

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

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

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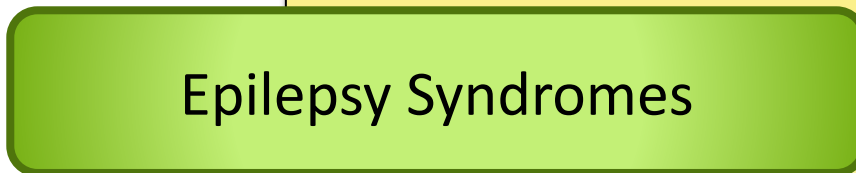
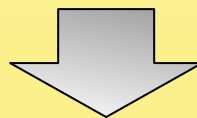
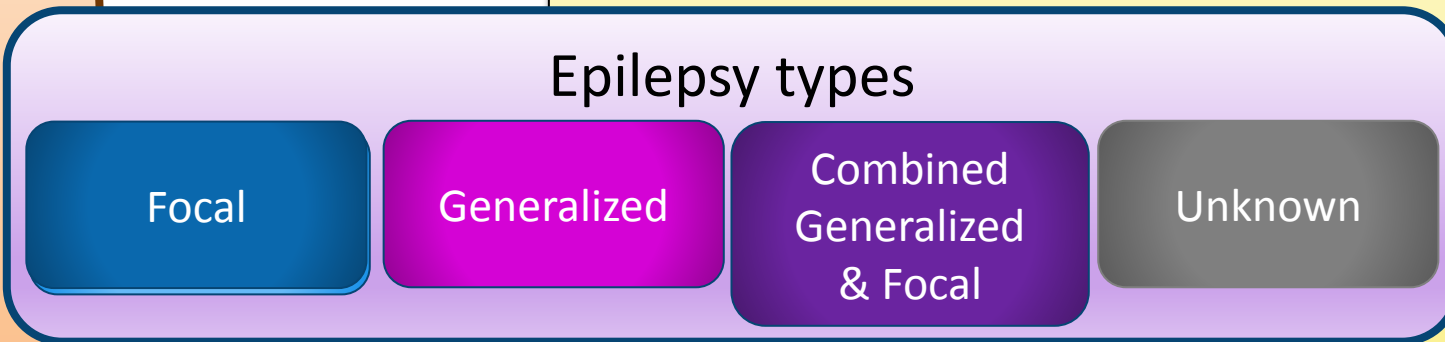
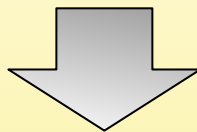
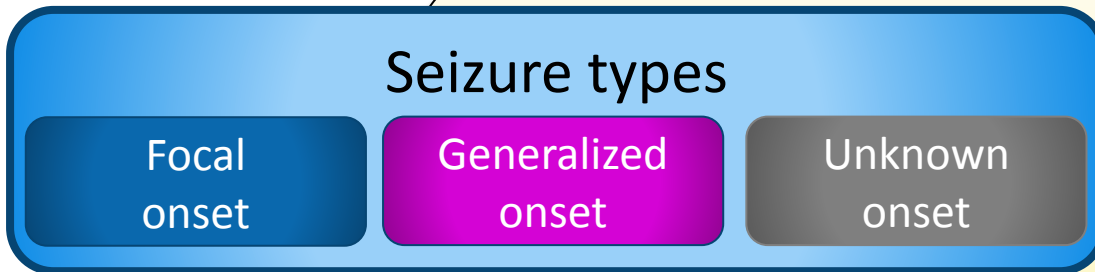
Classification of the Epilepsies

Purpose: for clinical diagnosis

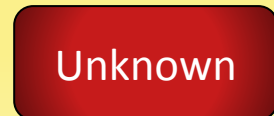


Transparent language: use words that mean what they say

Co-morbidities



Etiology



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

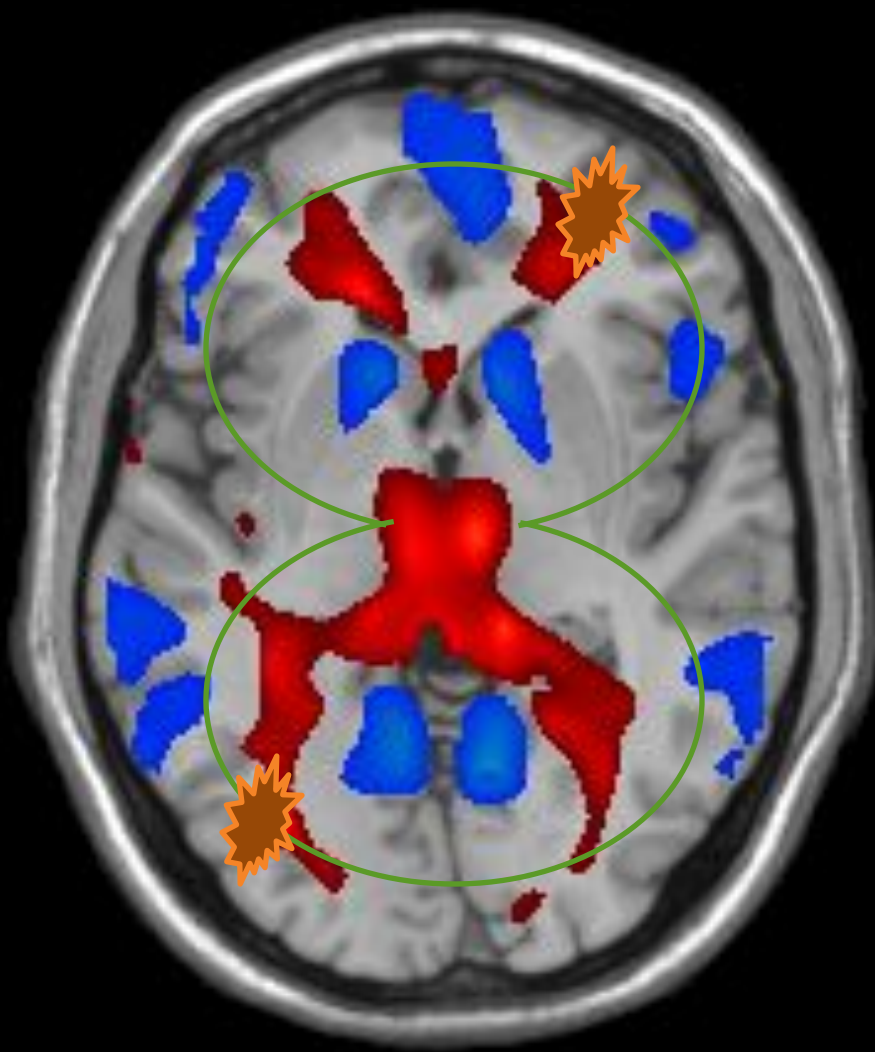
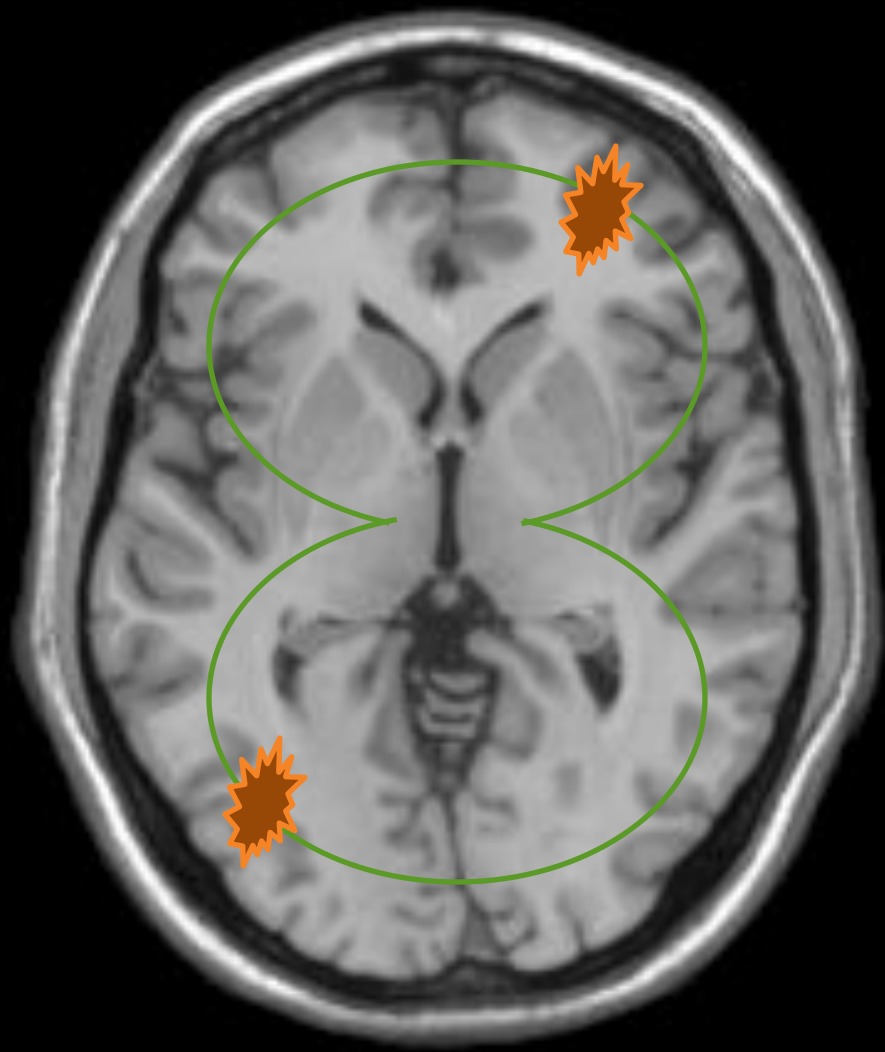
Generalized
onset

Unknown
onset

Generalized seizures

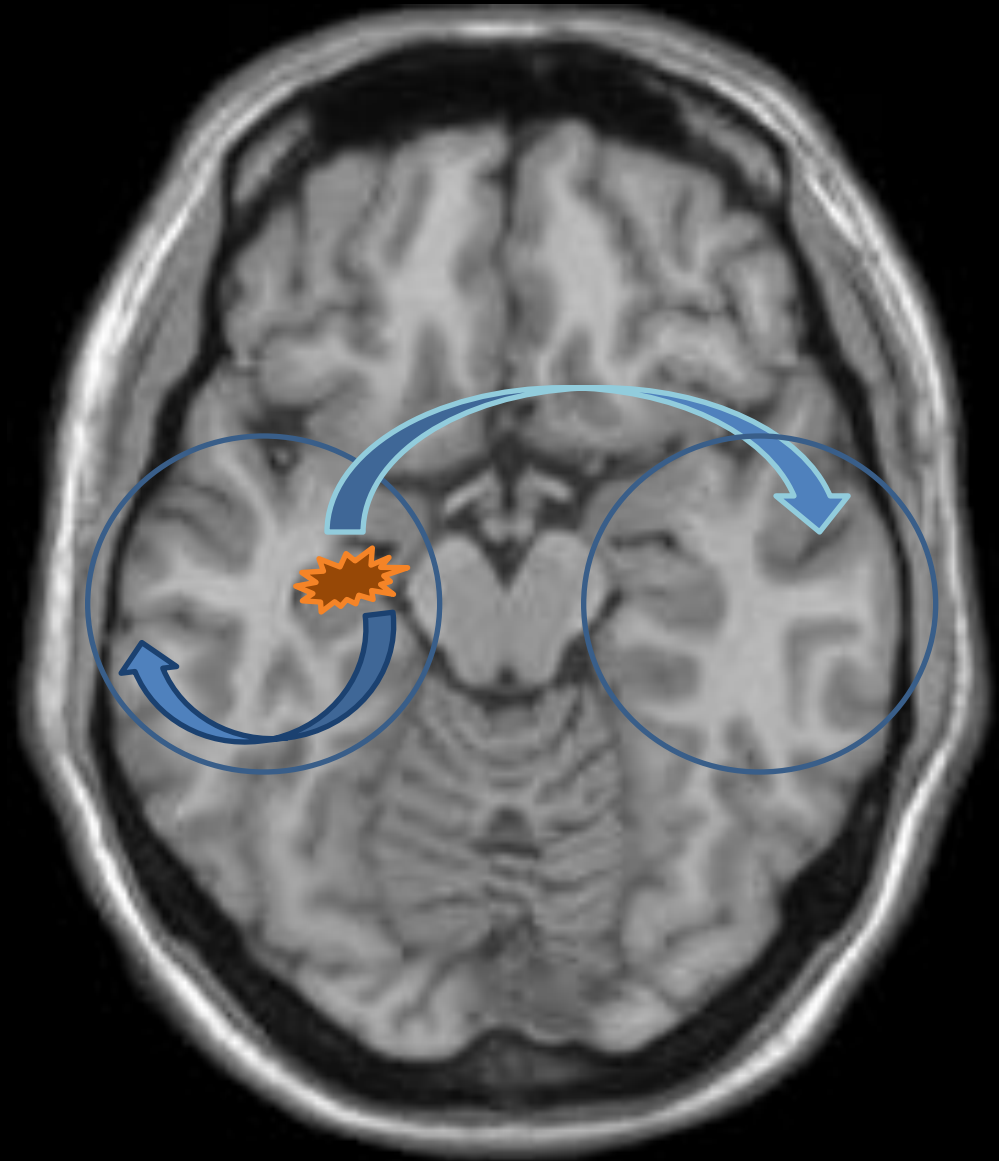
- Originate at some point within and rapidly engage bilaterally distributed networks
- Can include cortical and subcortical structures but not necessarily the entire cortex



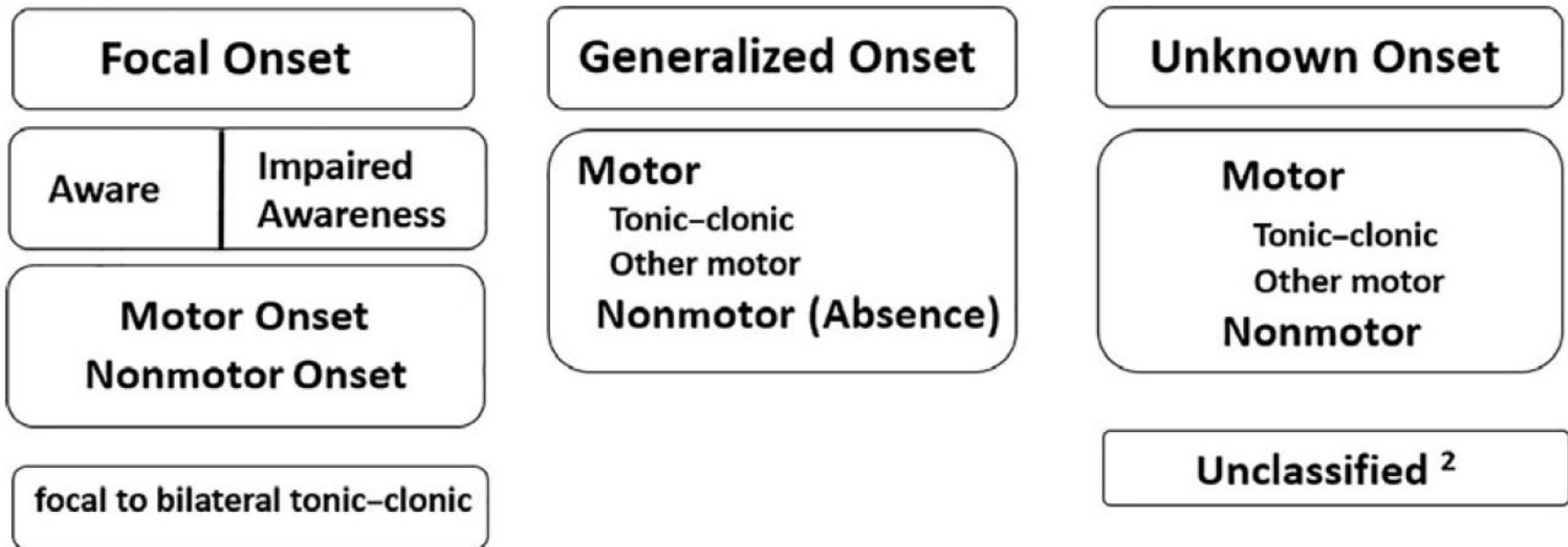


Focal seizures

- 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

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

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

Generalized Onset

Motor

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

Nonmotor (absence)

typical
atypical
myoclonic
eyelid myoclonia

focal to bilateral tonic-clonic

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

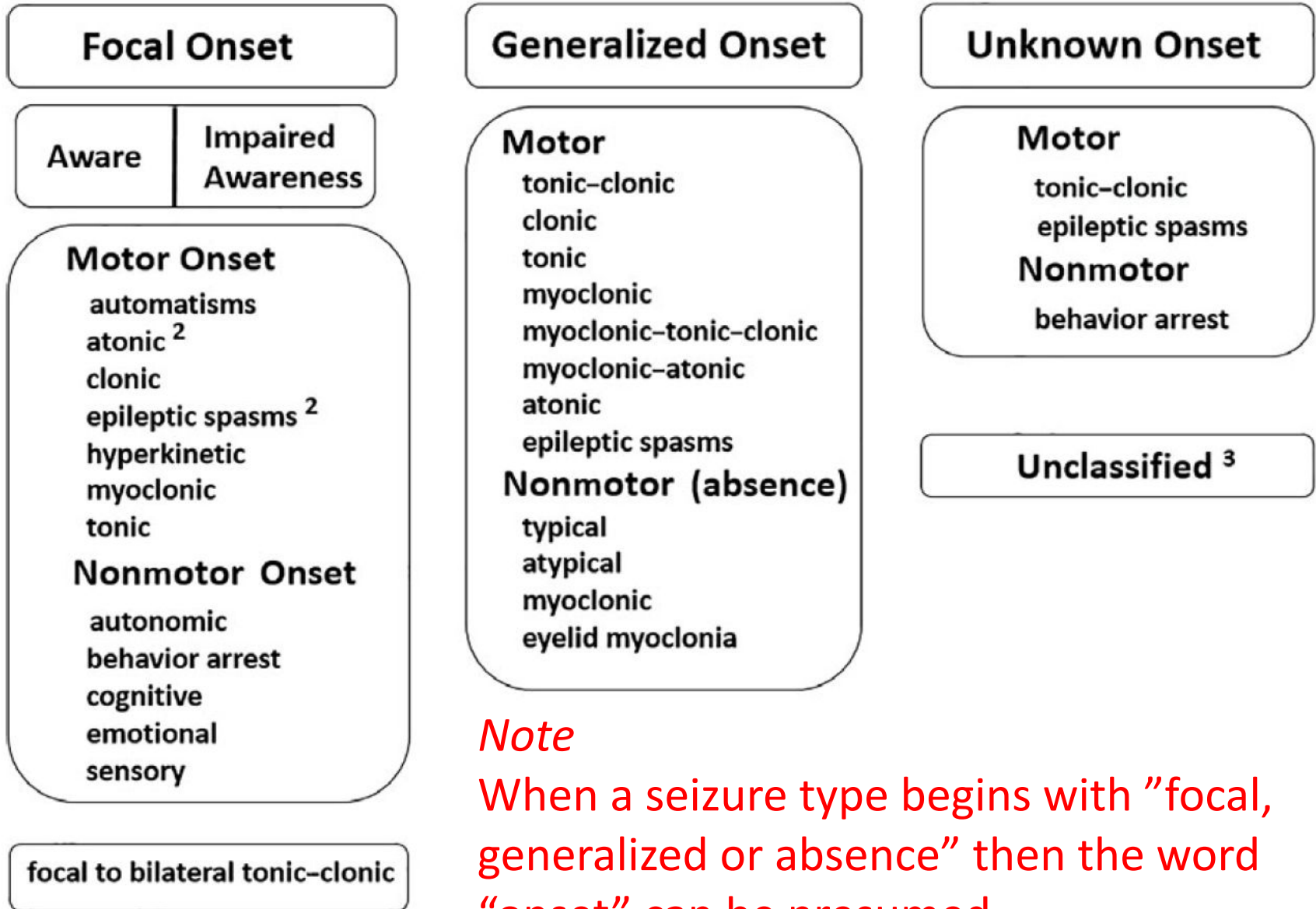
tonic-clonic
epileptic spasms

Nonmotor

behavior arrest

Unclassified ³

ILAE 2017 Classification of Seizure Types Expanded Version ¹



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

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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

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 from Ref. 12

Ins

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

Table 3.

Old term for

Absence

Absence, atypical

Absence, typical

Akinetic

Astatic

Atonic

Aura

Clonic

Complex partial

Convulsion

Dacrystic

Dialeptic

Drop attack

Fencer's posture

(asymmetric tonic)

Figure-of-4

clonic], focal to bilateral tonic-clonic

Focal [aware or impaired awareness]

emotional (dacrystic)

Focal impaired awareness

[Focal/generalized] atonic,

[focal/generalized] tonic

Focal [aware or impaired awareness]

motor tonic

Focal [aware or impaired awareness]

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

the following conditions: (1) At least two unprovoked seizures; (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. The diagnosis of epilepsy is not resolved for individuals who had an age-dependent

Summary of rules for

- 1 Onset: Decide whether the seizure is generalized, focal, or focal to bilateral tonic-clonic. If the onset is unclear, the seizure is classified as focal impaired awareness.
- 2 Awareness: Decide whether the patient is aware during the seizure. If the patient is not aware, the seizure is classified as focal impaired awareness or focal to bilateral tonic-clonic.
- 3 Impaired awareness: Decide whether the patient is aware during the seizure. If the patient is not aware, the seizure is classified as focal impaired awareness or focal to bilateral tonic-clonic.
- 4 Onset pattern: Decide whether the seizure is focal or focal to bilateral tonic-clonic. If the onset is unclear, the seizure is classified as focal impaired awareness or focal to bilateral tonic-clonic.
- 5 Behavior: Decide whether the patient is aware during the seizure. If the patient is not aware, the seizure is classified as focal impaired awareness or focal to bilateral tonic-clonic.
- 6 Motor/non-motor: Decide whether the seizure is focal or focal to bilateral tonic-clonic. If the onset is unclear, the seizure is classified as focal impaired awareness or focal to bilateral tonic-clonic.

Adapted from Ref. 12

New 12

12

Adapted from Ref. 12

New New

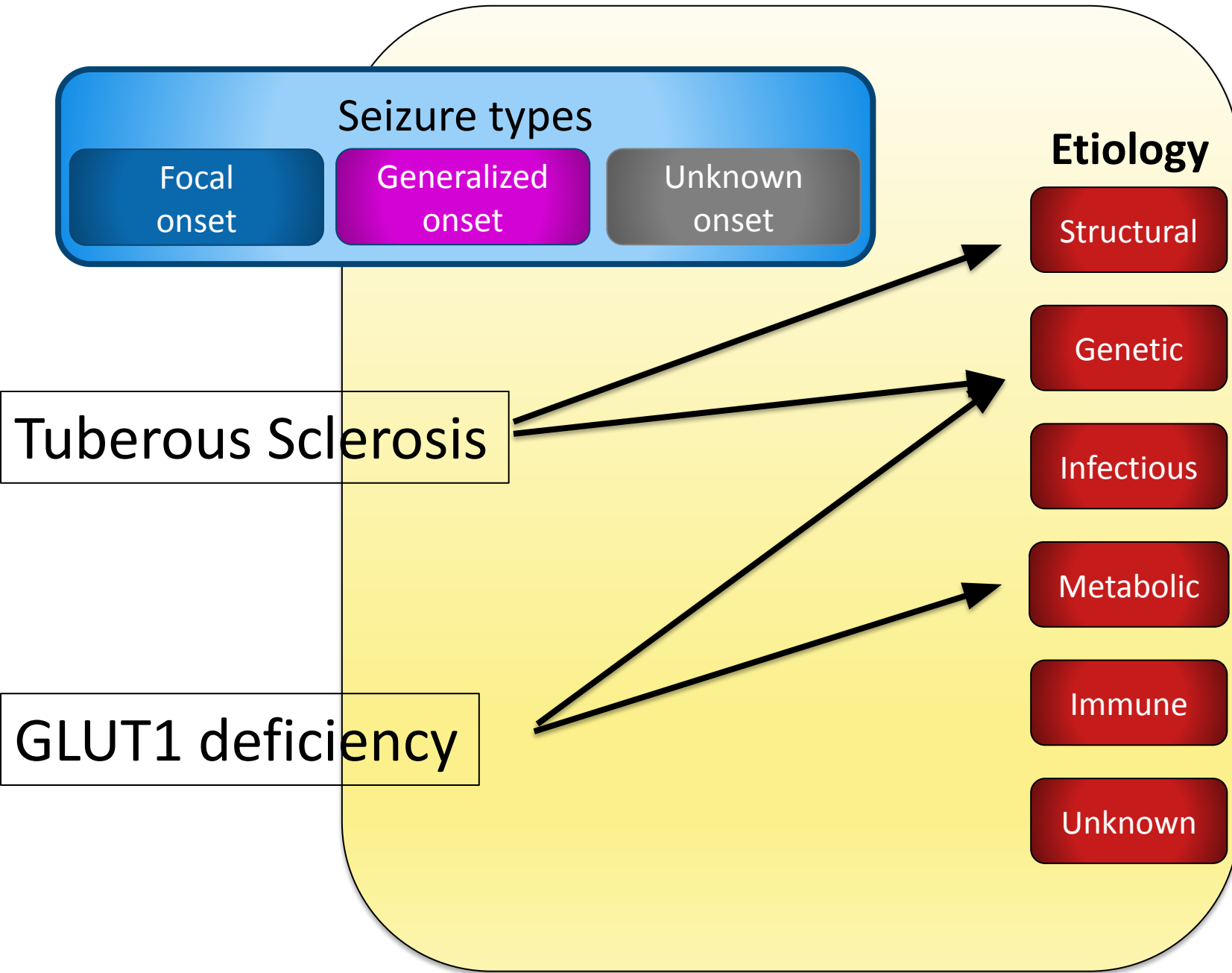
Adapted from Ref. 12

New

12 Adapted from Ref. 12

Adapted from Ref. 12

3



Seizure types

Focal onset

Generalized onset

Unknown onset

Etiology

Structural

Genetic

Infectious

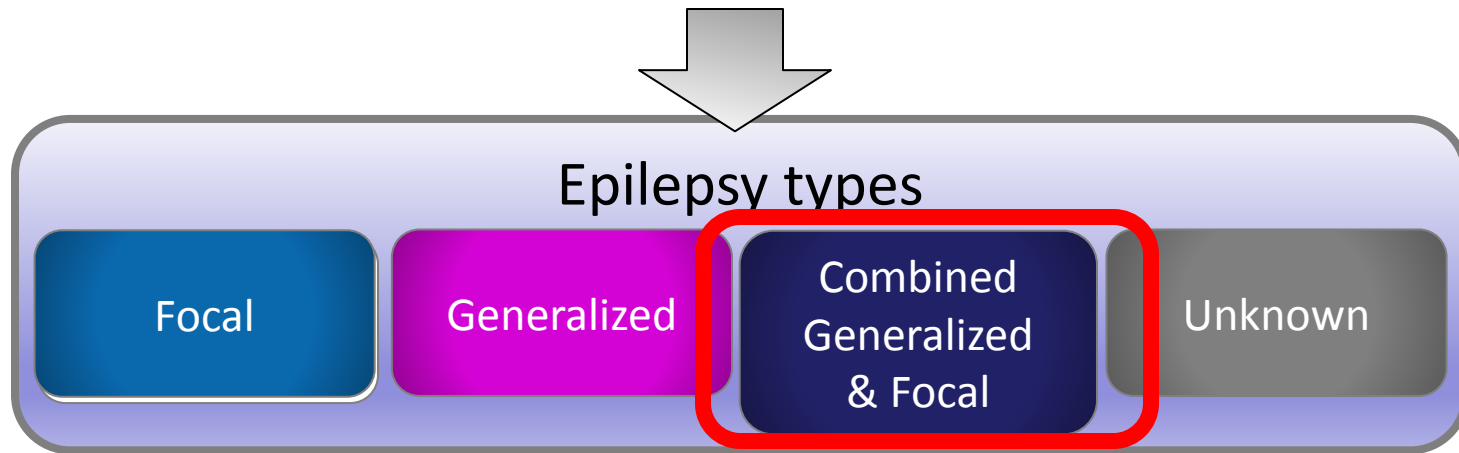
Metabolic

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

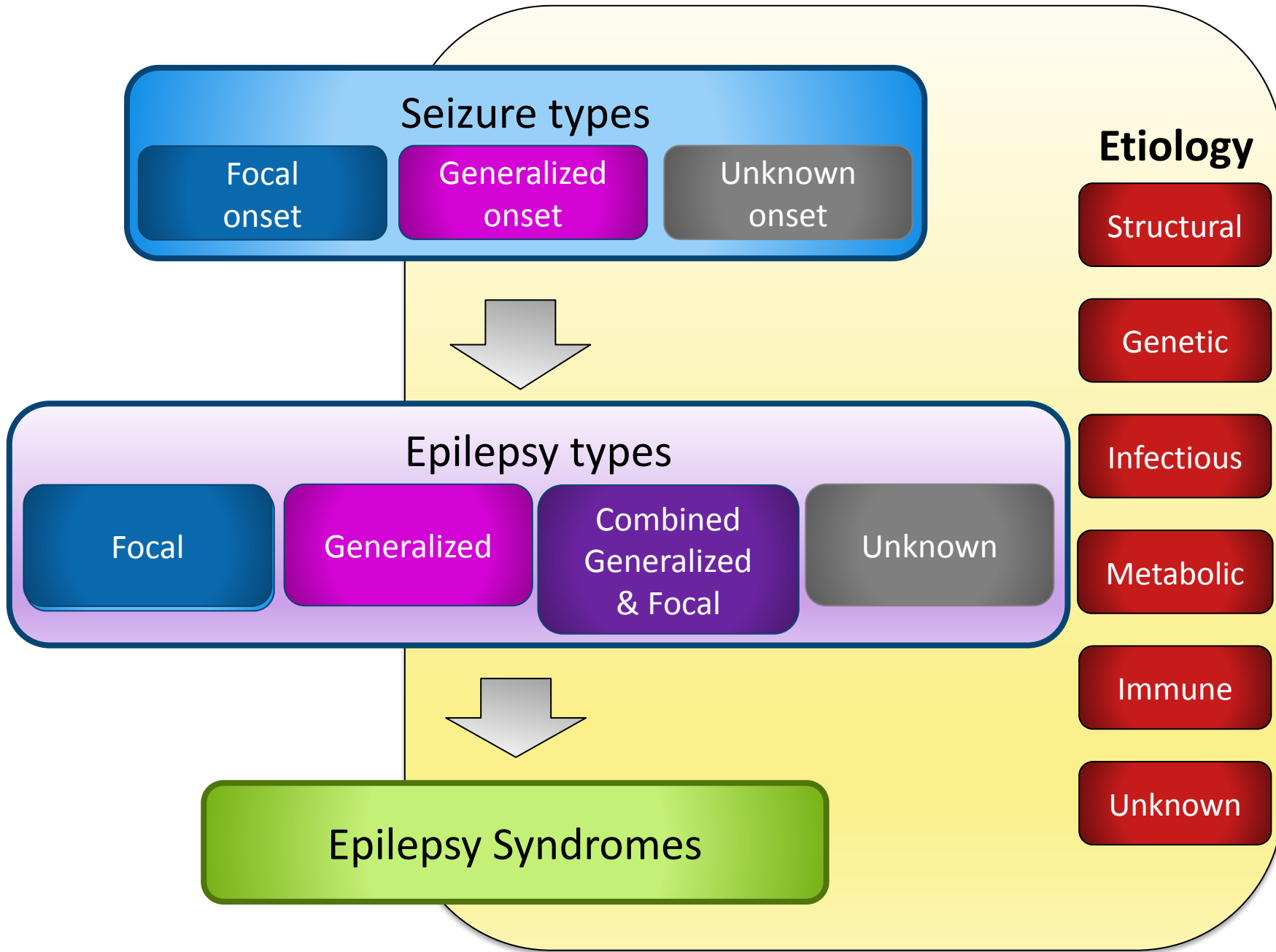
Examples

- Dravet syndrome

- What do with

- Multifocal epilepsies? focal

- Hemispheric epilepsies? focal



Seizure types

Focal onset

Generalized onset

Unknown onset

Etiology

Structural

Genetic

Infectious

Metabolic

Immune

Unknown

Epilepsy types

Focal

Generalized

Combined
Generalized
& Focal

Unknown

Epilepsy Syndromes

Old term

'Idiopathic Generalized Epilepsies'

Idiopathic Generalized Epilepsies

Childhood
Absence
Epilepsy

Juvenile
Absence
Epilepsy

Juvenile
Myoclonic
Epilepsy

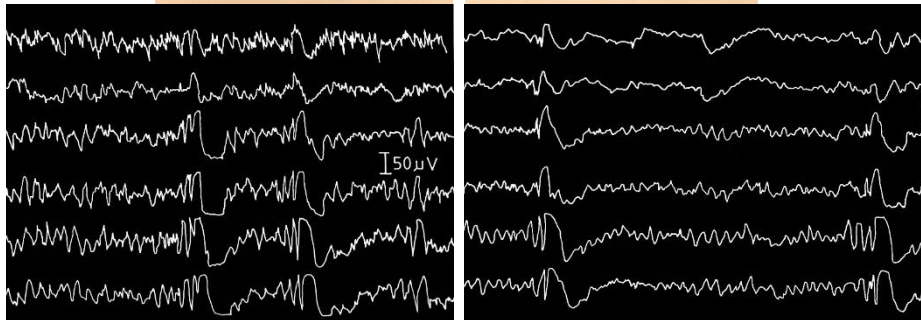
Generalized
Tonic-Clonic
Seizures Alone

Genetic *versus* idiopathic

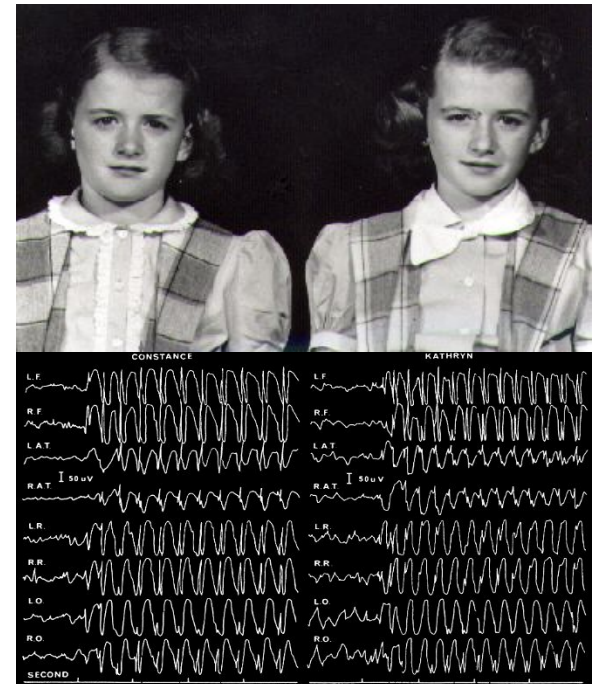
- 'Idiopathic' = presumed hereditary predisposition
- Genetic \neq inherited
 - Importance *of de novo* mutations in both mild and severe epilepsies
- Critical problem of stigma in some parts of the world

Genetic \neq Gene testing

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

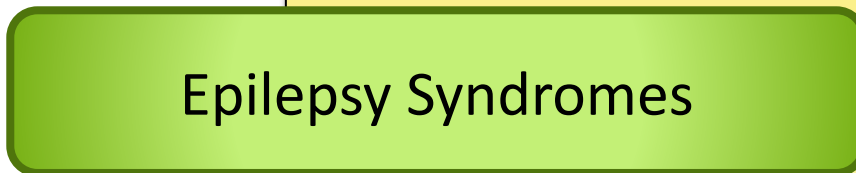
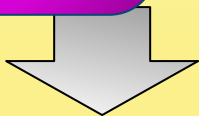
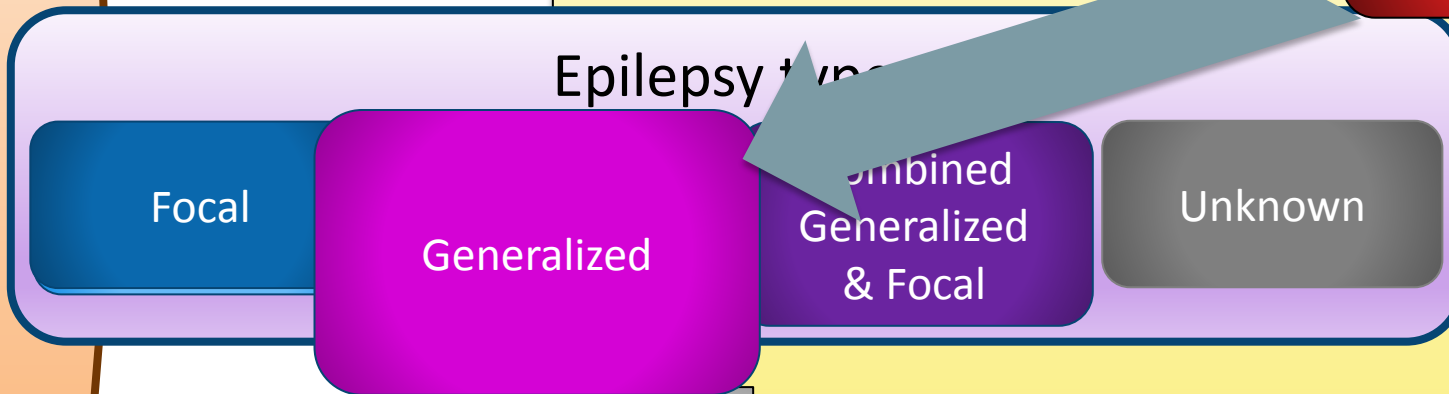
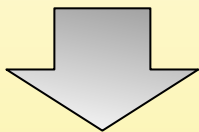
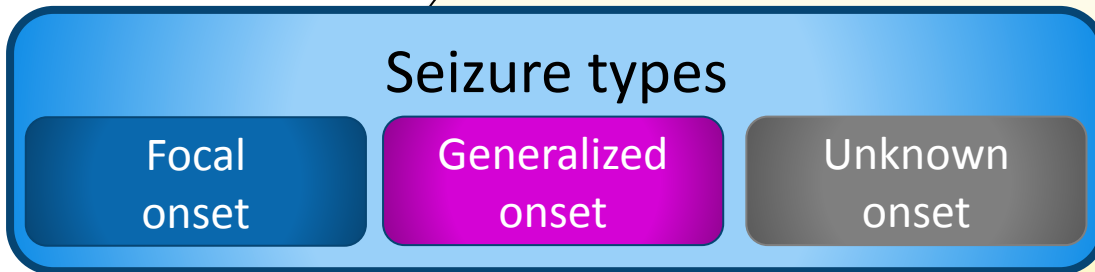


JME pair; Lennox 1941



CAE pair; Lennox 1950

Co-morbidities



Etiology

Structural

Genetic

Infectious

Metabolic

Immune

Unknown

Epilepsy syndromes

- There are **no** approved ILAE epilepsy syndromes

<https://www.epilepsydiagnosis.org>



International League Against Epilepsy
Working toward a world where no person's life is limited by epilepsy



> **Overview**

Log In For Videos

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 >

EpilepsyDiagnosis.org

The ILAE Commission on Classification and Terminology welcomes you to EpilepsyDiagnosis.org, a cutting edge online diagnostic manual of the epilepsies.

Goal

The goal of ***epilepsydiagnosis.org*** is to make available, in an easy to understand form, latest concepts relating to seizures and the epilepsies. The principle goal is to assist clinicians who look after people with epilepsy anywhere in the world to diagnose seizure type(s), classify epilepsy, diagnose epilepsy syndromes and define the etiology of the epilepsy. The site is principally designed for clinicians in primary and secondary care settings caring for people with epilepsy and we hope will also serve as a useful teaching aid.

Structure

The structure of this site reflects the importance of seizure type, syndrome, and etiology in clinical practice. On this website, you will find current classification concepts for seizures, with their clinical features, video examples, EEG correlate, differential diagnosis and related epilepsy syndromes. Epilepsy syndromes are detailed by their clinical features, seizure types, EEG, imaging and genetic correlates and differential diagnoses. The site includes sections on etiologies of epilepsies and epilepsy imitators with cross-referencing between these sections and seizure and syndrome sections.

Definition of epilepsy

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

- At least two unprovoked (or reflex) seizures occurring more than 24 hours apart



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

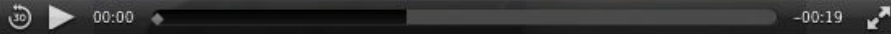
Two absence seizures occur, with the eyes opening, staring and 3 Hz eyelid flutter associated with the events. GSW appearing on the video as it plays denotes when generalized spike-and-wave appears on the patient's EEG.



International League Against Epilepsy
Working toward a world where no person's life is limited by epilepsy



Typical Absence Seizure Video 1



▶ 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 patient's EEG.

> Generalized seizure

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 ▶

Welcome *scheffer!*

Logout

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. Hypnagogic 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

R09:25:12

Fp2-F8

F8-T4

T4-T6

T6-O2

Fp1-F7

F7-T3

T3-T5

T5-O1

Fp2-F4

F4-C4

C4-P4

P4-O2

Fp1-F3

F3-C3

C3-P3

P3-O1

**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

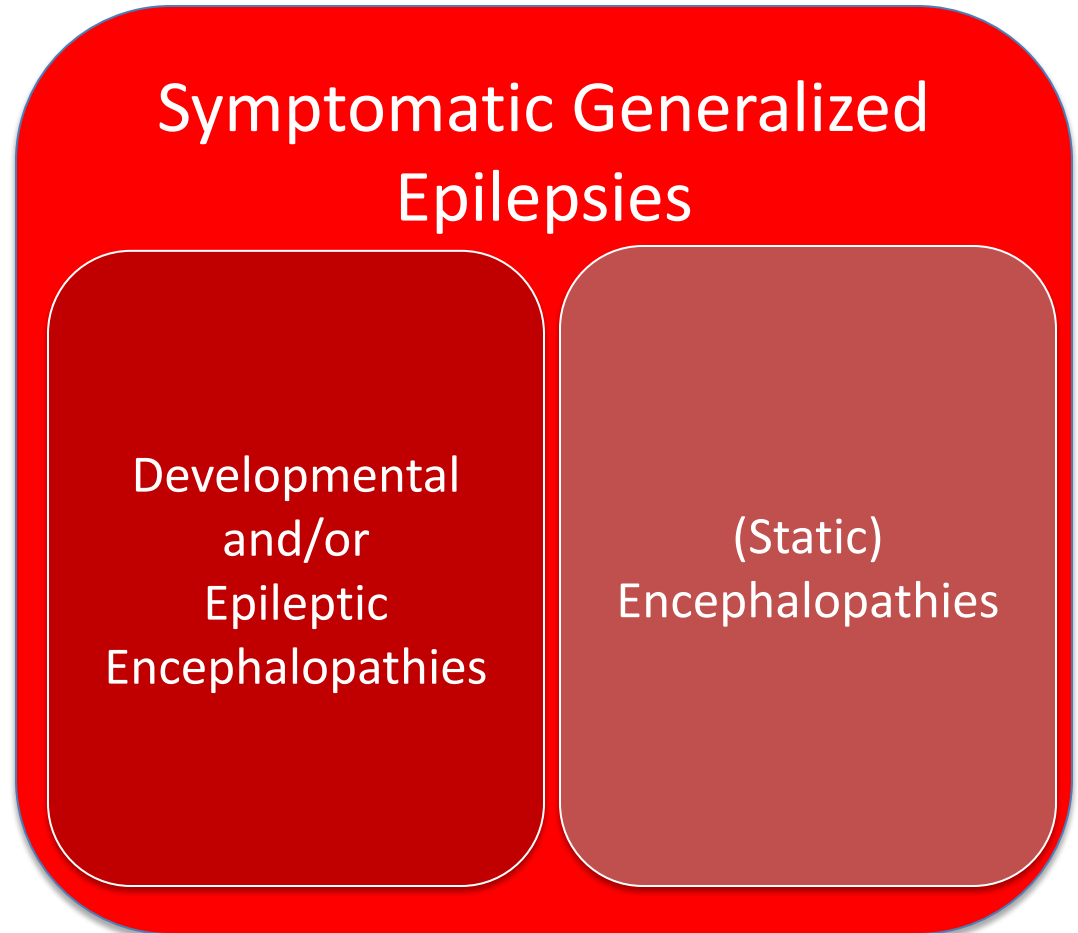
Developmental *and/or* Epileptic Encephalopathy

- 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 Epilepsies 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
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