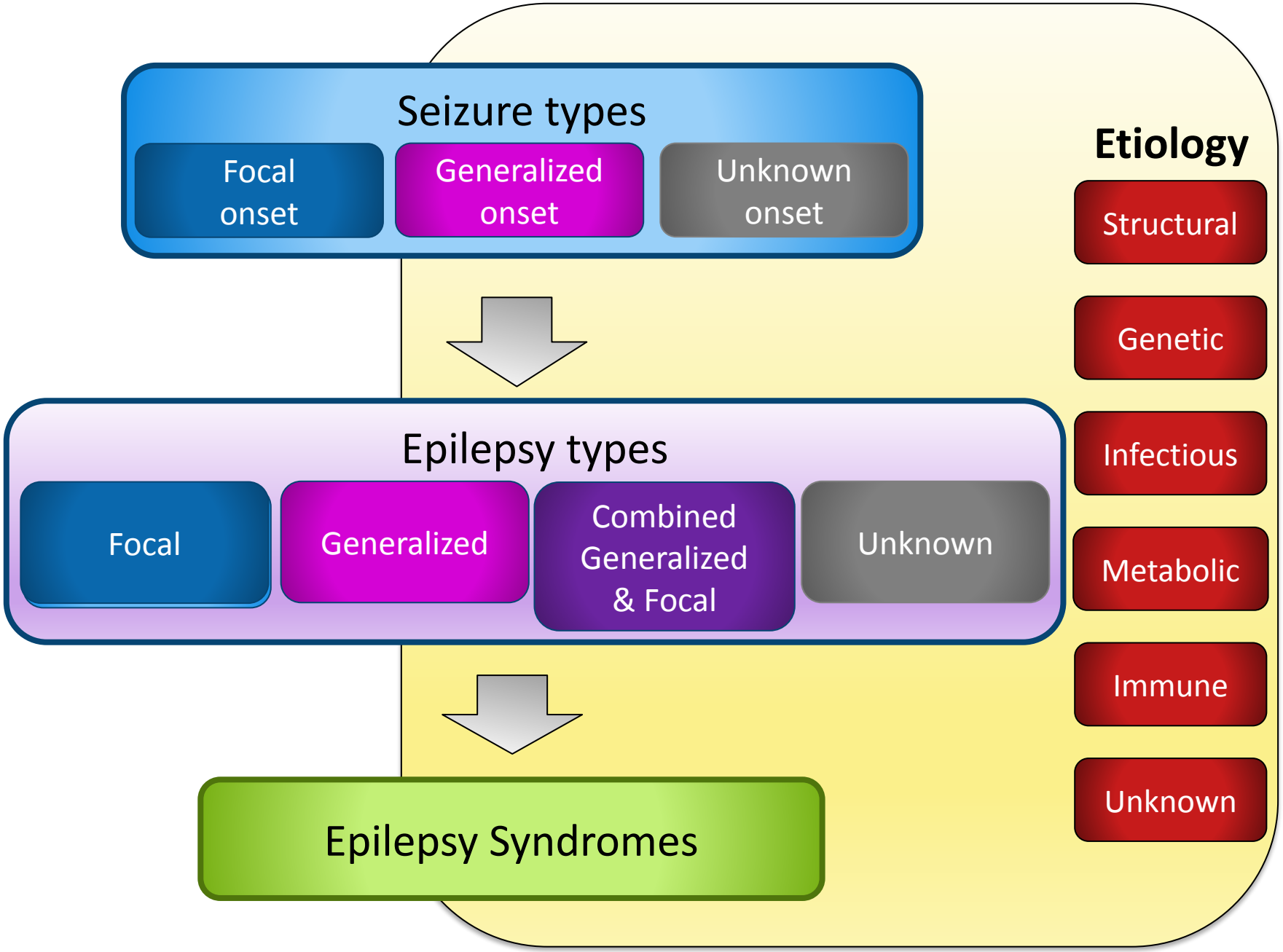
Generalized *and* Focal Epilepsies

- Combined focal and generalized epilepsies

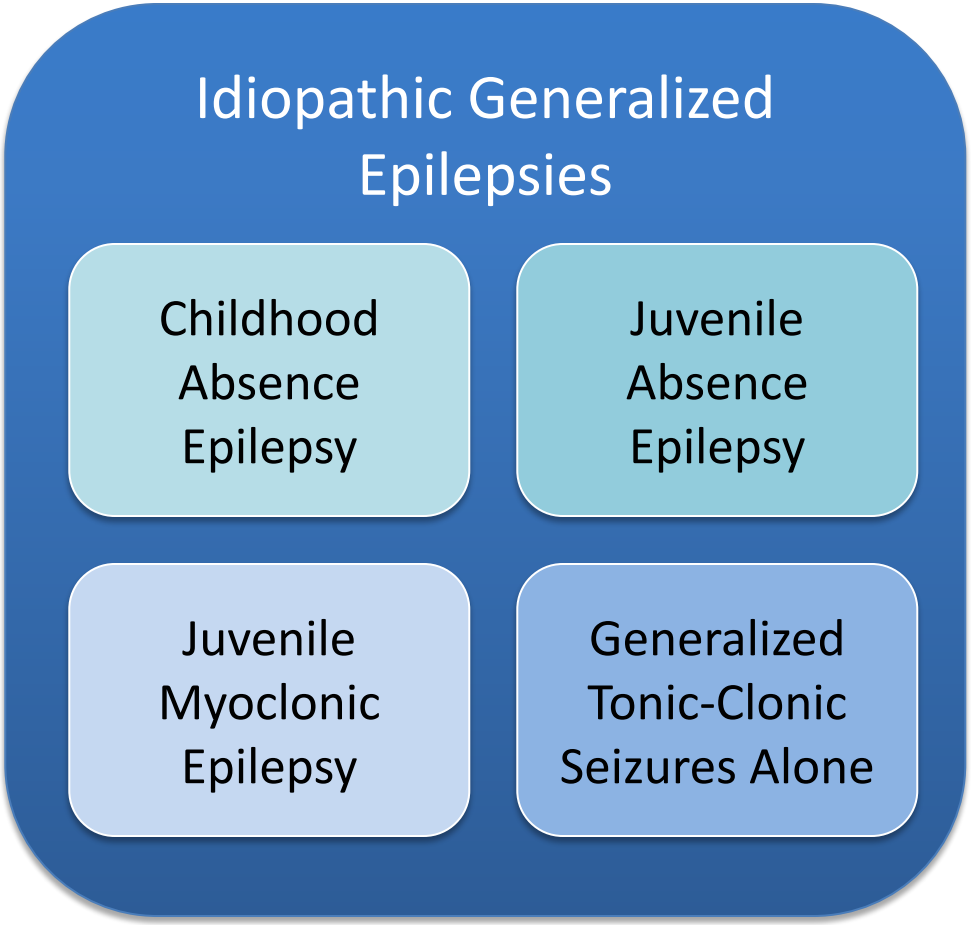
  Examples

  – Dravet syndrome

- What do with
  – Multifocal epilepsies? □ focal
  – Hemispheric epilepsies? □ focal


Seizure types
Focal onset
Generalized onset
Unknown onset
Epilepsy types
Focal
Generalized
Combined Generalized & Focal
Unknown
Epilepsy Syndromes
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown

Old term
‘Idiopathic Generalized Epilepsies’



Genetic *versus* idiopathic

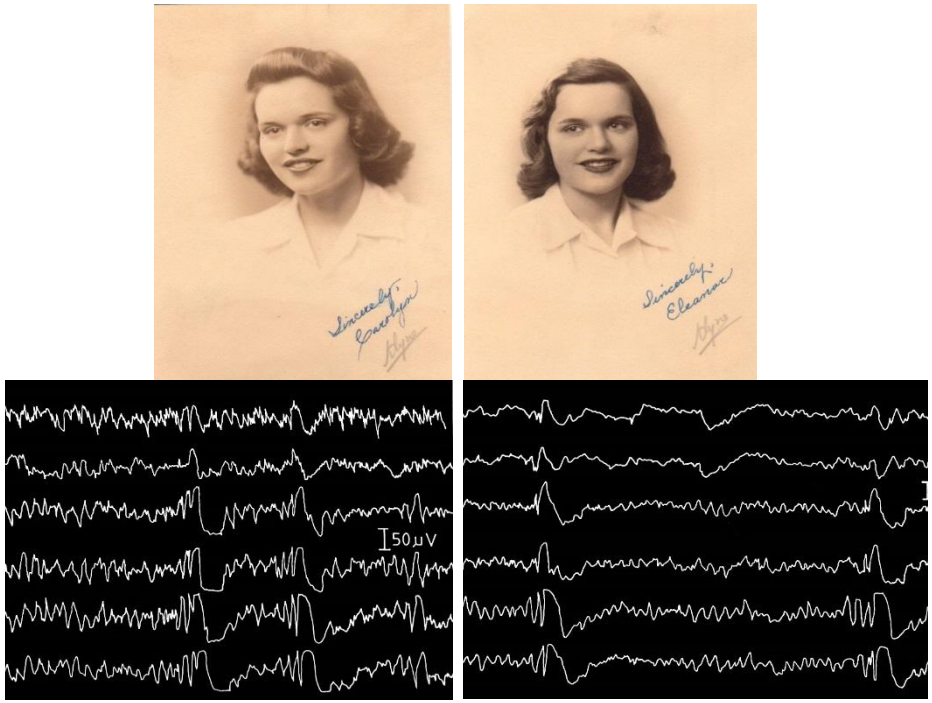
- 'Idiopathic' = presumed hereditary predisposition

- Genetic ≠ inherited
  - Importance *of de novo* mutations in both mild and severe epilepsies

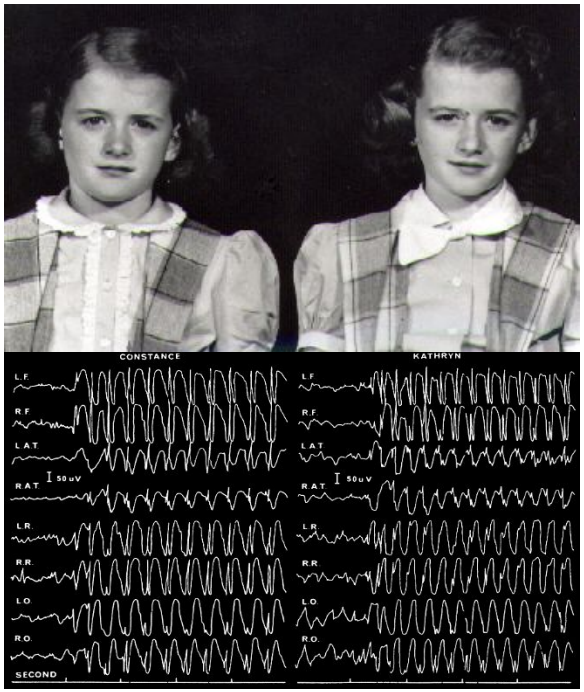
- Critical problem of stigma in some parts of the world

Genetic ≠ Gene testing

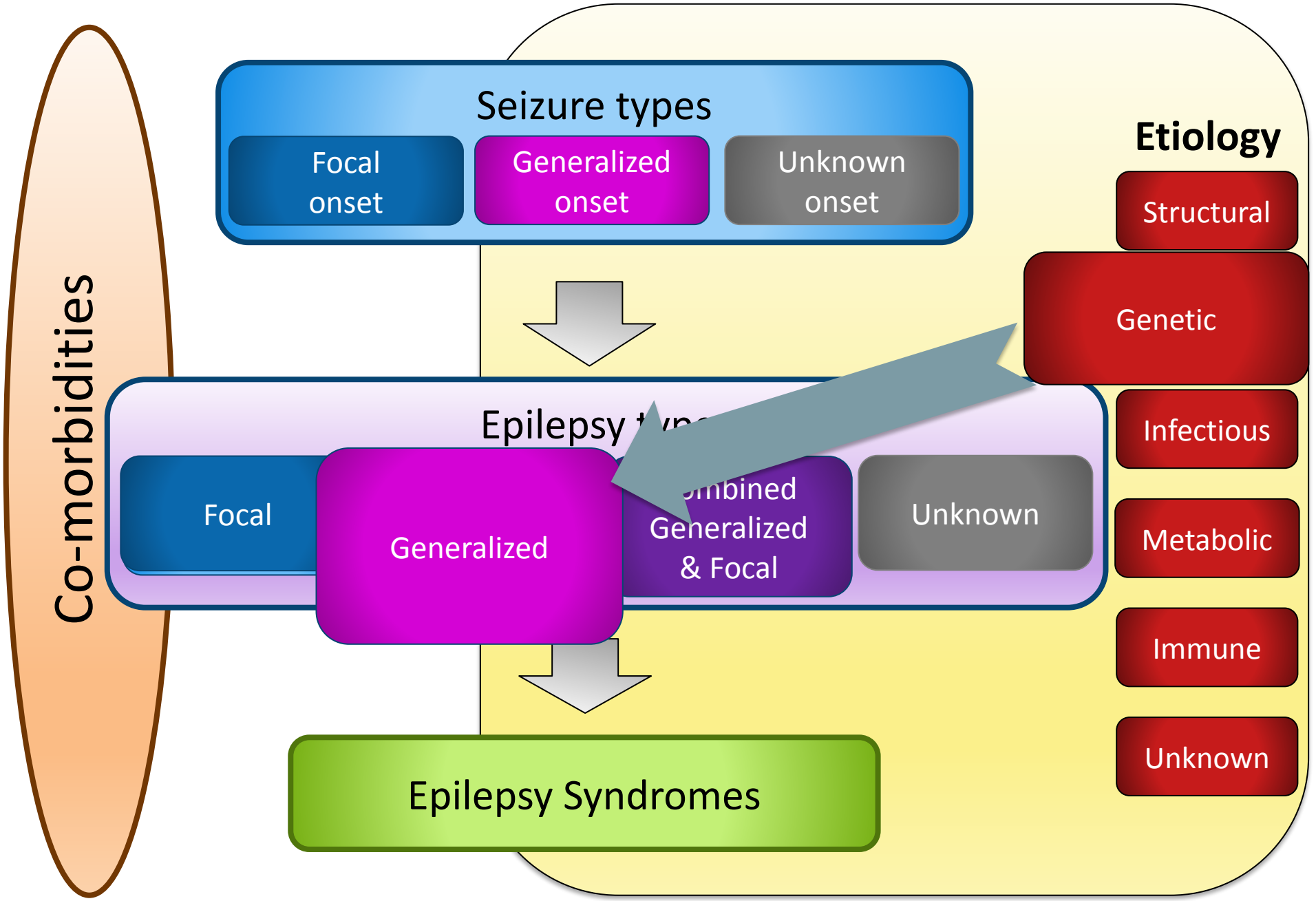
- Usually the mutation is *not* known
- Access to molecular genetic testing *not* necessary
- Diagnosed on clinical research eg. twin, family studies



JME pair; Lennox 1941



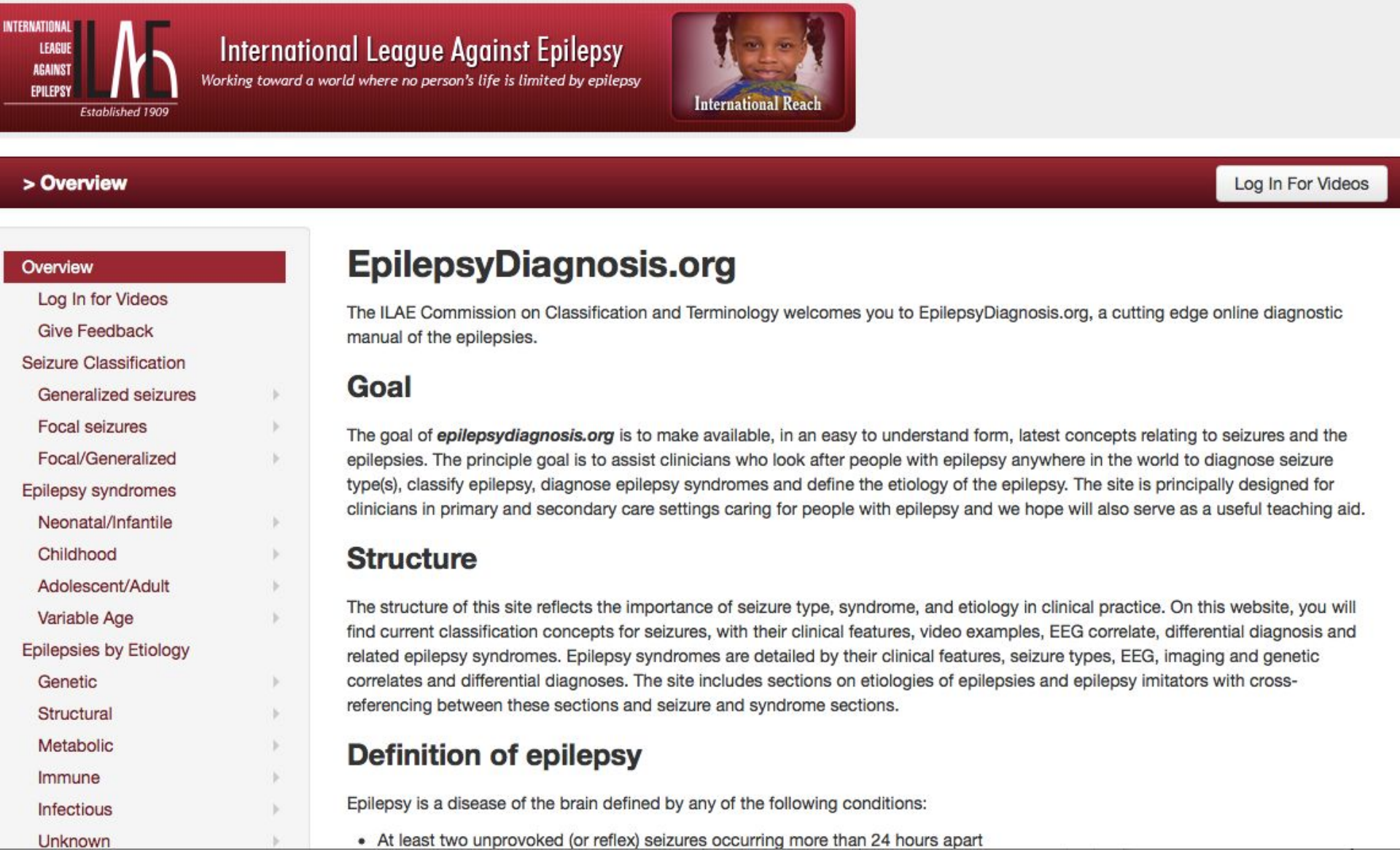
CAE pair; Lennox 1950


Co-morbidities
Seizure types
Focal onset
Generalized onset
Unknown onset
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown
Epilepsy t
Focal
Generalized
mbined Generalized & Focal
Unknown
Epilepsy Syndromes

Epilepsy syndromes

- There are no approved ILAE epilepsy syndromes

https://www.epilepsydiagnosis.org



Overview
- Log In for Videos
- Give Feedback

Seizure Classification
- Generalized seizures
- Focal seizures
- Focal/Generalized

Epilepsy syndromes
- Neonatal/Infantile
- Childhood
- Adolescent/Adult
- Variable Age

Epilepsies by Etiology
- Genetic
- Structural

ABSENCE - TYPICAL

Clinical Overview | Videos | EEG | Differential diagnoses | Related syndromes

A **typical absence** seizure is a generalized seizure with abrupt onset and offset of altered awareness which can vary in severity (see specific syndromes). Memory for events during the seizures is usually impaired although there may be some retained awareness particularly for adolescents. Clonic movements of eyelids, head, eyebrows, chin, perioral or other facial parts may occur, most typically at 3Hz. Myoclonus of limbs can rarely occur. Oral and manual automatisms are common and there may be perseveration of behaviors occurring prior to seizure onset. Absence seizures were previously known as 'petit mal' seizures. Absence status epilepticus can occur.

CAUTION Individual absence seizure longer than 45 seconds or with a post-ictal phase → consider focal seizure.

CAUTION Onset of absence seizures < 4 years → consider glucose transporter disorders.

Overview
Log In for Videos
Give Feedback
Seizure Classification
Generalized seizures
Focal seizures
Focal/Generalized
Epilepsy syndromes
Neonatal/Infantile
Childhood
Adolescent/Adult
Variable Age
Epilepsies by Etiology
Genetic
Structural
Metabolic
Immune
Infectious
Unknown

ABSENCE - TYPICAL

Clinical Overview | Videos | EEG | Differential diagnoses | Related syndromes

Videos

Typical Absence Seizure Video 1

An absence seizure occurs in hyperventilation with behavioural arrest, and upward deviation of the eyes.

Typical Absence Seizure Video 2

An absence seizure occurs in hyperventilation with behavioral arrest (stops hyperventilating) and impaired responsiveness. GSW appearing on the video as it plays denotes when generalized spike-and-wave appears on the patient's EEG.

Typical Absence Seizure Video 3

An absence seizure occurs in hyperventilation, with arrest of activity, staring and loss of responsiveness.

Typical Absence Seizure Video 4

An absence seizure occurs in hyperventilation with arrest of activity, mild oral automatisms, and lack of responsiveness until the seizure ends.

Typical Absence Seizure Video 5

Two absence seizures occur, with the eyes opening, staring and 3 Hz eyelid flutter associated with the events. GSW appearing on the video as it plays denotes when generalized spike-and-wave appears on the patient's EEG.

Overview
Log In for Videos
Give Feedback
Seizure Classification
Generalized seizures
Focal seizures
Focal/Generalized
Epilepsy syndromes
Neonatal/Infantile
Childhood
Adolescent/Adult
Variable Age
Epilepsies by Etiology
Genetic
Structural
Metabolic
Immune
Infectious
Unknown

Typical Absence Seizure Video 1

n of the eyes.

) and impaired responsiveness. GSW

he patient's EEG.

Typical Absence Seizure Video 3

An absence seizure occurs in hyperventilation, with arrest of activity, staring and loss of responsiveness.

Typical Absence Seizure Video 4

An absence seizure occurs in hyperventilation with arrest of activity, mild oral automatisms, and lack of responsiveness until the seizure ends.

Typical Absence Seizure Video 5

Two absence seizures occur, with the eyes opening, staring and 3 Hz eyelid flutter associated with the events. GSW appearing on the

Overview
Log In for Videos
Give Feedback
Seizure Classification
Generalized seizures
Focal seizures
Focal/Generalized
Epilepsy syndromes
Neonatal/Infantile
Childhood
Adolescent/Adult
Variable Age
Epilepsies by Etiology
Genetic
Structural
Metabolic
Immune
Infectious
Unknown
Unclassified epilepsies
Epilepsy imitators
Glossary

EPILEPSY IMITATORS

1. OVERVIEW
2. SYNCOPE AND ANOXIC SEIZURES
   a. Vasovagal syncope
   b. Reflex anoxic seizures
   c. Breath-holding attacks
   d. Hyperventilation syncope
   e. Compulsive valsalva
   f. Neurological syncope
   g. Imposed upper airways obstruction
   h. Orthostatic intolerance
   i. Long QT and cardiac syncope
   j. Hyper-cyanotic spells
3. BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS
   a. Daydreaming /inattention
   b. Infantile gratification
   c. Eidetic imagery
   d. Tantrums and rage reactions
   e. Out of body experiences
   f. Panic attacks
   g. Dissociative states
   h. Non-epileptic seizures
   i. Hallucinations in psychiatric disorders
   j. Fabricated / factitious illness
4. SLEEP RELATED CONDITIONS
   a. Sleep related rhythmic movement disorders
   b. Hypnogogic jerks
   c. Parasomnias

Benign

- Many epilepsies not benign
  - CAE – psychosocial impact
  - BECTS – learning concerns
- Replaced by terms:
  - Self-limited
  - Pharmacoresponsive
- No longer use
  - Malignant
  - Catastrophic

Developmental and/or Epileptic encephalopathies

Epileptic activity itself contributes to severe cognitive and behavioral impairment above and beyond that expected from the underlying pathology and that these can worsen over time

Berg et al

Developmental *and/or* Epileptic Encephalopathy

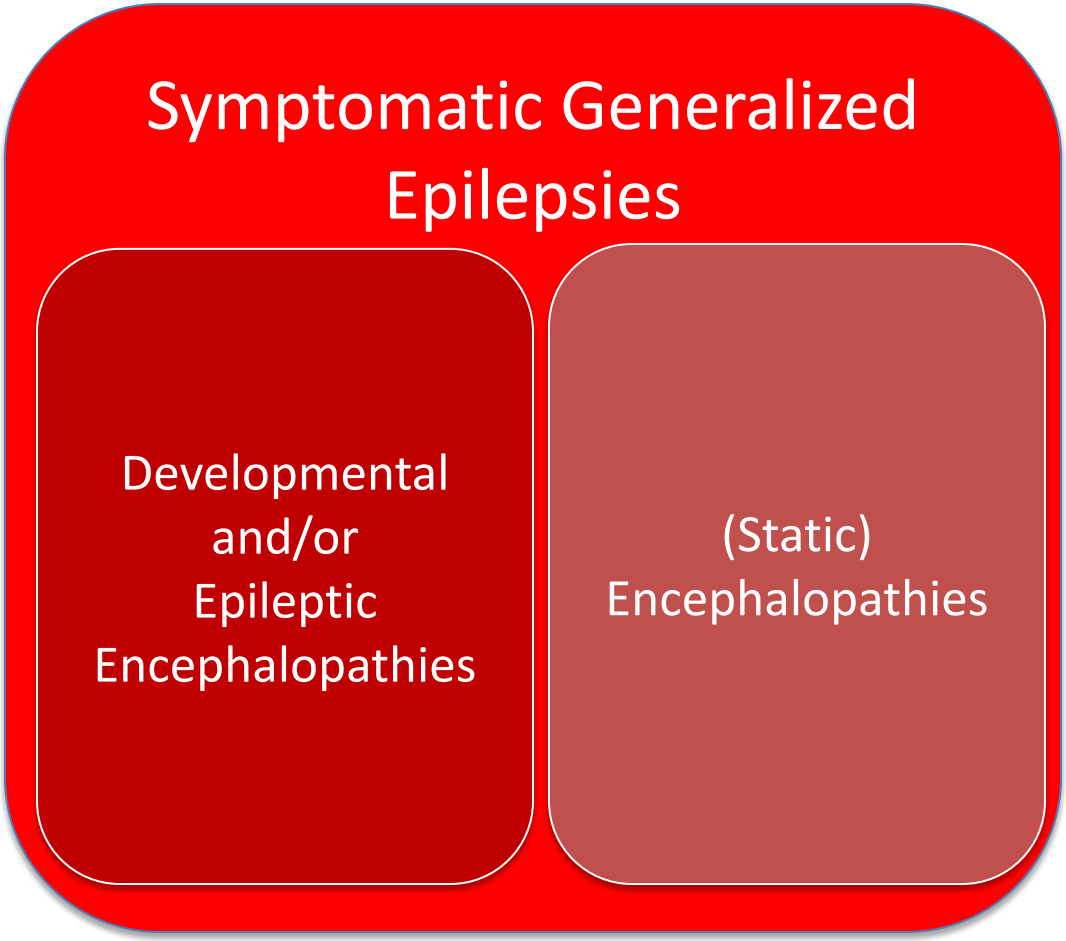
- For many encephalopathies, there is a developmental component *independent* of the epileptic encephalopathy
- Developmental delay may precede seizure onset
- Co-morbidities
  eg. cerebral palsy, autism spectrum disorder, intellectual disability
- Outcome poor even though seizures stop
  eg. KCNQ2, STXBP1 encephalopathies

Developmental *and/or* Epileptic Encephalopathy

- Developmental encephalopathy
  - May begin in utero
  - Post birth
- Epileptic encephalopathy
  - Can occur at any age
  - May have remediable component – right vs wrong AED
- Move towards *GENE* encephalopathy
  - eg. *CDKL5* encephalopathy, *SCN2A* encephalopathy

Old terms
‘Symptomatic Generalized Epilepsies’

- Used for two different groups of disorders



ILAE Classification of the Epilepsies

- Simplified the framework
- Etiology – consider at all stages
- Developmental and/or Epileptic Encephalopathies
- Self-limited, pharmacoresponsive
- Genetic Generalized Epilepsies
  - Idiopathic Generalized Epilepsies = CAE, JAE, JME, GTCA
- Symptomatic Generalized Epiliepsies used for both
  - Developmental and Epileptic Encephalopathies
  - (static) Encephalopathy with Epilepsy

Impact on Clinical Care and Practice

- New classification framework will
  - Change the approach to diagnosis in the clinic
  - Be applied to patients and guide management
- Updates terminology to reflect current thinking
  - Scientific advances

ILAE Classification Task Force 2013-7

Torbjörn Tomson, Emilio Perucca, Ingrid Scheffer, Jackie French, Yue-Hua Zhang
Satish Jain, Gary Mathern, Sam Wiebe, Edouard Hirsch, Sameer Zuberi, Nico Moshe