

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

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

*Epilepsia*, \*\*(\*):1–10, 2017

doi: 10.1111/epi.13709

## **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

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# Classification of the Epilepsies

**Purpose: for clinical diagnosis**



Transparent language: use words that mean what they say

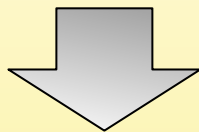
Co-morbidities

## Seizure types

Focal  
onset

Generalized  
onset

Unknown  
onset



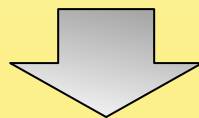
## Epilepsy types

Focal

Generalized

Combined  
Generalized  
& Focal

Unknown



Epilepsy Syndromes

## Etiology

Structural

Genetic

Infectious

Metabolic

Immune

Unknown

## 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

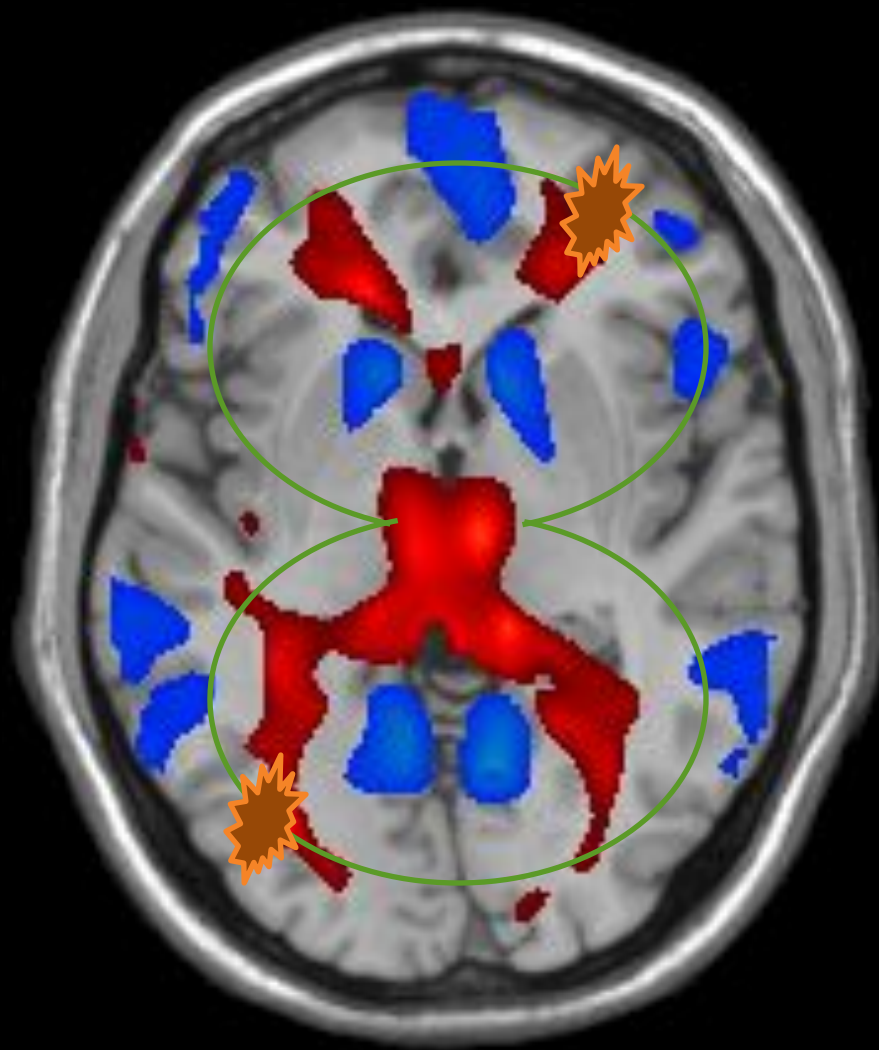
Unknown  
onset

# 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

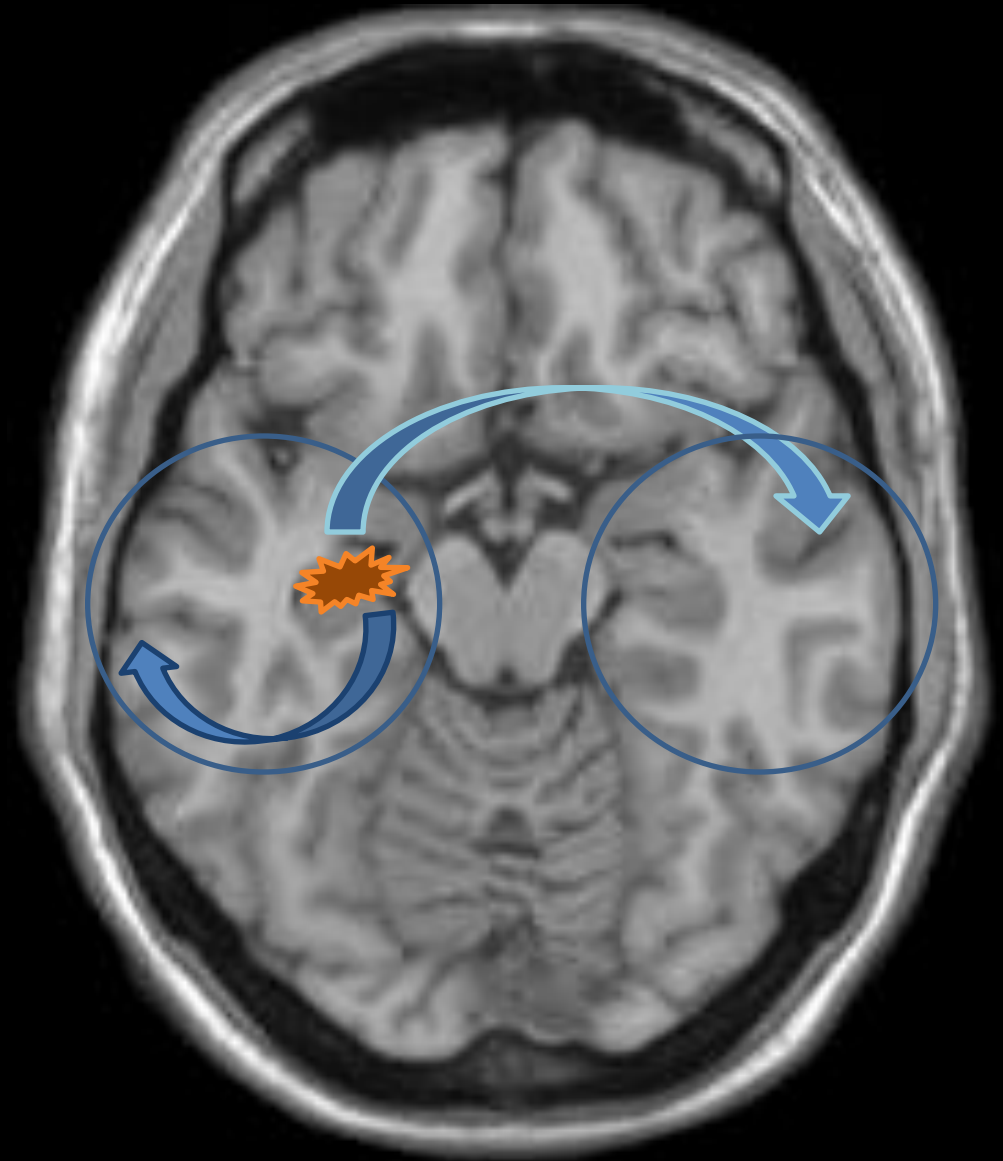






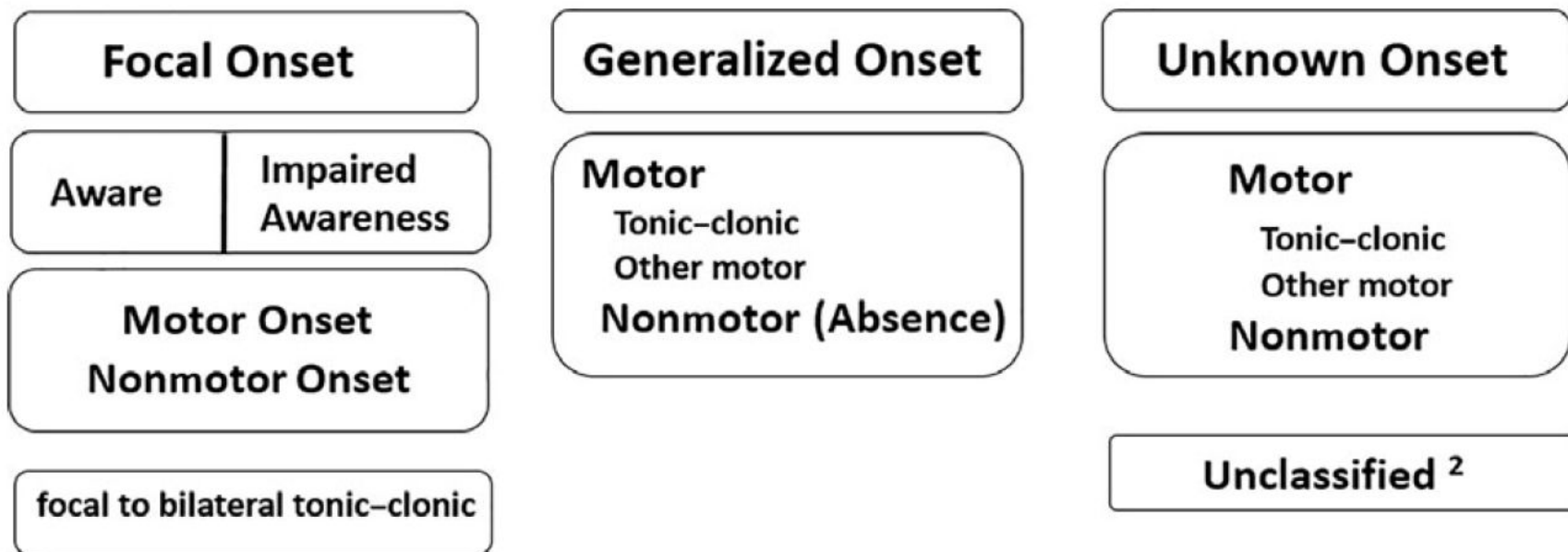
# Focal seizures

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

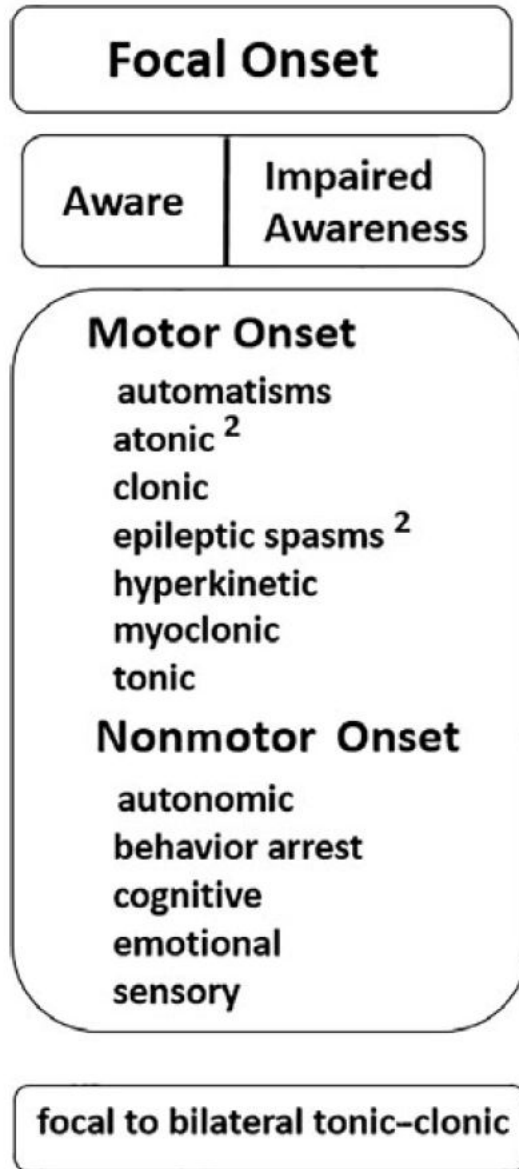




## ILAE 2017 Classification of Seizure Types Basic Version <sup>1</sup>



# ILAE 2017 Classification of Seizure Types Expanded Version <sup>1</sup>



## 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 <sup>1</sup>

## Focal Onset

Aware

Impaired  
Awareness

### Motor Onset

automatisms  
atonic <sup>2</sup>  
clonic  
epileptic spasms <sup>2</sup>  
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 <sup>1</sup>

## Focal Onset

Aware

Impaired  
Awareness

### Motor Onset

automatisms  
atonic <sup>2</sup>  
clonic  
epileptic spasms <sup>2</sup>  
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

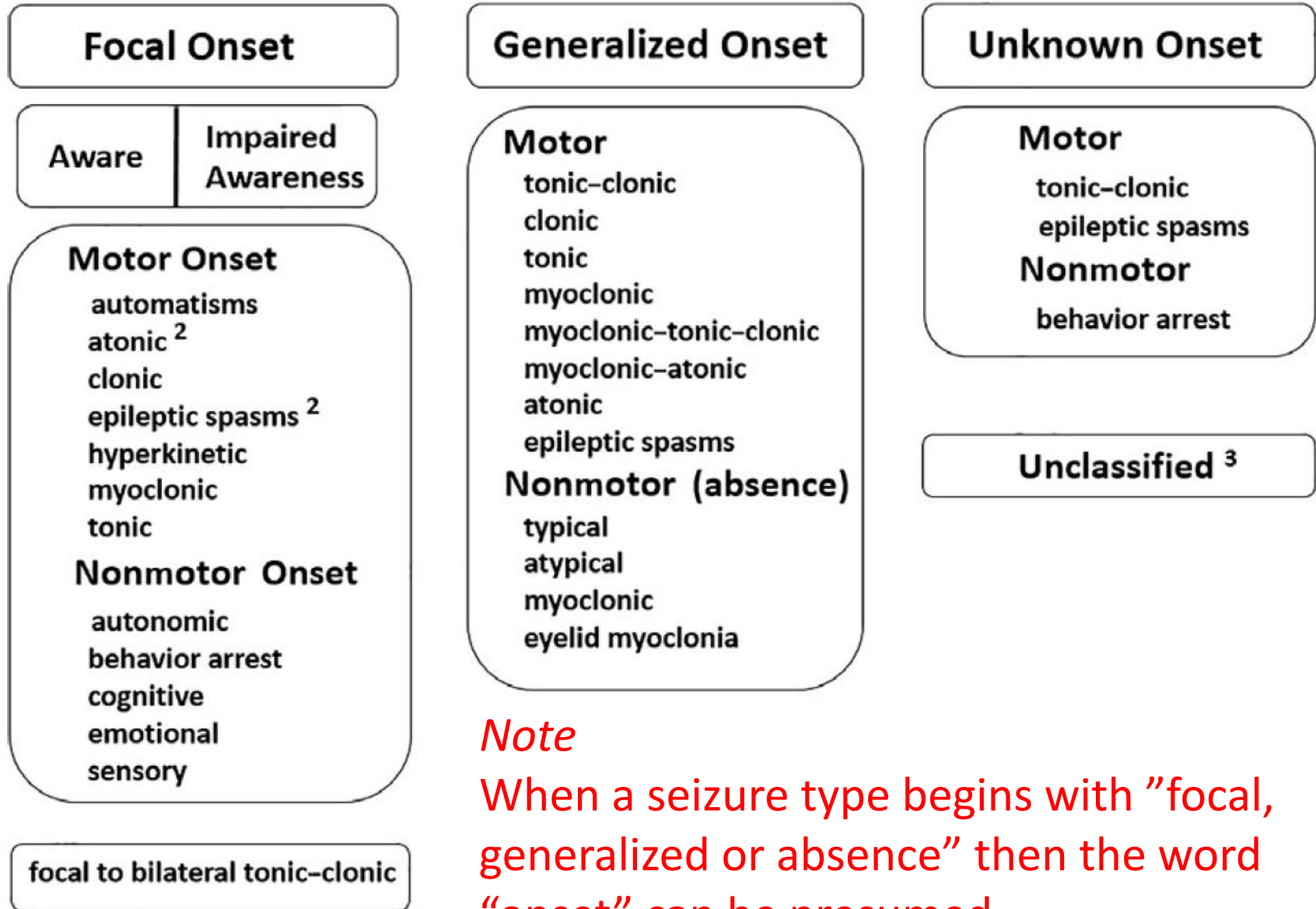
tonic-clonic  
epileptic spasms

### Nonmotor

behavior arrest

### Unclassified <sup>3</sup>

# ILAE 2017 Classification of Seizure Types Expanded Version <sup>1</sup>



## Note

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



# Terms no longer in use

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



# **Instruction manual for the ILAE 2017 operational classification of seizure types**

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

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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

Dialectic

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]

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  
part; (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

Adapted from  
Ref. 12

New

Ref. 12

12

New

New

Adapted from

New

New

Ref. 12

Adapted from

12

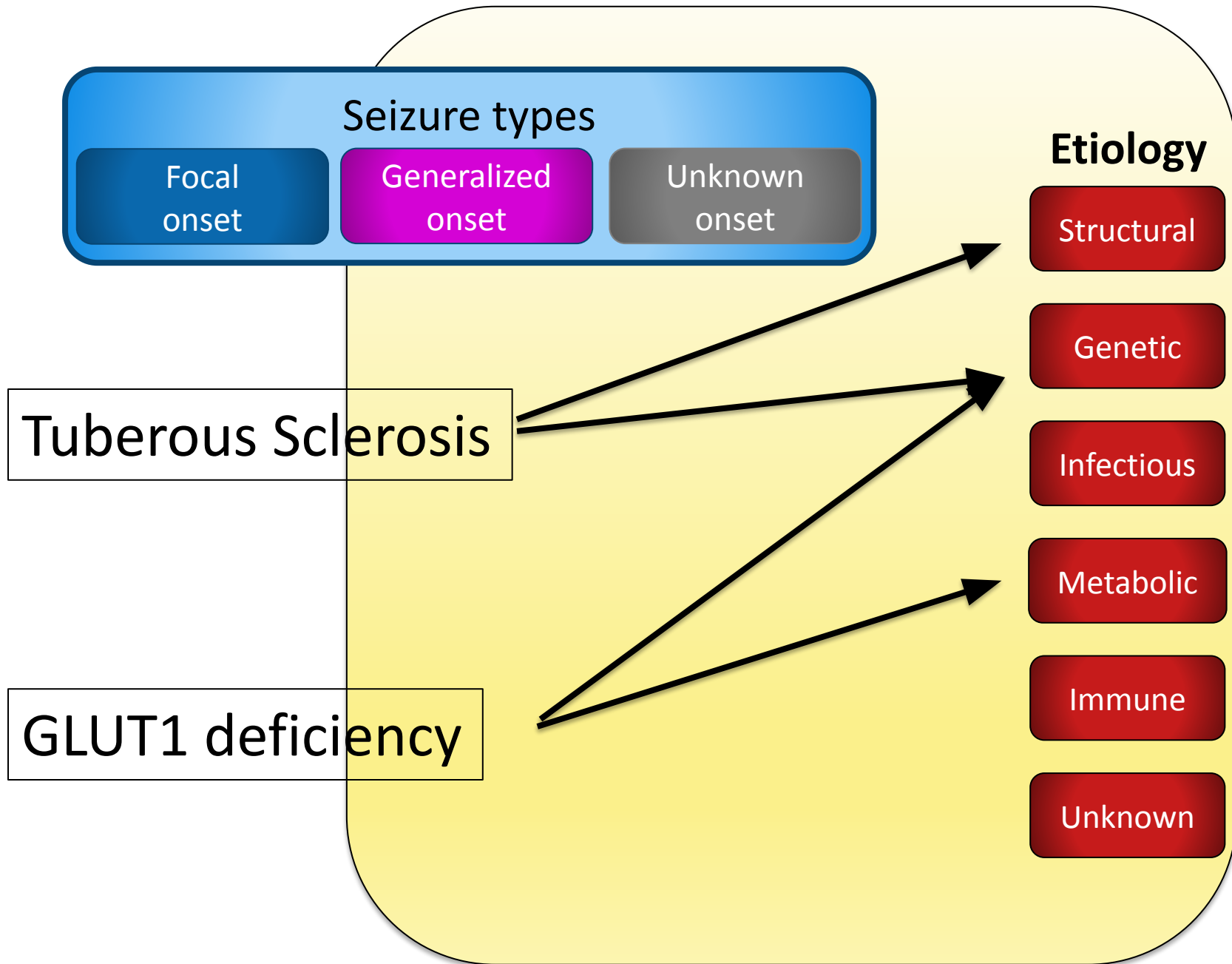
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New

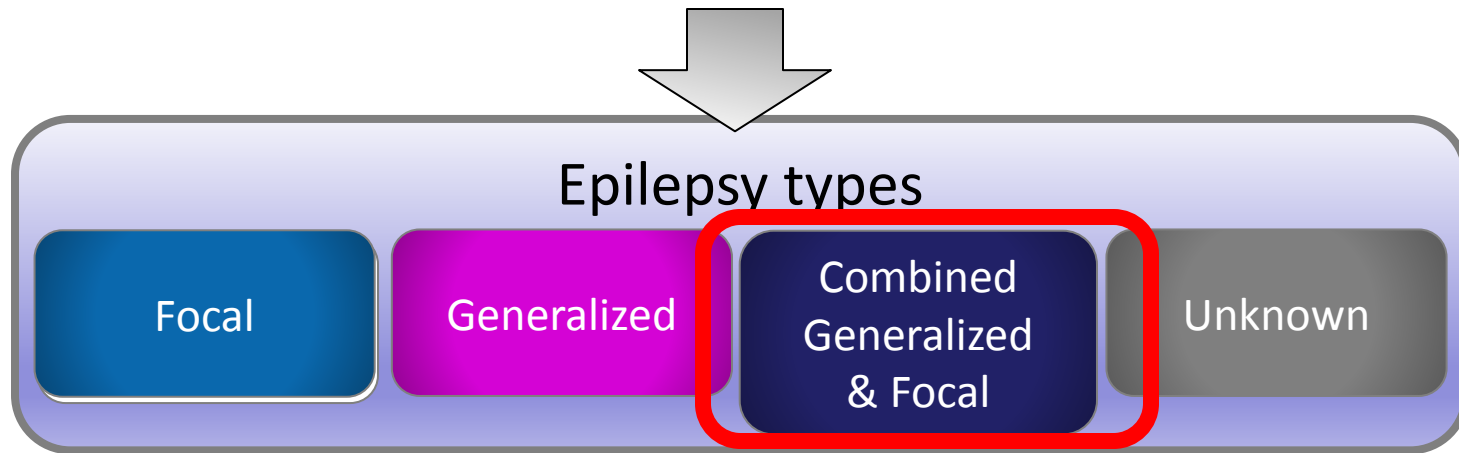
Ref. 12

Adapted from

12







- 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

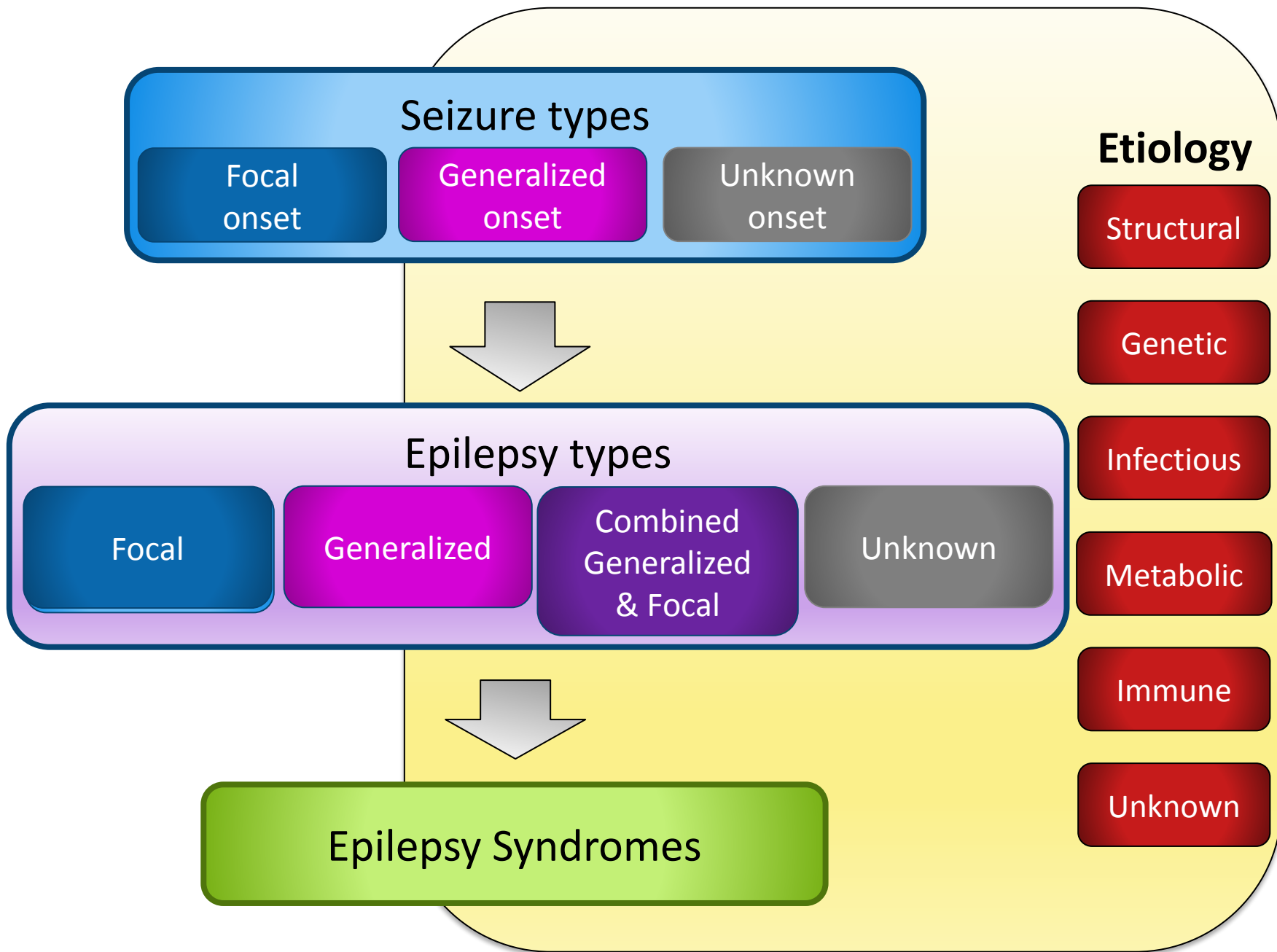
Examples

- Dravet syndrome

- What do with

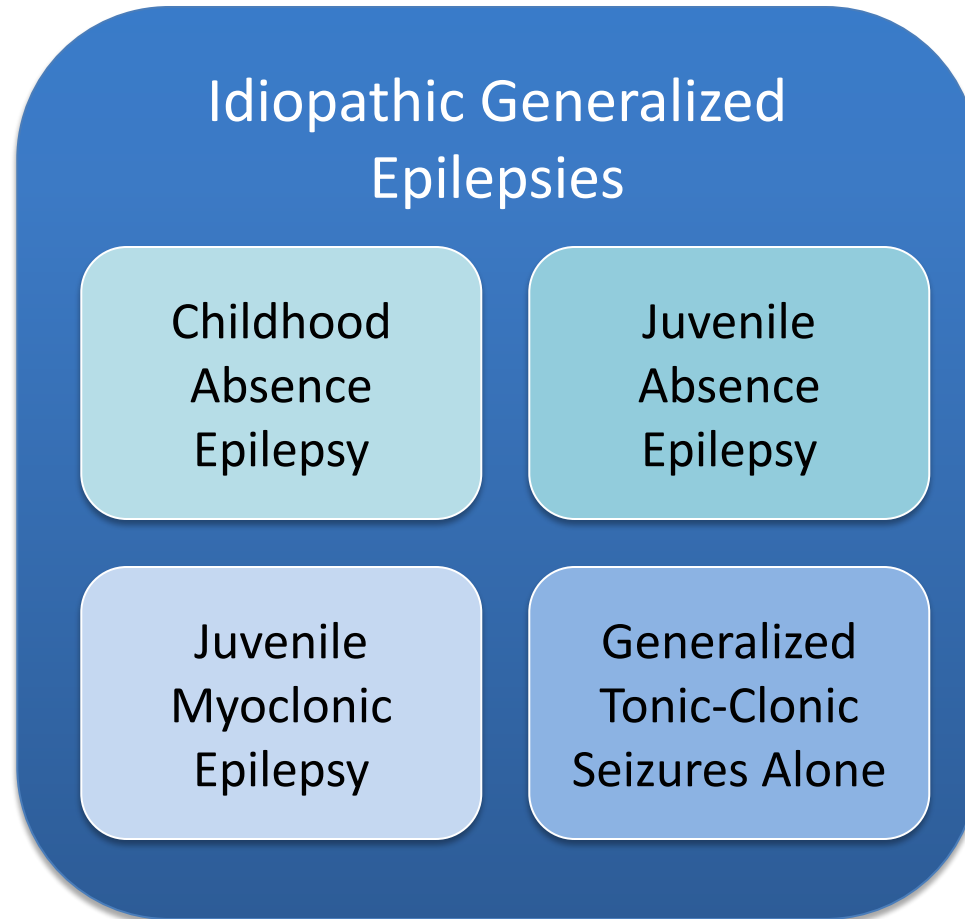
- Multifocal epilepsies? ☐ focal

- Hemispheric epilepsies? ☐ focal



# Old term

## 'Idiopathic Generalized Epilepsies'



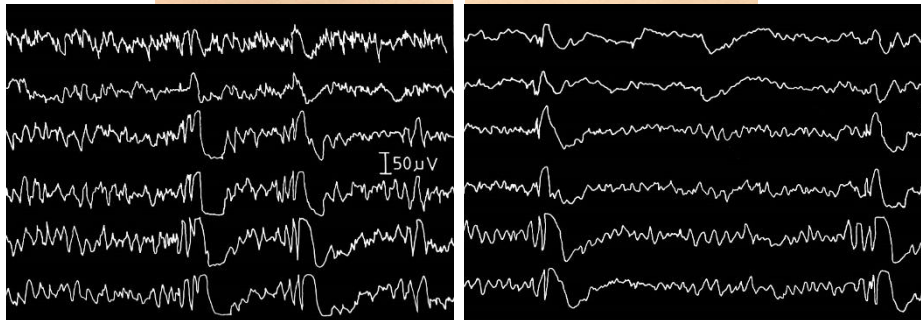
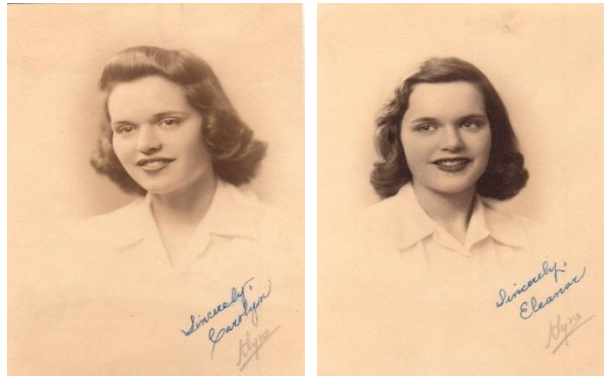
# Genetic *versus* idiopathic

- '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

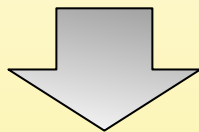
Co-morbidities

## Seizure types

Focal  
onset

Generalized  
onset

Unknown  
onset



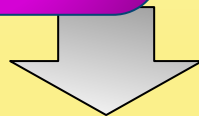
## Epilepsy types

Focal

Generalized

Combined  
Generalized  
& Focal

Unknown



Epilepsy Syndromes

## Etiology

Structural

Genetic

Infectious

Metabolic

Immune

Unknown



# 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**

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**Overview**

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**Seizure Classification**

[Generalized seizures](#)

[Focal seizures](#)

[Focal/Generalized](#)

**Epilepsy syndromes**

[Neonatal/Infantile](#)

[Childhood](#)

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[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



## > Generalized seizure

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### Overview

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### 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

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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



## > Generalized seizure

### Overview

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### Seizure Classification

**Generalized seizures**

[Focal seizures](#)

[Focal/Generalized](#)

### Epilepsy syndromes

[Neonatal/Infantile](#)

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[Adolescent/Adult](#)

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### Epilepsies by Etiology

[Genetic](#)

[Structural](#)

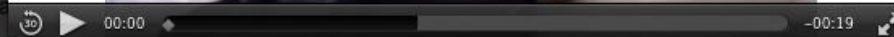
[Metabolic](#)

[Immune](#)

[Infectious](#)

[Unknown](#)

### 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.

Welcome *scheffer*!

[Logout](#)

## > Epilepsy Imitators

### 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

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- 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

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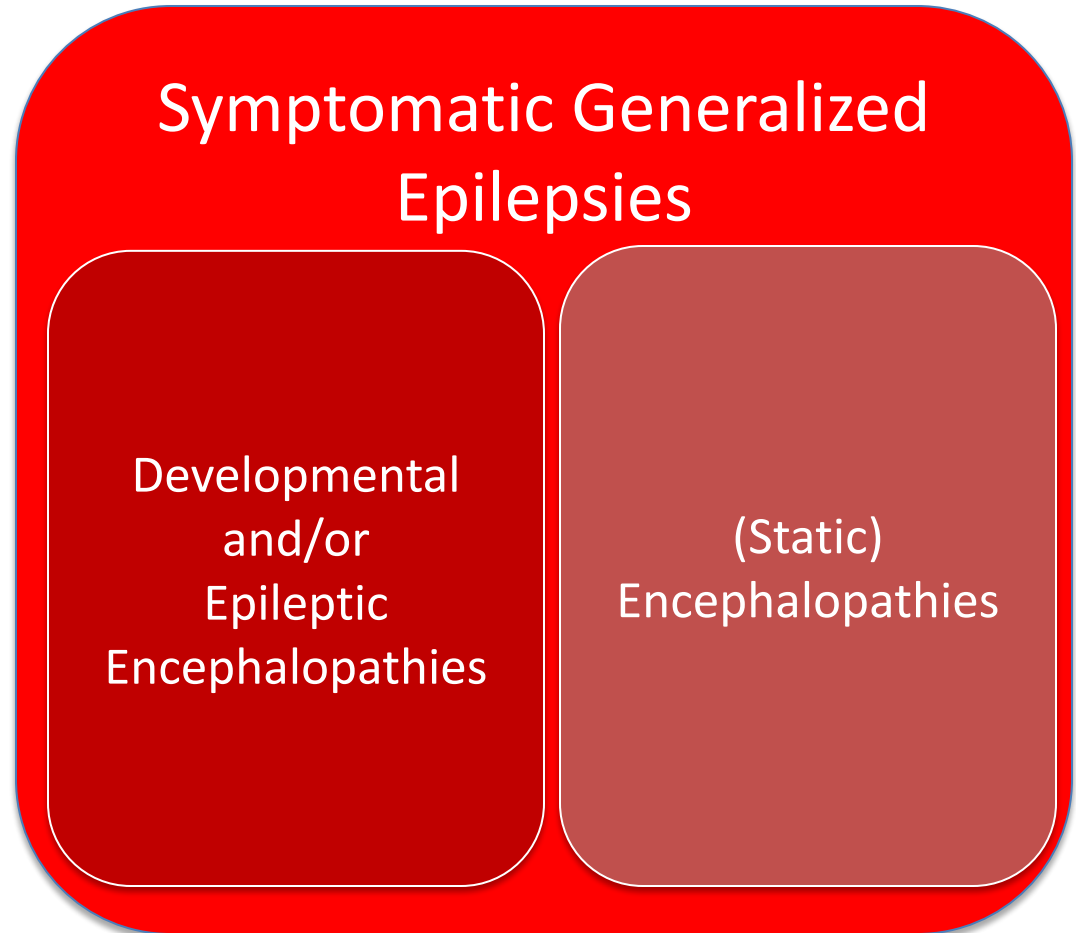
- Developmental encephalopathy
  - May begin in utero
  - Post birth
- Epileptic encephalopathy
  - Can occur at any age
  - May have remediable component – right vs wrong AED
- Move towards *GENE* encephalopathy
  - eg. *CDKL5* encephalopathy, *SCN2A* encephalopathy

# Old terms

## ‘Symptomatic Generalized Epilepsies’

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- Used for two different groups of disorders





# ILAE Classification of the Epilepsies

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- 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

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- 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