Types of Seizures and Common Epilepsy Syndromes in Children

February 26, 2019

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American Academy of Pediatrics
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Types of Seizures

- Generalized
  - Absence, myoclonic, clonic, tonic, tonic-clonic, atonic and combinations thereof
- Partial
  - Simple partial
  - Complex partial
  - Secondary generalized
- Unclassified epileptic seizures

Classification of Epilepsy

[ILAE 1989]

- Localization related
- Generalized epilepsies and syndromes
- Undetermined whether focal or generalized
- Special Syndromes

Epilepsia, 30(4):389-399, 1989
Raven Press, Ltd., New York

Proposal for Revised Classification of Epilepsies and Epileptic Syndromes

Commission on Classification and Terminology of the International League Against Epilepsy

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Idiopathic Focal

Focal Seizures

Normal intellect/PE

Normal EEG background

Normal neuroimaging

Good Prognosis

Ex: Benign Rolandic Epilepsy

Symtomatic Focal

Focal seizures

Gross/subtle cognitive and/or exam abnormalities

Abnormal neuroimaging

Variable prognosis

Ex: Temporal lobe epilepsy, Post-traumatic epilepsy

Idoiopathic Generalized

Generalized Seizures

Normal intellect/PE

Normal EEG background

Normal neuroimaging

Good prognosis

Ex: Childhood Absence, Juvenile Myoclonic Epilepsy

Symptomatic Generalized

Generalized seizures (many types)

Subnormal intellect/Abnormal exam

Abnormal EEG background

Abnormal neuroimaging

Poor Prognosis

Ex: Lennox-Gastaut syndrome

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Source: Joshi and Shellhaas 2014

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Problems with the “Old” System

- Difficult to apply the localization related/focal vs. generalized epilepsy paradigm in some cases
  - Eg: infant with infantile spasms who evolves to focal seizures
- Etiology: Cryptogenic or symptomatic
  - New onset focal epilepsy I an otherwise healthy young adult (normal imaging, abnormal EEG)
  - Developmentally delayed patient with generalized seizures (doesn’t fit criteria for LGS), etiology not obvious
- Advances in neuroscience have improved the understanding and etiology of some epilepsies
  - Genetic
  - Neuroimaging

New Organization of Epilepsies

Epilepsia, 51(4):676–685, 2010
doi: 10.1111/j.1528-1167.2010.02522.x

SPECIAL REPORT

Revised terminology and concepts for organization of seizures and epilepsies: Report of the ILAE Commission on Classification and Terminology, 2005–2009

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Etiology: New Concepts

Old Terms

- Etiology
  - Idiopathic
  - Symptomatic
  - Cryptogenic
- Localization-related

New Terms

- Etiology
  - Genetic
  - Structural/Metabolic
  - Immune
  - Unknown
- Focal (replaces localization related and partial

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In the New System…

- Terms no longer used
  - Simple partial
  - Complex partial
  - Secondarily generalized
- Replaced by
  - With or without impairment of consciousness/awareness
  - Dyscognitive
  - Evolving to a convulsive seizure

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Some Common Childhood Epilepsy Syndromes

- Childhood Absence Epilepsy
- Benign Focal Epilepsies of Childhood
- Juvenile Myoclonic Epilepsy
- Infantile Spasms/West Syndrome
- Lennox Gastaut Syndrome

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Epilepsy Syndrome

- Diagnosed based on:
  - History (type of seizure/s, age of onset, personal & family history)
  - Neurological examination (including cognition, development)
  - EEG ± Neuroimaging
  - Etiology (where known)
- Importance of syndrome identification
  - Better understand prognosis
  - Define appropriate work-up and treatment

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Epilepsy Syndrome

- Formerly epilepsy syndromes were classified simply based on type of seizure and whether an etiology was known or not.
- New epilepsy classification considers genetic, metabolic, structural immune etiologies in describing type of epilepsy

Case #1

A 6 year old boy presented with frequent episodes of staring, during which he appeared to be “daydreaming.” The boy would abruptly stop whatever he was doing, stare straight ahead, and was totally unresponsive to verbal mild physical stimulation. The episodes were quite brief, typically lasting 5-10 seconds, and occurred many times per day.

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Diagnosis?

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Childhood Absence Epilepsy

- Age of onset 5-9 years
- Typical absence seizures
  - Simple absence (behavioral arrest & unresponsiveness)
  - Complex
    - With mild clonic or myoclonic component
    - With changes in tone
    - With automatisms
    - With autonomic components
- Intellect and neurological examination normal
  - Mild cognitive abnormalities may be found
- Neuroimaging studies normal (and usually not needed)
- EEG: 3Hz generalized spike wave

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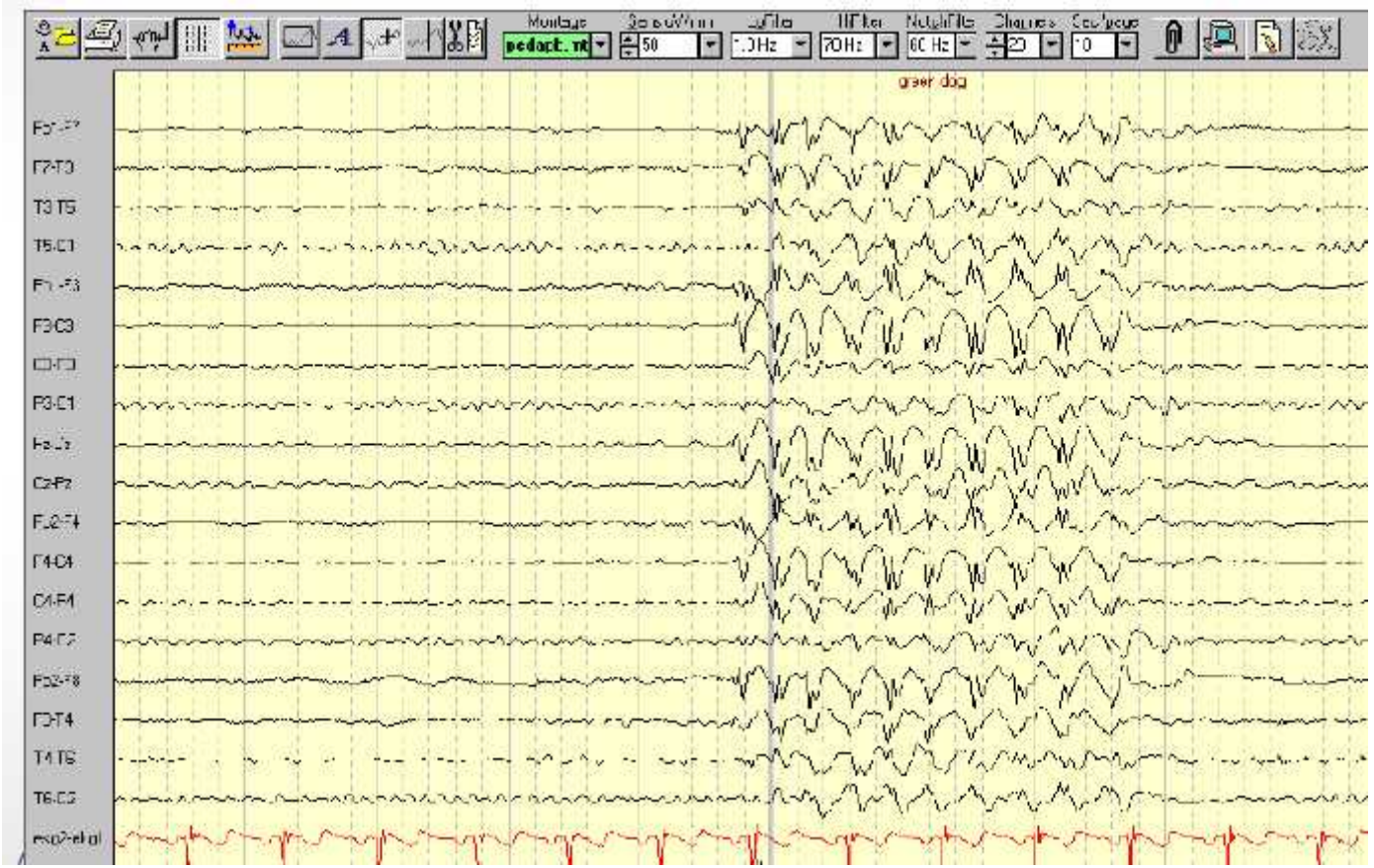
EEG in Absence Epilepsy



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Childhood Absence Epilepsy

- Typical absence seizures remit in ~ 80%
  - Remission rates lower if generalized tonic-clonic seizures co-exist
  - Average age of remission =10-11 years
- Favorable prognostic signs for “outgrowing” absence seizures:
  - Negative family history
  - Normal background EEG
  - Normal intelligence

Child Absence Epilepsy Treatment

- Drug of Choice: Ethosuximide

- Randomized-controlled trial:
  - Ethosuximide = valproic acid for efficacy
  - Lamotrigine lower efficacy
  - Lamotrigine > ethosuximide > valproic acid for cognitive side effects

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Case #2

A 15-year-old girl was brought in following a generalized convulsion. It occurred shortly after waking up, while she was getting ready for school. The seizure lasted 45 seconds, followed by 30 minutes of confusion, and then recovery to her baseline. Birth, developmental, past medical and family histories were normal.

The girl also described feeling “shaky” in the morning. Her hands often jerked, and she had dropped her hairbrush or cup on occasion. These symptoms were present for a few months preceding the convulsion.

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Diagnosis?

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Juvenile Myoclonic Epilepsy

- Onset: 12-18 years
- Myonclonus
  - Early morning/photic stimulation-induced
  - May be recognized only in retrospect
- Generalized Convulsive Seizures occur in almost all patients and are often the representing symptom
- Absence seizures in 15-40%
- Prognosis:
  - JME usually persists for life
  - AED withdrawal not recommeneded
- Treatment:
  - Valproic acid used to be the drug of choice
  - Recently levetiracetam, lamotrigine, zonisamide

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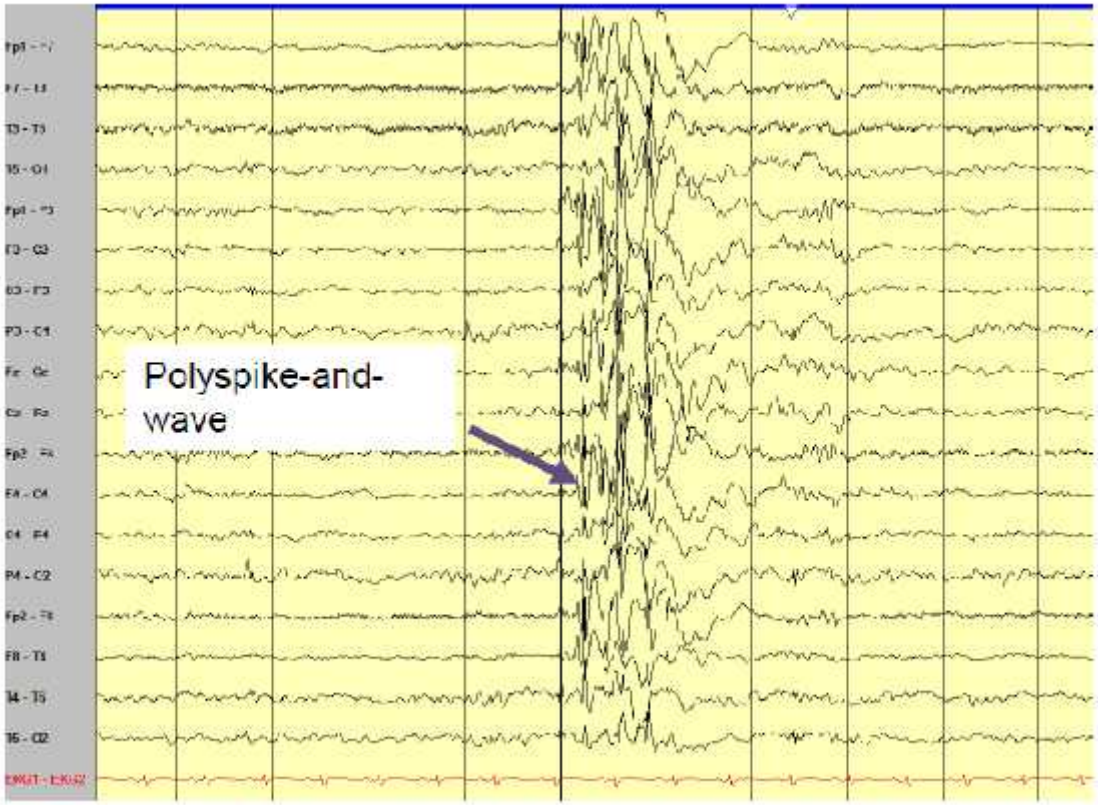
EEG in JME



Source:
Joshi and Shellhaas 2014

Case # 3

A 9 year old boy was referred because of possible seizures. He had been completely healthy until a month before, when he was awakened by his parents coming into their bedroom at 4:00 a.m. He looked somewhat frightened and appeared to be attempting to talk, but was able to make only unintelligible sounds. He was drooling from the right side of his mouth, which was twitching mildly. This continued for a few minutes, after which he gradually began to talk normally.

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Case #3, continued…

- At that time, he said that he had awakened from a sound sleep, aware that he couldn't talk or swallow, and remembered that he had come into his parents' room. He returned to normal functioning after 20 minutes. He was brought to his pediatrician for evaluation after he had a similar episode two weeks later. He had a normal neurologic examination.

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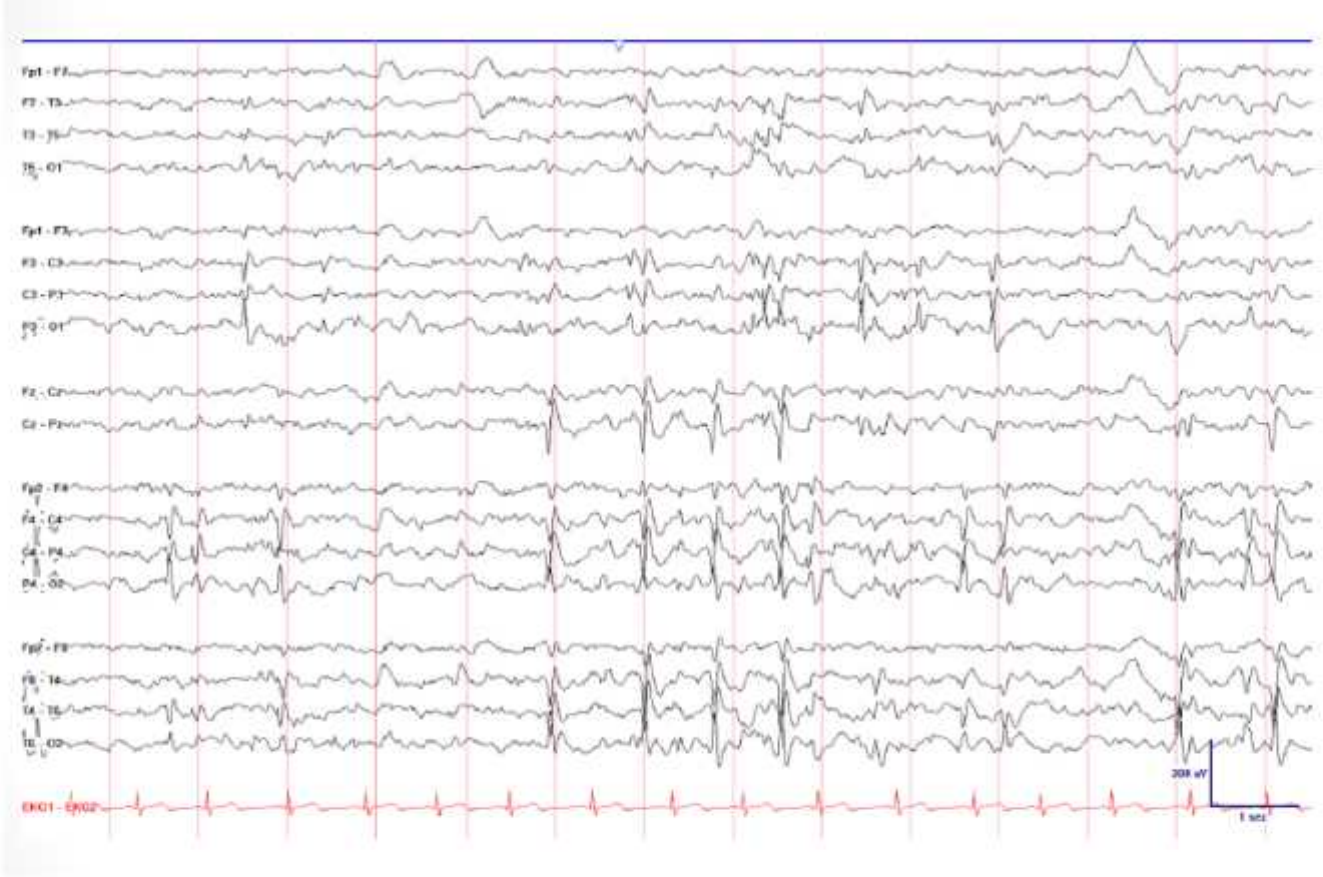

Source:
Joshi and Shellhaas 2014

Diagnosis?

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Benign Rolandic Epilepsy

(Benign Childhood Epilepsy with Centeral-Temporal Spikes)

- Most common focal epilepsy I childhood
- Age of onset 3-13 years
  - Peak = 7-8 years
- Typical scenario:
  - Predominantly nocturnal seizures
  - Focal seizure with motor symptoms involving face and arm
  - Seizure often secondarily generalize
- Cognitively normal child
  - Except language-based learning disorders
- Normal examination

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Benign Rolandic Epilepsy continued

- EEG shows characteristic interictal central-temporal focal sharp waves, activated by drowsiness/sleep
- Normal neuroimaging studies
  - Not needed if EEG is characteristic
- Good response to AED treatment
  - Oxcarbazepine
- Remission always occurs in second decade
  - Learning difficulties may persist

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Case #4

A 6-month-old infant presented with episodes of sudden forward bending at the waist during the two previous weeks. His mother worried that he was having abdominal pain, since he would let out a cry during the episodes. He had clusters of these episodes (with 10 or more in a cluster) several times per day, particularly upon awakening.

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Case #4, continued

His development seemed normal previously, but ever since these episodes started, he had become lethargic and had stopped sitting up by himself. He did not seem as playful and interactive as he had been, and no longer showed much interest in his toys.

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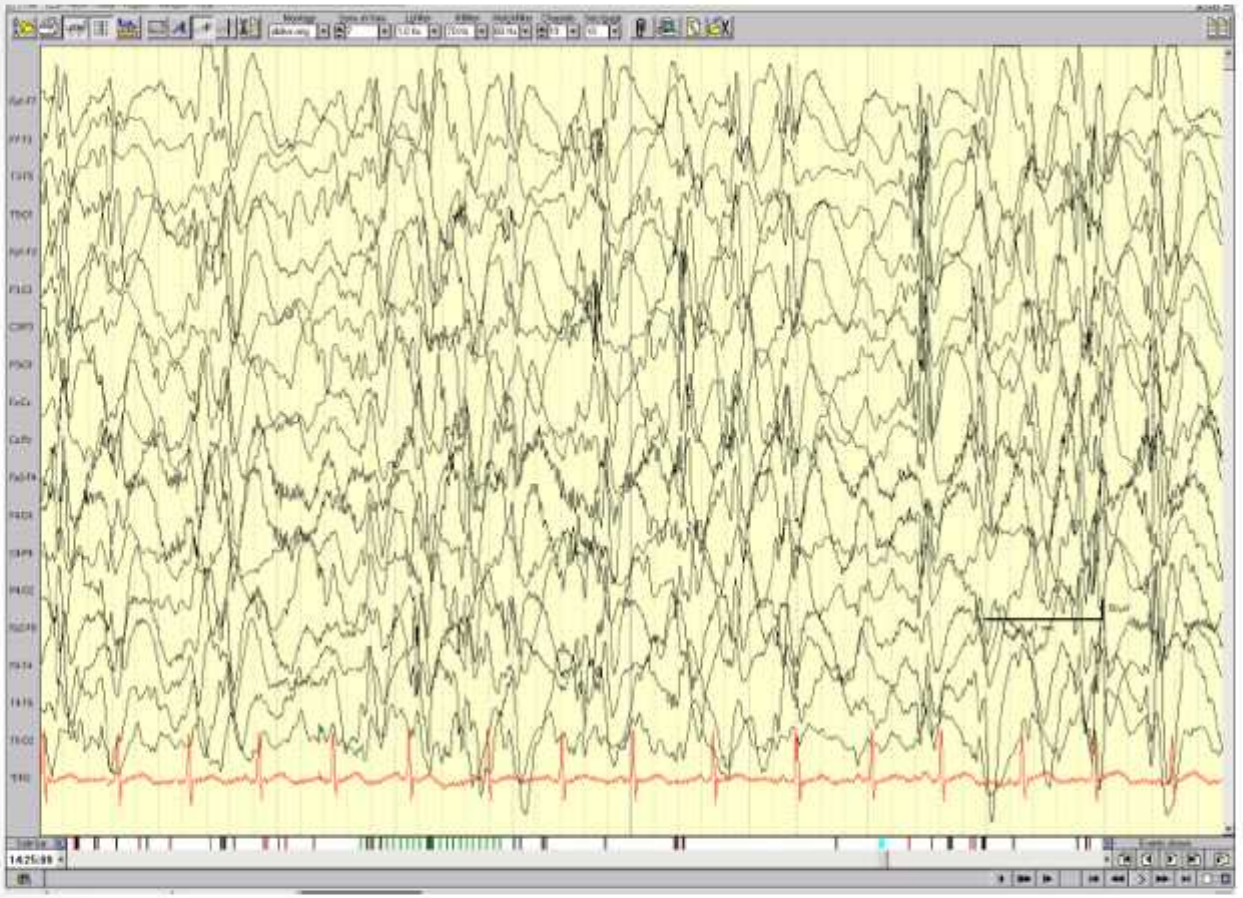

Source:
Joshi and Shellhaas 2014

Diagnosis?

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Infantile Spasms

- Incidence: 1 in 4000-6000 live births
- Onset: usually 4-8 months
- Clusters of flexor > mixed flexor-extensor > extensor spasms,
  - Often upon awakening

Infantile Spasms

- Cryptogenic/Unknown etiology: 10-15%
  - Normal exam and development before onset of spasms, normal imaging, no known etiology
- Symptomatic
  - Genetic: Tuberous sclerosis, Down Syndrome & other chromosomal abnormalities, ARX mutations, CDKL5 mutations, etc.
  - Prenatal: cerebral dysgenesis, IU infections (CMV), stroke
  - Perinatal: HIE, CNS infection
  - Postnatal: inborn errors of metabolism (NKH), head trauma, CNS infection, HIE, ICH
- 85% have intellectual disability

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West Syndrome

- Described by Dr. West in his own child
- Triad of:
  - Infantile spasms
  - Hypasar[r]hythmia
  - Mental retardation

Infantile Spasms – Treatment

- Earlier treatment = better prognosis
- ACTH or high-dose prednisone
- Other options:
  - Vigabatrin (indicated in TS)
  - Topiramate
  - Benzodiazepines
  - Valproic acid
  - Ketogenic diet
  - Pyridoxine

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Lennox Gastaut Syndrome

- Infantile spasm patients may evolve to LGS
  - Age of onset 2-8 years; very poor diagnosis
- Diagnostic Criteria:
  - Multiple seizure types, including: tonic, atonic, atypical absence, GTCs
  - EEG: interictal slow spike and wave (1.5-2 Hz), generalized paroxysmal fast activity
  - Cognitive dysfunction/mental retardation
  - Often difficult to treat

PAROXYSMAL NON-EPILEPTIC EVENTS (PNEE)

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PNEE: Overview

- Diagnostic challenge
- Relies on good description of events
- Difficult to get accurate description
  - Event can be frightening to witness, may not be the most reliable or accurate in their description (especially at night)
  - May not get first hand description (event occurs at school/daycare)
  - Terms like “shaking” “zone out” are non-specific

- Misdiagnosis can be as high as 39% (Uldall et al 2006)

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PNEE

- Misdiagnosis
  - Unnecessary prescribing of AED's
  - Lifestyle restricitions
  - Social (and other) stigma
- Accurate diagnosis is important
- Specific treatment
- Some PNES carry their own morbidity (e.g. cardiac arrhythmias mimicking seizures)

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PNEE Events

- Can present with symptoms including shaking that mimic seizures
- Some distinguishing features of PNEE:
  - Longer duration (several minutes)
  - Often eyes are closed during a “convulsive” event
  - Symptoms can have a start-stop quality to them
  - Injury (tongue biting), bladder incontinence can occur with PNEE

PNEE Events

- May occur in patients with epilepsy
- Non-epileptic seizures in children seldom represent malingering
- Management should include collaboration between the neurology and psychiatry/psychology
- Important to recognize as a diagnostic entity to avoid overdiagnosis and/or over-treatment and associated risks

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Differentiating seizures from non-epileptic events

- Eyes are usually open during a seizure
- Negative phenomena: pallor, visual loss, bradycardia are less common with seizures
- Non-epileptic symptoms can co-exist with epilepsy

Spectrum of Non-epileptic events in children

- Breath holding spells
- BPPV
- Sandifer syndrome
- Self stimulatory behaviors
- Parasomnias
- Syncope and cardiac dysrhythmias
- Psychogenic events
- Hyperekplexia
- Alternating hemiplegia of childhood
- Migraine
- Movement d/o (tics, dystonia)
- Others…

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Case

- A developmentally normal 18 month old presented with episodes of loss of consciousness. After a toy was taken away from her, she began crying and then became apneic and developed circumoral cyanosis and lost consciousness for 15-20 seconds. She also became stiff and had a few extremity jerks. When she awoke she seemed fine.

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Breath Holding Spells

- Common PNEE in childhood
- Peak age of occurrence
- Two types:
  - Cyanotic
  - Pallid

BHS: Work up and treatment

- Workup:
  - EEG, Neuroimaging: not required
  - CBC: Iron deficiency anemia
- Treatment
  - Parent education and reassurance
  - Iron supplements (5-6mg/kg/day supplemental iron) can reduce frequency of cyanotic BHS if anemia present

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Case

- A 9 month old is brought in for episodic of back arching, turning his head to one side in irritability. This often happens around feeds. He is developmentally normal, but weight gain has been a little slow.

Sandifer Syndrome

- Dystonic posturing of the trunk +/- extremities or torticollis in association with gastroesophageal reflux.
- Typically presents in infancy (older ages have been described)
- May have associated hiatal hernia (not required for diagnosis)
- Reflux symptoms may not always be obvious

Case

- A 15 month old girl presents with episodes of leg stiffening. Several times a day, especially while seated in a chair or car seat, she extends and stiffens her legs for a few seconds at a time, sometimes repeatedly over a couple of minutes. She is awake and interactive during these, but unaware of what she is doing.

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Self Stimulatory Behaviors

- Also described as (infantile) masturbation or self-gratification behaviors
- Commonly seen in toddlers and young children, more often in girls

Staring Spells

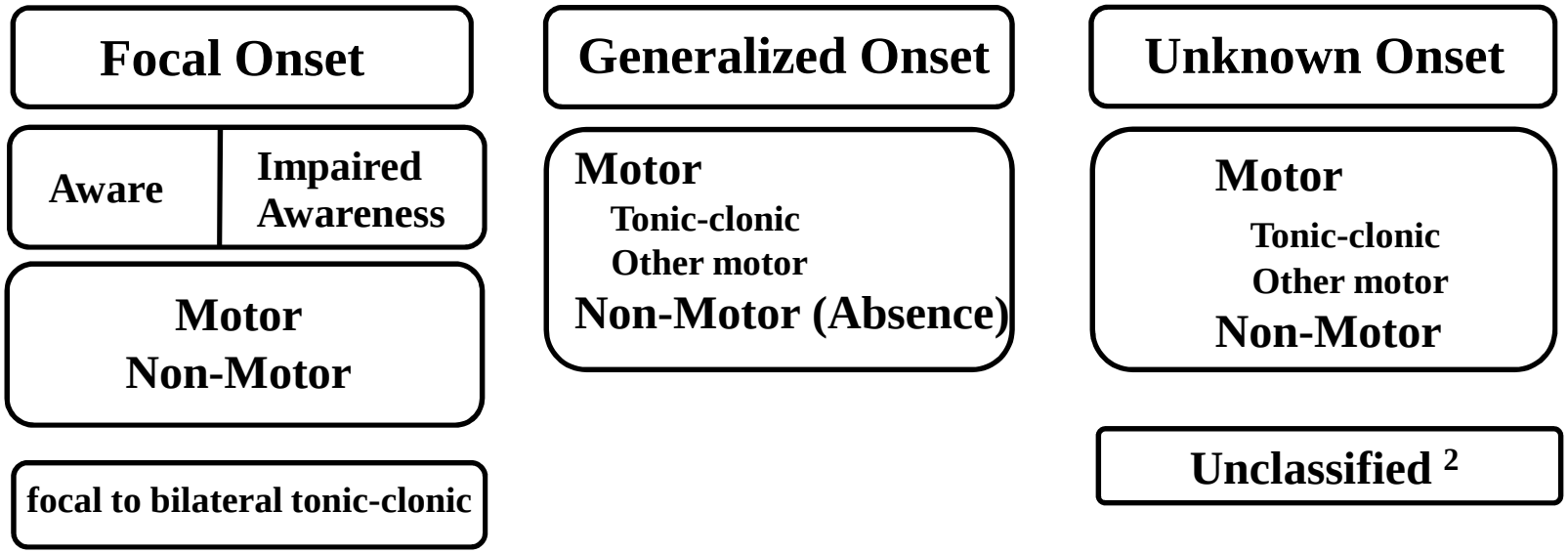
- Common presentation in children
- Can be a sign of absence or focal seizures
- Also seen in behavioral inattention, ADHD, daydreaming
- Vigorous tactile stimulation can help to assess responsiveness

The 2017 ILAE Classification of Seizures

Robert S. Fisher, MD, PhD
Maslah Saul MD Professor of Neurology
Director, Stanford Epilepsy Center

In 2017, the ILAE released a new classification of seizure types, largely based upon the existing classification formulated in 1981. Primary differences include specific listing of certain new focal seizure types that may previously only have been in the generalized category, use of awareness as a surrogate for consciousness, emphasis on classifying focal seizures by the first clinical manifestation (except for altered awareness), a few new generalized seizure types, ability to classify some seizures when onset is unknown, and renaming of certain terms to improve clarity of meaning.

ILAE 2017 Classification of Seizure Types Basic Version [1]



[1] Definitions, other seizure types and descriptors are listed in the accompanying paper & glossary of terms

[2] Due to inadequate information or inability to place in other categories

From *Fisher et al. Instruction manual for the ILAE 2017 operational classification of seizure types. Epilepsia 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

Non-Motor 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[2]

Non-Motor (absence)
- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

Motor
- tonic-clonic
- epileptic spasms

Non-Motor
- behavior arrest

Unclassified[3]

1 Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms.

2 These could be focal or generalized, with or without alteration of awareness

3 Due to inadequate information or inability to place in other categories

From *Fisher et al. Instruction manual for the ILAE 2017 operational classification of seizure types. Epilepsia doi: 10.1111/epi.13671*

INTERNATIONAL CLASSIFICATION OF SEIZURES 1981

Partial Seizures (start in one place)

- **Simple (no loss of consciousness of memory)**
  - **Sensory**
  - **Motor**
  - **Sensory-Motor**
  - **Psychic (abnormal thoughts or perceptions)**
  - **Autonomic (heat, nausea, flushing, etc.)**
- **Complex (consciousness or memory impaired)**
  - **With or without aura (warning)**
  - **With or without automatisms**
- **Secondarily generalized**

Generalized Seizures (apparent start over wide areas of brain)

- **Absence (petit mal)**
- **Tonic-clonic (grand mal)**
- **Atonic (drop seizures)**
- **Myoclonic**
- **Other**

Unclassifiable seizures

Dreifuss et al. Proposal for revised clinical and electroencephalographic classification of epileptic seizures. From the Commission on Classification and Terminology of the International League Against Epilepsy. Epilepsia. 1981;22:489-501.

Motivation for Revision

- **Some seizure types, for example tonic seizures or epileptic spasms, can have either a focal or generalized onset.**
- **Lack of knowledge about the onset makes a seizure unclassifiable and difficult to discuss with the 1981 system.**
- **Retrospective seizure descriptions often do not specify a level of consciousness, and altered consciousness, while central to many seizures, is a confusing concept.**
- **Some terms in current use do not have high levels of community acceptance or public understanding, such as “psychic,” “partial,” “simple partial,” “complex partial”, and “dyscognitive.”**
- **Some important seizure types are not included.**

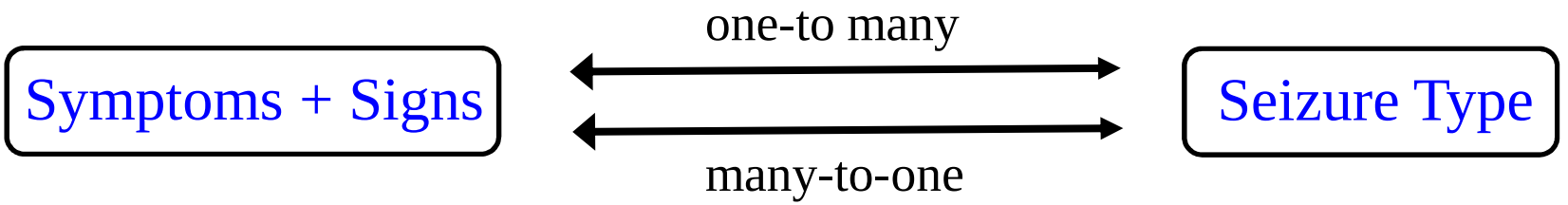
Possible Seizure Classifications Could be Based On:

Pathophysiology	Anatomy	Networks	Practical, by:	Modify Existing
But this is currently impossible with our limited understanding	Temporal Frontal Parietal Occipital Diencephalic Brainstem	Neocortical Limbic Thalamo-Cortical Brainstem	AED response Surgical target Disabling EEG pattern Many others	1981 ILAE System 2010 ILAE update

- In the absence of fundamental knowledge, ILAE chose to extend the existing classification
- The is an operational (practical) system, not a true scientific classification
- Others might devise special operational classifications for specific use, e.g., neonatal, ICU
- This classification is predominantly for clinicians

How Do Clinicians Classify Seizures ?

- **Elicit symptoms and signs of event (semiology)**
- **Look for familiar patterns in symptoms and signs**
- **Sometimes use ancillary data, e.g., EEG, MRI, genes, antibodies, etc.**



examples

Key Seizure Signs and Symptoms?

Symptoms	Medical Term
automatic behaviors	automatisms
emotions or appearance of emotions	emotions
extension or flexion postures	tonic
flushing/sweating/piloerection	autonomic
jerking arrhythmically	myoclonus
jerking rhythmically	clonus
language or thinking problems, deja vu	cognitive
lid jerks	eyelid myoclonia
limp	atonic
numb/tingling, sounds, smells, tastes visions, vertigo	sensations
pausing, freezing, activity arrest	behavior arrest
thrashing/pedaling	hyperkinetic
trunk flexion	spasm

The Elements of Change

- **Allow some seizures to be either focal or generalized onset**
- **Classify seizures of unknown onset**
- **Clarify “impairment of consciousness”**
- **Include a few previously unclassified types**
- **Update word usage for greater public clarity**
- **Validate use of supportive information, e.g. EEG**
- **Conform with ICD 11 and 12**
- **Update the 2001 glossary of seizure terms**
- **Standardize common descriptors to describe seizures**
- **Map old to new terms**

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Some Seizure Onsets can be Focal or Generalized

Focal Onset

atonic

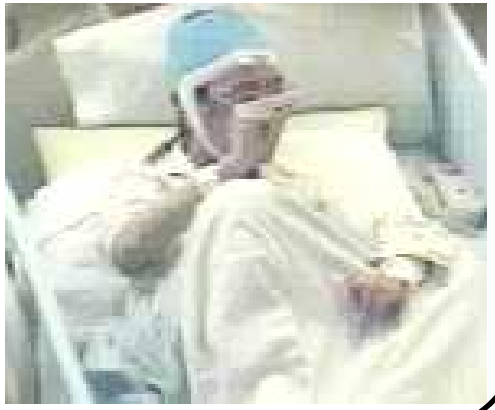
clonic

epileptic spasms

myoclonic

tonic →

tonic-clonic



Generalized Onset

atonic

clonic

epileptic spasms

myoclonic

← tonic

tonic-clonic

The Elements of Change

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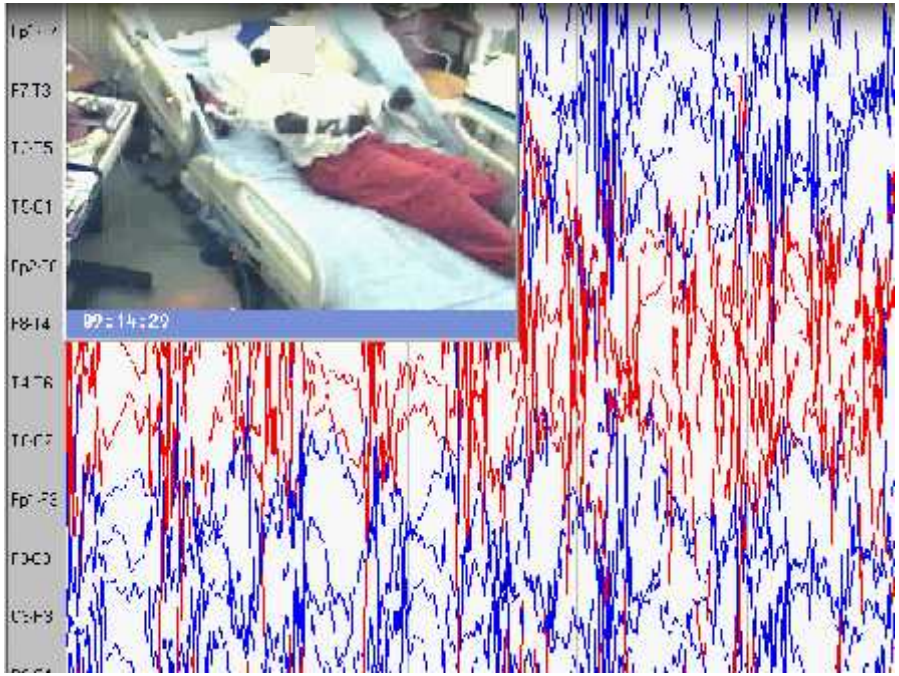
Seizures of Unknown Onset

Hypothetical case: You hear a noise and enter the video-EEG room to find the patient in bed, grunting, eyes rolled up, all limbs stiff, then rhythmically jerking for a minute. He was off-camera at the start. What seizure type is this?



Some seizure types are worth describing even if onset is unknown:

- **tonic-clonic**
- **epileptic spasms**
- **behavior arrest**

The Elements of Change

- **Allow some seizures to be either focal or generalized onset**
- **Classify seizures of unknown onset**
- **Clarify "impairment of consciousness"**
- **Include a few previously unclassified types**
- **Update word usage for greater public clarity**
- **Validate use of supportive information, e.g. EEG**
- **Conform with ICD 11 and 12**
- **Update the 2001 glossary of seizure terms**
- **Standardize common descriptors to describe seizures**
- **Map old to new terms**

Key Role of Impaired Consciousness

Among many possible behaviors during a seizure, impairment of consciousness has always had a key role in classifying the seizure, because of practical importance for:

- **Driving**
- **Safety during seizures**
- **Employability**
- **Interference with schooling and learning**

Loss (or Impairment) of Consciousness

Two types of seizures with loss of consciousness

How well does the public understand LOC during a complex partial seizure?

Loss (or Impairment) of Consciousness

Elements of consciousness

- **Awareness** of ongoing activities
- **Memory** for time during the event
- **Responsiveness** to verbal or nonverbal stimuli
- **Sense of self** as being distinct from others

Which would be the best surrogate marker ?

- **The 2017 Classification chooses awareness**
- **Consciousness remains in the classification but "awareness" is in the seizure name**
- **In several languages, these words are the same**
- **Awareness is not used to classify generalized onset seizures**

The Elements of Change

- **Allow some seizures to be either focal or generalized onset**
- **Classify seizures of unknown onset**
- **Clarify “impairment of consciousness”**
- **Include a few previously unclassified types**
- **Update word usage for greater public clarity**
- **Validate use of supportive information, e.g. EEG**
- **Conform with ICD 11 and 12**
- **Update the 2001 glossary of seizure terms**
- **Standardize common descriptors to describe seizures**
- **Map old to new terms**

New Seizure Types

New Focal Seizures

Motor	Non-Motor
atonic	behavior arrest
automatisms	(autonomic)
clonic	(cognitive)
epileptic spasms	emotional
hyperkinetic	(sensory)
myoclonic	
tonic	

New generalized seizures

absence with eyelid myoclonia
epileptic spasms (infantile spasms)
myoclonic-atonic (e.g., Doose)
myoclonic-tonic-clonic (e.g., JME)

New combined seizures

(focal to bilateral tonic-clonic)

(parentheses) indicates prior existence, but renaming

The Elements of Change

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Wording Changes

O L D T E R M	N E W T E R M
Unconscious (still used, not in name)	Impaired awareness (surrogate)
Partial	Focal
Simple partial	Focal aware
Complex partial	Focal impaired awareness
Dyscognitive (word discontinued)	Focal impaired awareness
Psychic	Cognitive
Secondarily generalized tonic-clonic	Focal to bilateral tonic-clonic
Arrest, freeze, pause, interruption	Behavior arrest

The Elements of Change

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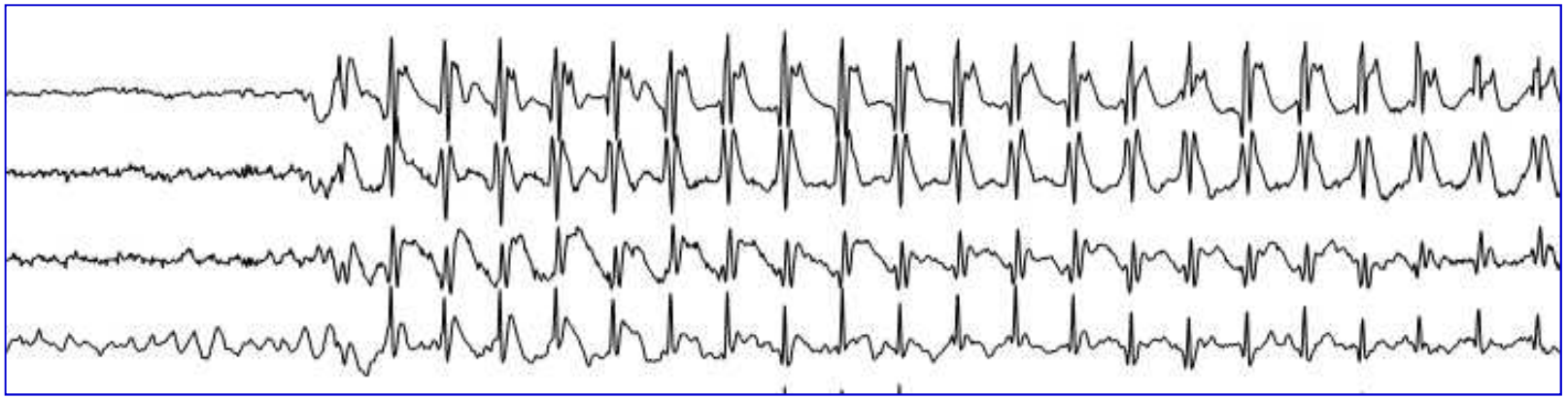
- **Map old to new terms**

Supportive Information

Seizures are usually classified by symptoms and signs

But supportive information may be helpful, when available:

- **Videos brought in by family**
- **EEG patterns**
- **Lesions detected by neuroimaging**
- **Laboratory results such as detection of anti-neuronal antibodies**
- **Gene mutations**
- **Diagnosis of an epilepsy syndrome diagnosis**



The Elements of Change

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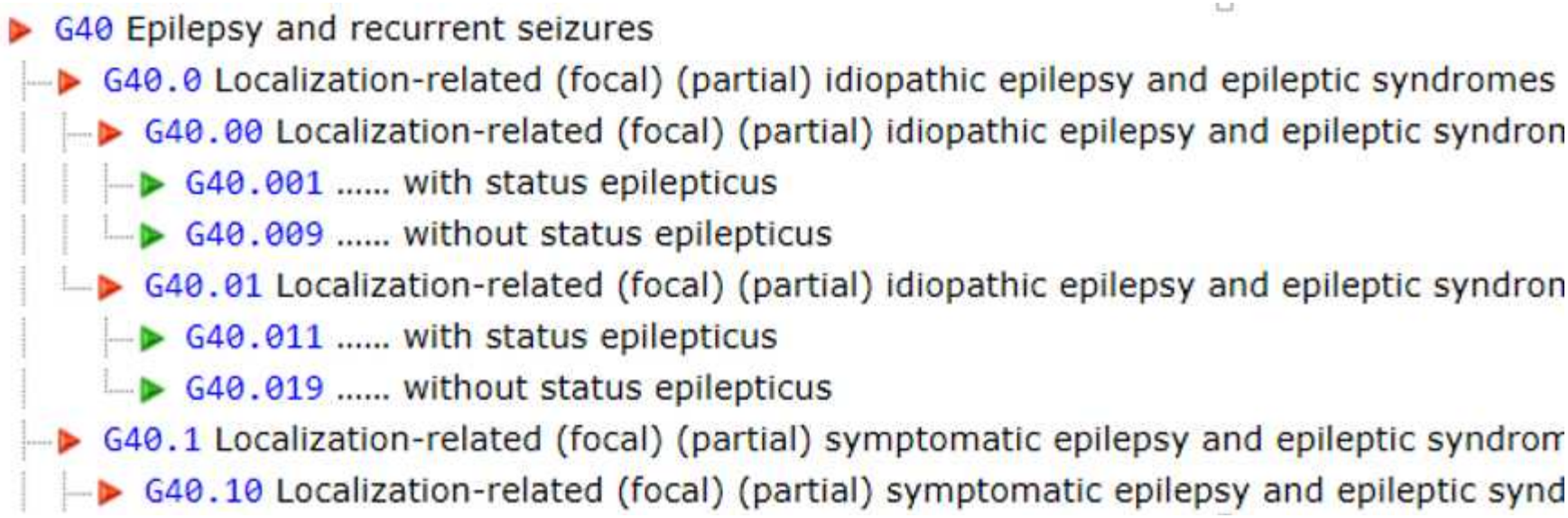
- **Map old to new terms**

ICD9, 10, 11, 12

- **ICD 9 & 10 are in use now with old terminology: petit mal, grand mal**
- **ICD 11 does not name seizure types, but ILAE syndromes and etiologies**
- **ICD 12 should conform to the new ILAE seizure type classification**



The Elements of Change

- **Allow some seizures to be either focal or generalized onset**
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Glossary: Full list in Epilepsia Paper

WORD	DEFINITION	SOURCE
absence, typical	a sudden onset, interruption of ongoing activities, a blank stare, possibly a brief up- ward 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 [11]
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 Dreifuss [1]
arrest	see behavioral arrest	new
atonic	sudden loss or diminution of muscle tone without apparent preceding myoclonic or tonic event lasting ~1 to 2 s, involving head, trunk, jaw, or limb musculature.	[11]
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 activity.	[11]

The Elements of Change

- **Allow some seizures to be either focal or generalized onset**
- **Classify seizures of unknown onset**
- **Clarify “impairment of consciousness”**
- **Include a few previously unclassified types**
- **Update word usage for greater public clarity**
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- **Conform with ICD 11 and 12**
- **Update the 2001 glossary of seizure terms**
- **Standardize common descriptors to describe seizures**
- **Map old to new terms**

Common Descriptors

of other symptoms and signs during seizures.

These are not seizure types, just suggested descriptive words.

A free text description is also highly encouraged.

Cognitive
- acalculia
- aphasia
- attention impairment
- déjà vu or jamais vu
- dissociation
- dysphasia
- hallucinations
- illusions
- memory impairment
- neglect
- forced thinking
- responsiveness impairment

Emotional or affective
- agitation
- anger
- anxiety
- crying (dacrystic)
- fear
- laughing (gelastic)
- paranoia
- pleasure

Autonomic
- asystole
- bradycardia
- erection
- flushing
- gastrointestinal
- hyper/hypoventilation
- nausea or vomiting
- pallor
- palpitations
- piloerection
- respiratory changes
- tachycardia

Automatisms
- aggression
- eye-blinking
- head-nodding
- manual
- oral-facial
- pedaling
- pelvic thrusting
- perseveration
- running (cursive)
- sexual
- undressing
- vocalization/speech
- walking

Motor
- dysarthria
- dystonic
- fencer's posture (figure-4)
- incoordination
- Jacksonian
- paralysis
- paresis
- versive

Sensory
- auditory
- gustatory
- hot-cold sensations
- olfactory
- somatosensory
- vestibular
- visual

Laterality
- left
- right
- bilateral

The Elements of Change

- **Allow some seizures to be either focal or generalized onset**
- **Classify seizures of unknown onset**
- **Clarify “impairment of consciousness”**
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- **Update word usage for greater public clarity**
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Examples of Mapping Old to New Terms
Full List in Epilepsia paper

Old Term for Seizure The most important are in bold	New Term for Seizure [choice] (optional)
absence	**(generalized) absence**
absence, atypical	(generalized) absence, atypical
absence, typical	(generalized) absence, typical
akinetic	[focal/generalized] atonic
astatic	[focal/generalized] atonic
atonic	[focal/**generalized] atonic**
aura	focal aware
clonic	[focal/generalized] clonic
complex partial	**focal impaired awareness**
convulsion	[focal/generalized] motor [tonic-clonic, tonic, clonic], focal to bilateral tonic-clonic

Rules for Classifying Seizures (1 of 2)

Onset: Decide whether seizure onset is focal or generalized, using an 80% confidence level.

Awareness: For focal seizures, decide whether to classify by degree of awareness or to omit awareness as a classifier.

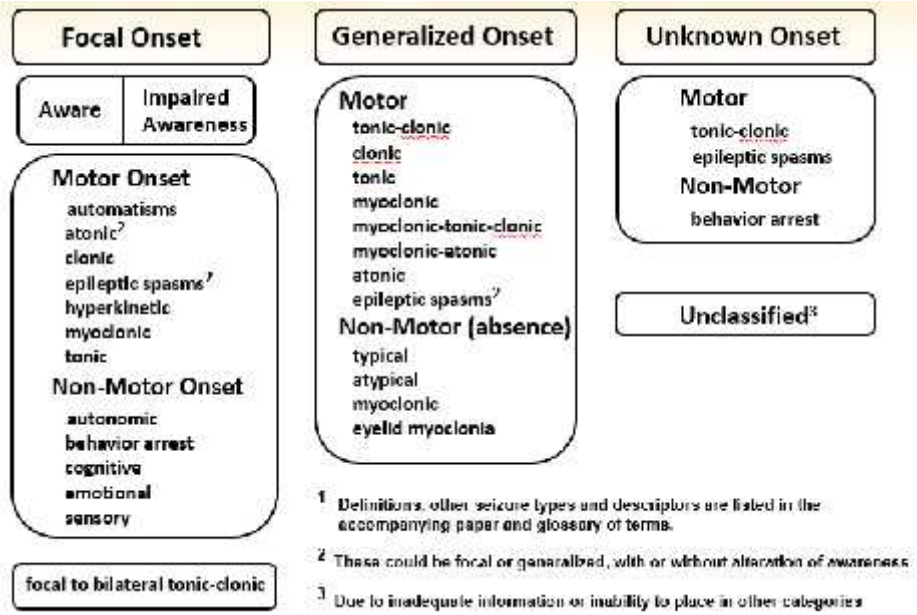
Impaired awareness at any point: A focal seizure is a *focal impaired awareness seizure* if awareness is impaired at any point during the seizure.

Onset predominates: Classify a focal seizure by its first prominent sign or symptom. Do not count transient behavior arrest.

Behavior arrest: A *focal behavior arrest seizure* shows arrest of behavior as the prominent feature of the entire seizure.

Motor/Non-motor: A *focal aware or impaired awareness seizure* maybe further sub-classified by motor or non-motor characteristics. Alternatively, a focal seizure can be characterized by motor or non-motor characteristics, without specifying level of awareness. Example, a *focal tonic seizure*.

Rules for Classifying Seizures (2 of 2)



Optional terms: Terms such as motor or non-motor may be omitted when the seizure type is otherwise unambiguous.

Additional descriptors: It is encouraged to add descriptions of other signs and symptoms, suggested descriptors or free text. These do not alter the seizure type. Example: *focal emotional seizure* with tonic right arm activity and hyperventilation.

Bilateral vs. generalized: Use the term "bilateral" for tonic-clonic seizures that propagate to both hemispheres and "generalized" for seizures that apparently originate simultaneously in both.

Atypical absence: Absence is atypical if it has slow onset or offset, marked changes in tone or EEG spike-waves at less than 3 per second.

Clonic vs. myoclonic: Clonic refers to sustain rhythmical jerking and myoclonic to a regular unsustained jerking.

Eyelid myoclonia: Absence with eyelid myoclonia refers to forced upward jerking of the eyelids during an absence seizure.

The Net Effect

The net effect of updating the Classification of Seizures should be the following:

1. Render the choice of a seizure type easier for seizures that did not fit into any prior categories;

2. Clarify what is meant when a seizure is said to be of a particular type;

3. Provide more transparency of terminology to the nonmedical community.

Examples

1. A woman awakens to find her husband having a seizure in bed. The onset is not witnessed, but she is able to describe bilateral stiffening followed by bilateral shaking. EEG and MRI are normal.

Examples

Old = unclassified
New = unknown onset tonic-clonic

1. A woman awakens to find her husband having a seizure in bed. The onset is not witnessed, but she is able to describe bilateral stiffening followed by bilateral shaking. EEG and MRI are normal. This seizure is classified as *onset unknown tonic-clonic*. There is no supplementary information to determine if the onset was focal or generalized. In the old classification, this seizure would have been unclassifiable.

Examples

2. In an alternate scenario of case #1, the EEG shows a clear right parietal slow wave focus. The MRI shows a right parietal region of cortical dysplasia.

Examples

Old = partial onset, secondarily generalized seizure
New = focal to bilateral tonic-clonic seizure

2. In an alternate scenario of case #1, the EEG shows a clear right parietal slow wave focus. The MRI shows a right parietal region of cortical dysplasia. In this circumstance, the seizure can be classified as focal to bilateral tonic-clonic, despite the lack of an observed onset, because a focal etiology has been identified, and the overwhelming likelihood is that the seizure had a focal onset. The old classification would have classified this seizure as partial onset, secondarily generalized seizure.

Examples

3. A child is diagnosed with Lennox-Gastaut syndrome of unknown etiology. EEG shows runs of slow spike-wave. Seizure types include absence and others.

Examples

Old = atypical absence seizures
New = atypical absence seizures

3. A child is diagnosed with Lennox-Gastaut syndrome of unknown etiology. EEG shows runs of slow spike-wave. Seizure types with this child include absence, tonic, and focal motor seizures. In this case, the absence seizures are classified as atypical absence (the word "generalized" may be assumed) due to the EEG pattern and underlying syndrome. The absence seizures would have had the same classification in the old system.

Examples

4. The same child as in #3 has seizures with stiffening of the right arm and leg, during which responsiveness and awareness are retained.

Examples

Old = tonic seizures
New = focal aware tonic seizures

4. The same child as in #3 has seizures with stiffening of the right arm and leg, during which responsiveness and awareness are retained. This seizure is a focal aware tonic seizures (the word "motor" can be assumed). In the old system, the seizures would have been called tonic seizures, with a perhaps incorrect assumption of generalized onset.

Examples

5: A 25 year old woman describes seizures beginning with 30 seconds of an intense feeling that “familiar music is playing.” She can hear other people talking, but afterwards realizes that she could not determine what they were saying. After an episode, she is mildly confused, and has to “reorient herself.”

Examples

Old = complex partial seizures
New = focal seizures with impaired awareness

5: A 25 year old woman describes seizures beginning with 30 seconds of an intense feeling that "familiar music is playing." She can hear other people talking, but afterwards realizes that she could not determine what they were saying. After an episode, she is mildly confused, and has to "reorient herself." The seizures would be classified as focal seizures with impaired awareness. Even though the patient is able to interact with her environment, she cannot interpret her environment, and is mildly confused.

Examples

6. A 22 year-old man has seizures during which he remains fully aware, with the “hair on my arms standing on edge” and a feeling of being flushed.

Examples

Old = simple partial autonomic seizures
New = focal aware autonomic seizures

6. A 22 year-old man has seizures during which he remains fully aware, with the "hair on my arms standing on edge" and a feeling of being flushed. These are classified as focal aware non-motor autonomic, or more succinctly focal aware autonomic. The old classification would have called them simple partial autonomic seizures.

Examples

7. A 4 year-old boy with myoclonic-atonic epilepsy (Doose syndrome) has seizures with a few arm jerks, then a limp drop to the ground.

Examples

Old = myoclonic astatic seizures
New = myoclonic-atonic seizures

7. A 4 year-old boy with myoclonic-atonic epilepsy (Doose syndrome) has seizures with a few arm jerks, then a limp drop to the ground. These are now classified as myoclonic-atonic seizures (the word “generalized” may be assumed). The old classification would have called these unclassified or unofficially, myoclonic-astatic seizures.

Examples

8. A 35 year-old man with juvenile myoclonic epilepsy has seizures beginning with a few bilateral arm jerks, followed by stiffening of all limbs and then rhythmic jerking of all limbs.

Examples

Old = myoclonic seizures followed by a tonic-clonic seizure
New = myoclonic-tonic-clonic seizures

8. A 35 year-old man with juvenile myoclonic epilepsy has seizures beginning with a few regularly-spaced jerks, followed by stiffening of all limbs and then rhythmic jerking of all limbs. This would be classified as generalized myoclonic-tonic-clonic seizures. No corresponding single seizure type existed in the old classification, but they might have been called myoclonic seizures followed by a tonic-clonic seizure.

Examples

9. A 14-month old girl has sudden flexion of both arms with head flexing forward for about 2 seconds. These seizures repeat in clusters. EEG shows hypsarrhythmia with bilateral spikes, most prominent over the left parietal region. MRI shows a left parietal dysplasia.

Examples

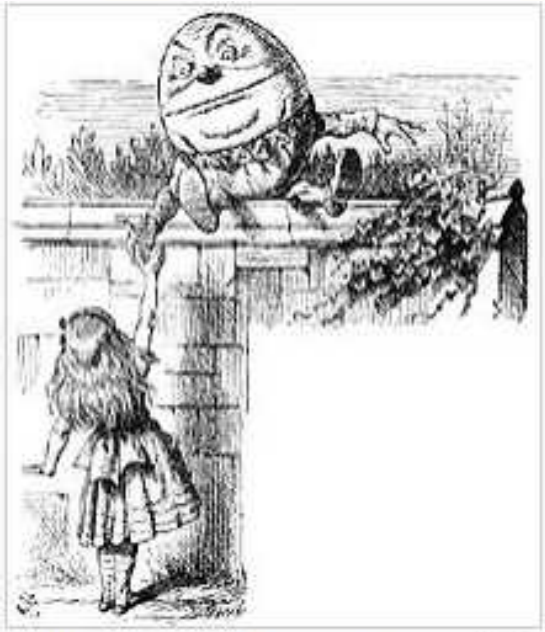
Old = infantile spasms (focality not specified)
New = focal epileptic spasms

9. A 14-month old girl has sudden flexion of both arms with head flexing forward for about 2 seconds. These seizures repeat in clusters. EEG shows hypsarrhythmia with bilateral spikes, most prominent over the left parietal region. MRI shows a left parietal dysplasia. Because of the ancillary information, the seizure type would be considered to be focal epileptic spasms (the term "motor" can be assumed). The previous classification would have called them infantile spasms, with information on focality not included.

Examples

10. A 75 year-old man reports an internal sense of body trembling. No other information is available.

The End



When *I* use a word,' Humpty Dumpty said in rather a scornful tone, 'it means just what I choose it to mean — neither more nor less.

"Words, words, words, I'm so sick of words!"
Eliza Doolittle, *My Fair Lady*

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

ILAE POSITION PAPER

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

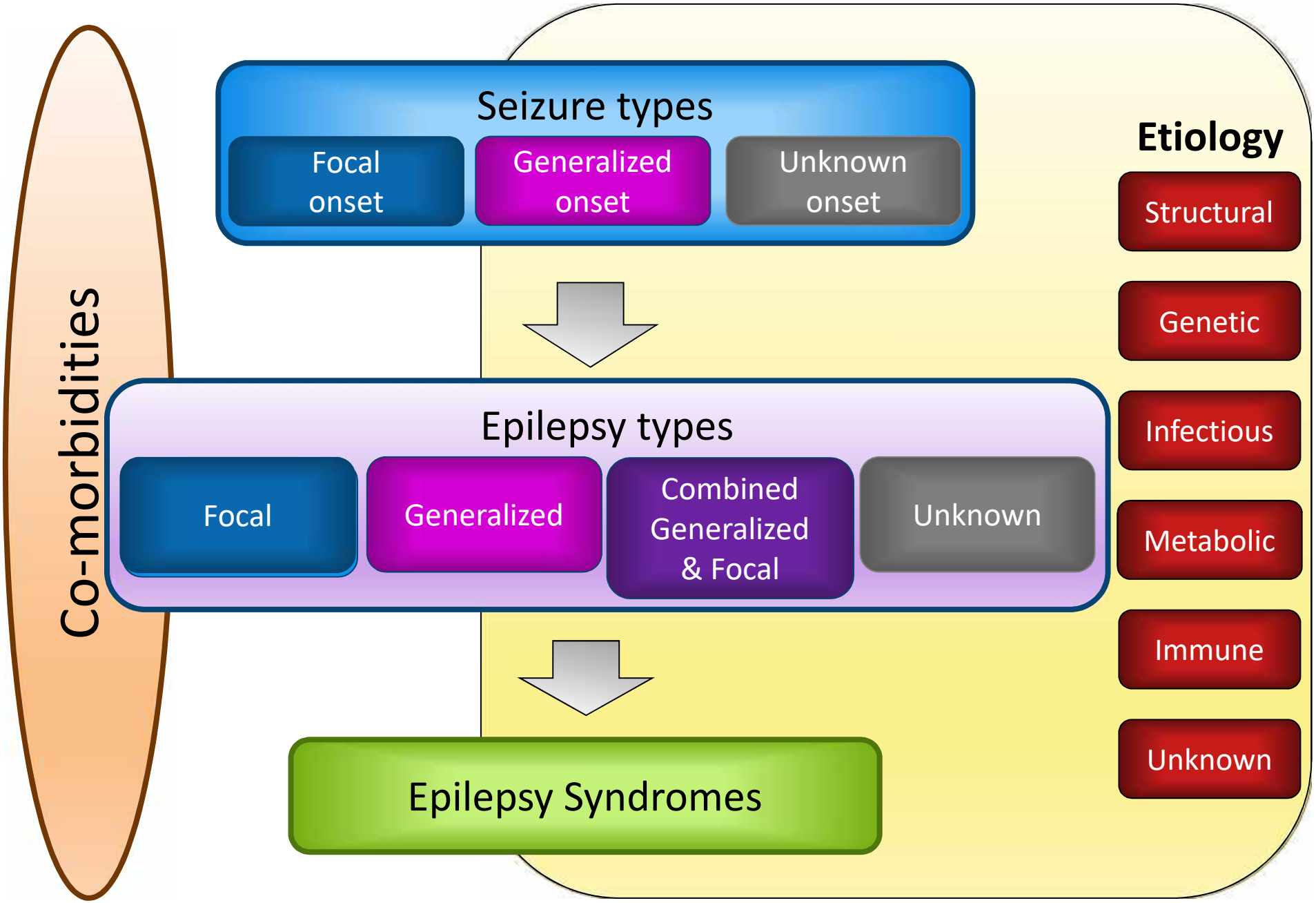
*Robert S. Fisher, †J. Helen Cross, ‡Jacqueline A. French, §Norimichi Higurashi, ¶Edouard Hirsch, #Floor E. Jansen, **Lieven Lagae, ††Solomon L. Moshé, ‡‡Jukka Peltola, §§Eliane Roulet Perez, ¶¶Ingrid E. Scheffer, and ##***Sameer M. Zuberi

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

Classification of the Epilepsies

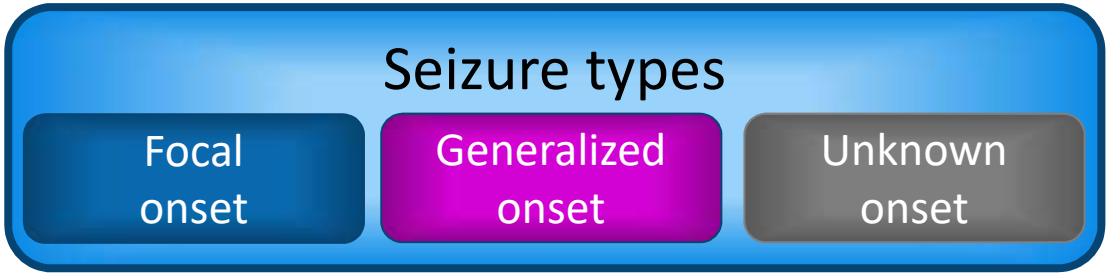
Purpose: for clinical diagnosis

Transparent language: use words that mean what they say


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

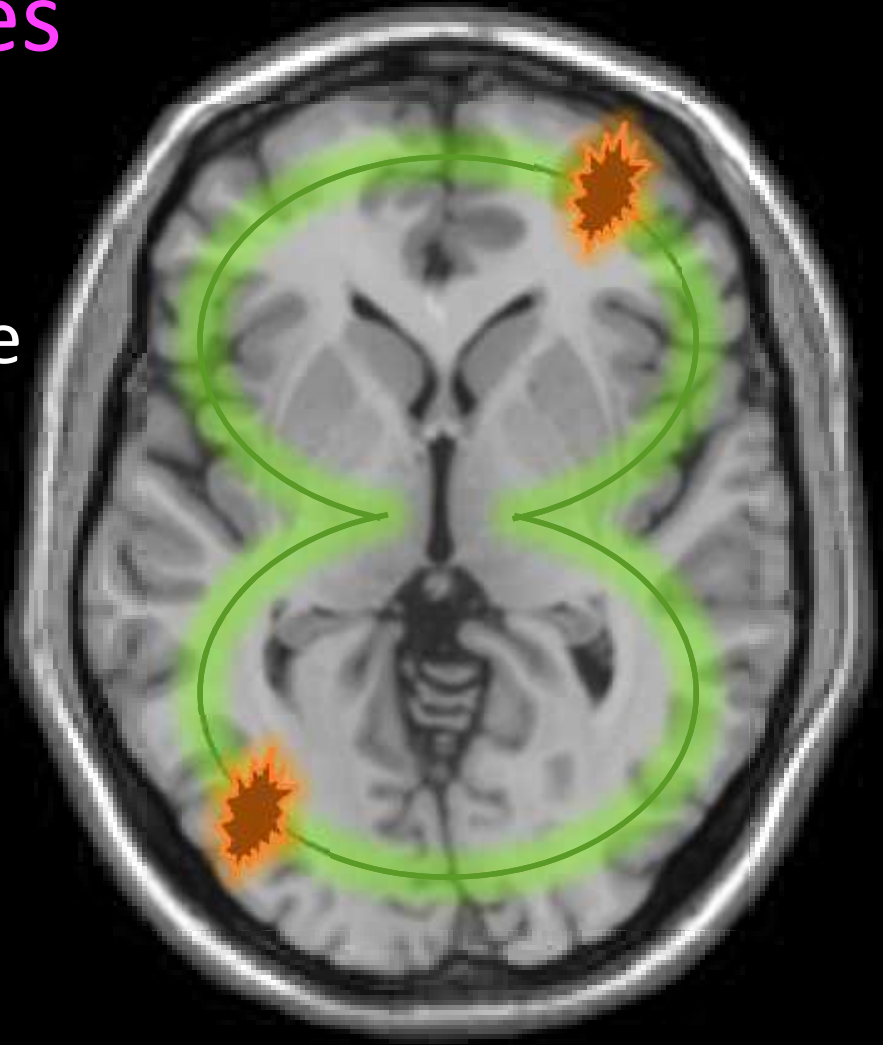
1. Seizure types

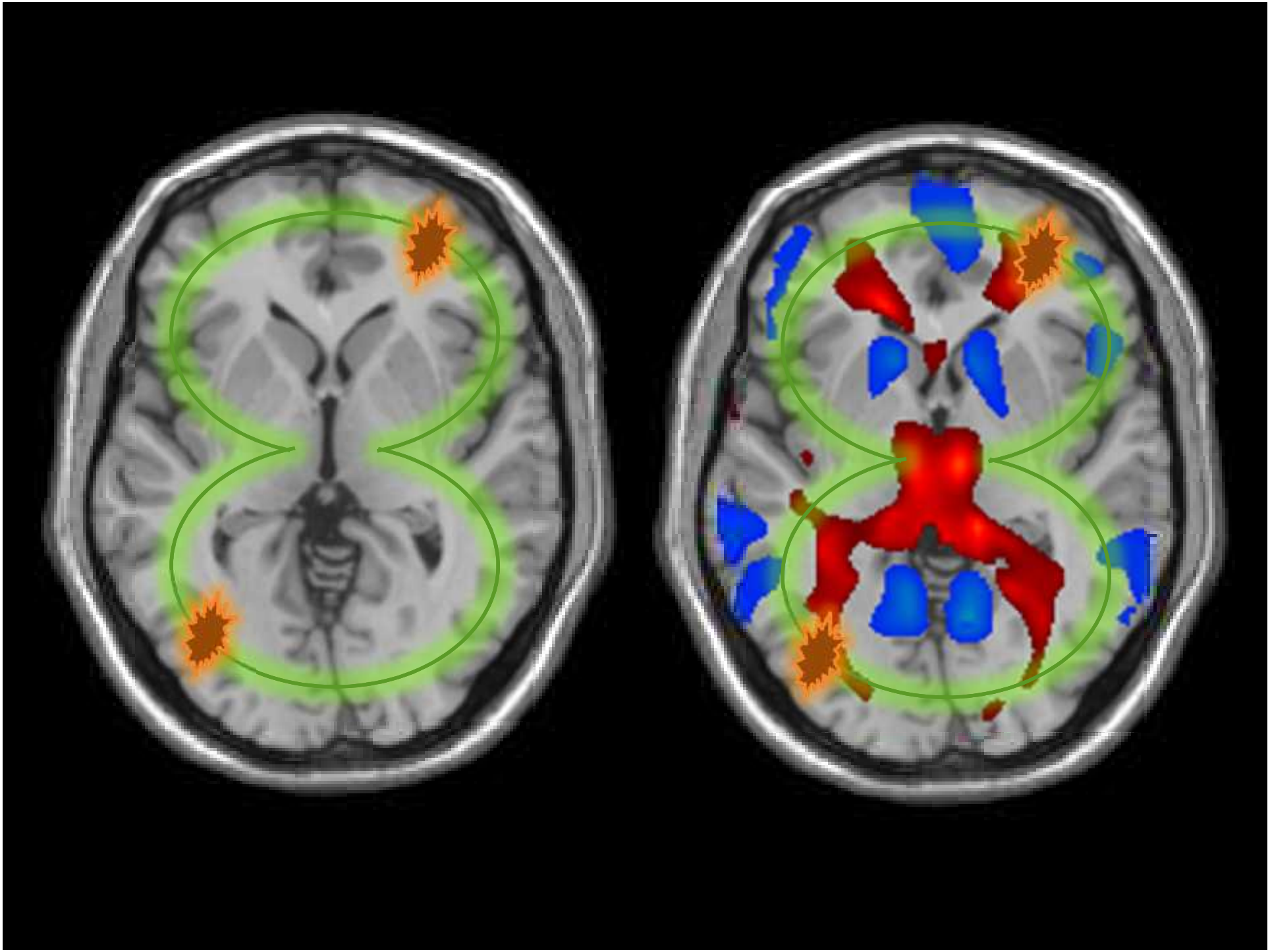
- Certain that events are epileptic seizures – **not** referring to distinguishing epileptic versus non-epileptic
- In some settings → classification according to seizure type may be maximum level of diagnosis possible
- In other cases → simply too little information to be able to make a higher level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


Seizure types
Focal onset
Generalized onset
Unknown onset

Generalized seizures

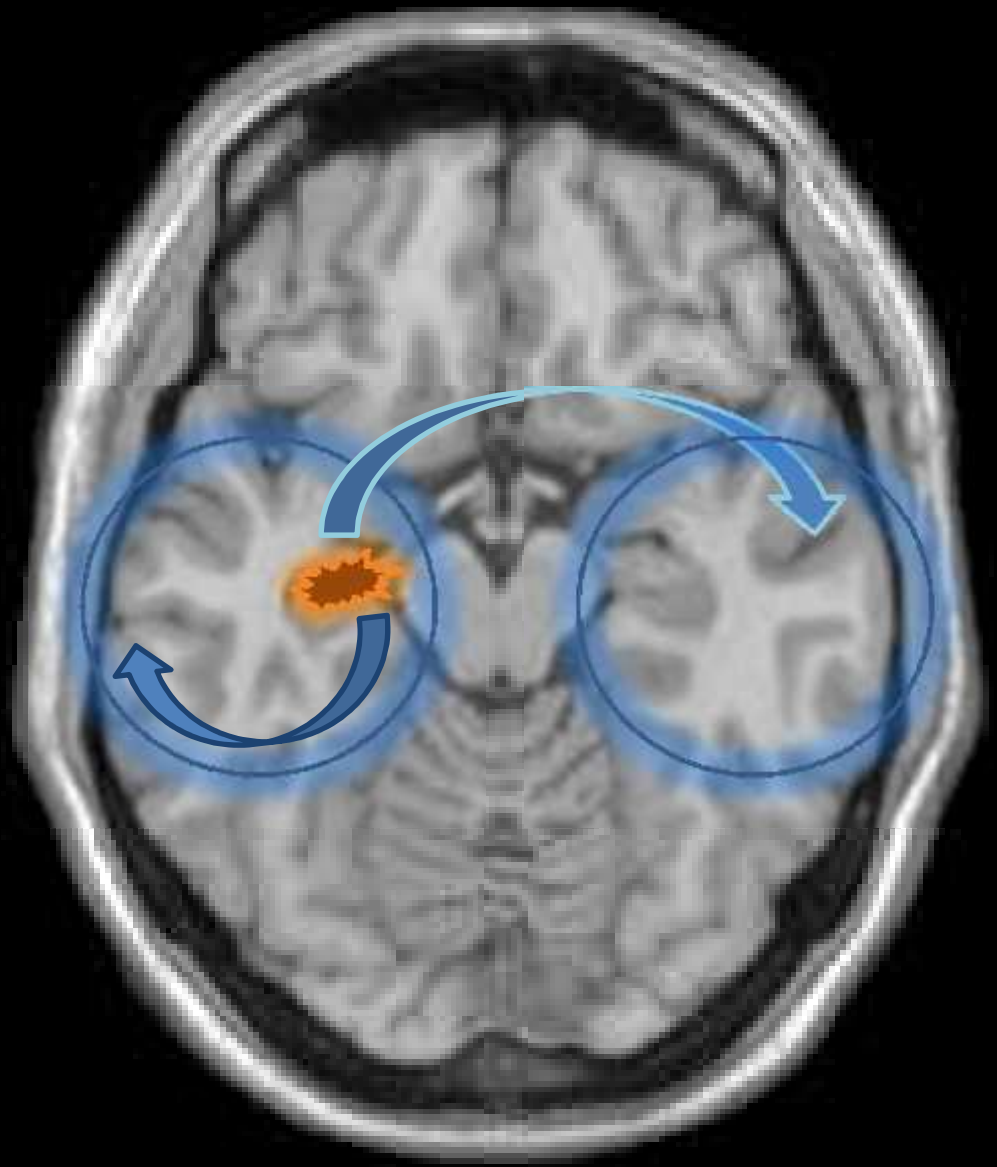
- Originate at some point within and rapidly engage bilaterally distributed networks
- Can include cortical and subcortical structures but not necessarily the entire cortex



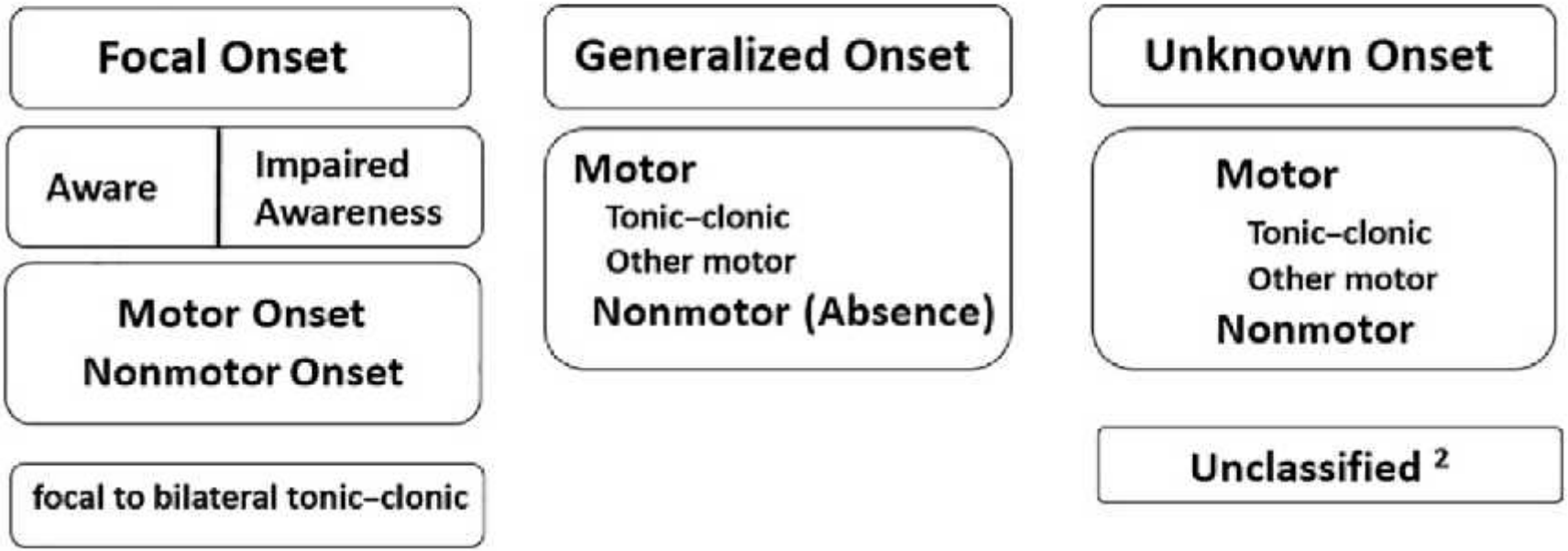


Focal seizures

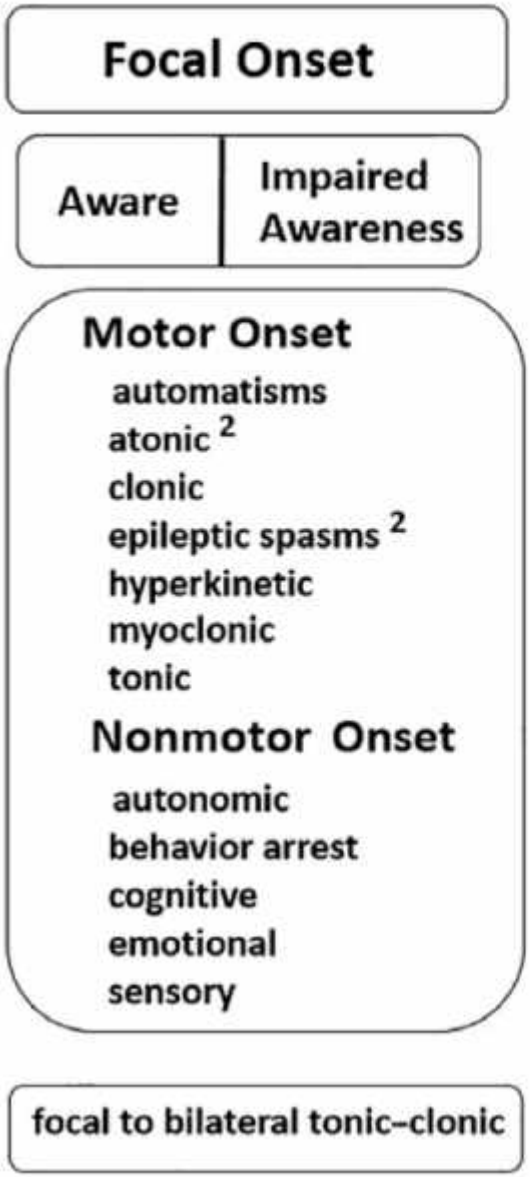
- Originate within networks limited to one hemisphere
- May be discretely localized or more widely distributed….



ILAE 2017 Classification of Seizure Types Basic Version [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

ILAE POSITION PAPER

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

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
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2 Awarene
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3 Impaired
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impaired
4 Onset pr
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sient beh
5 Behavior
shows ar
the entire
6 Motor/no

Table 3.

Old term for	
Absence	
Absence, aty	
Absence, typ	
Akinetic	
Astatic	
Atonic	
Aura	
Clonic	
Complex p	
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]

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

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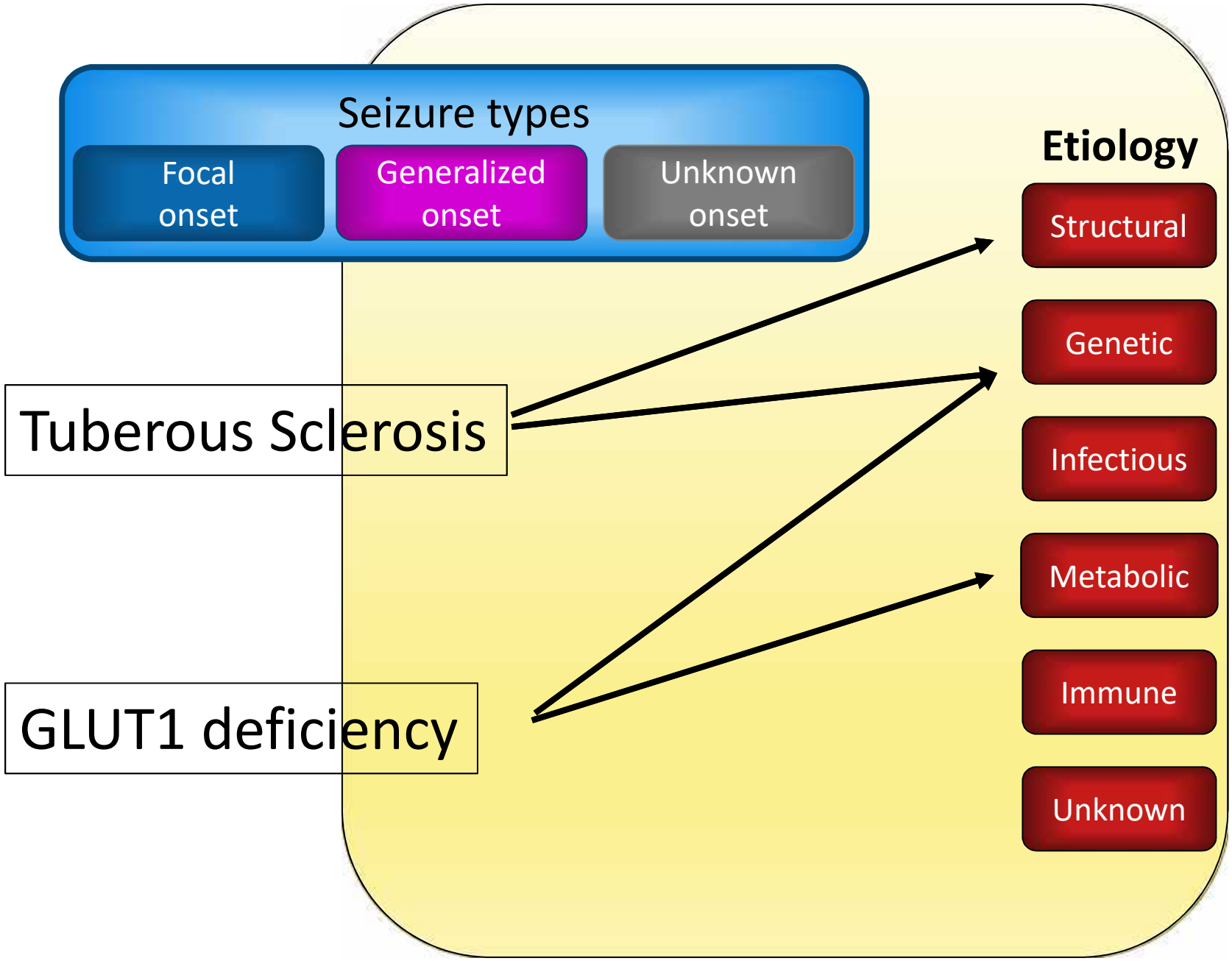
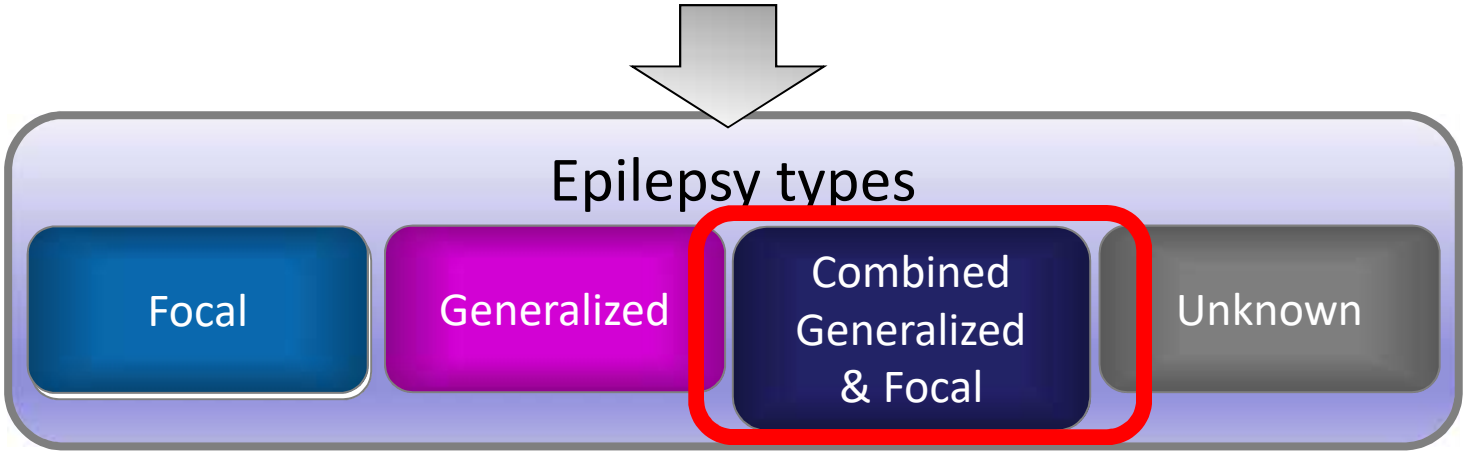
New New
Adapted fr Ref. 12 New
New
12 Adapted fr Ref. 12 New
Adapted fr Ref. 12

nms 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

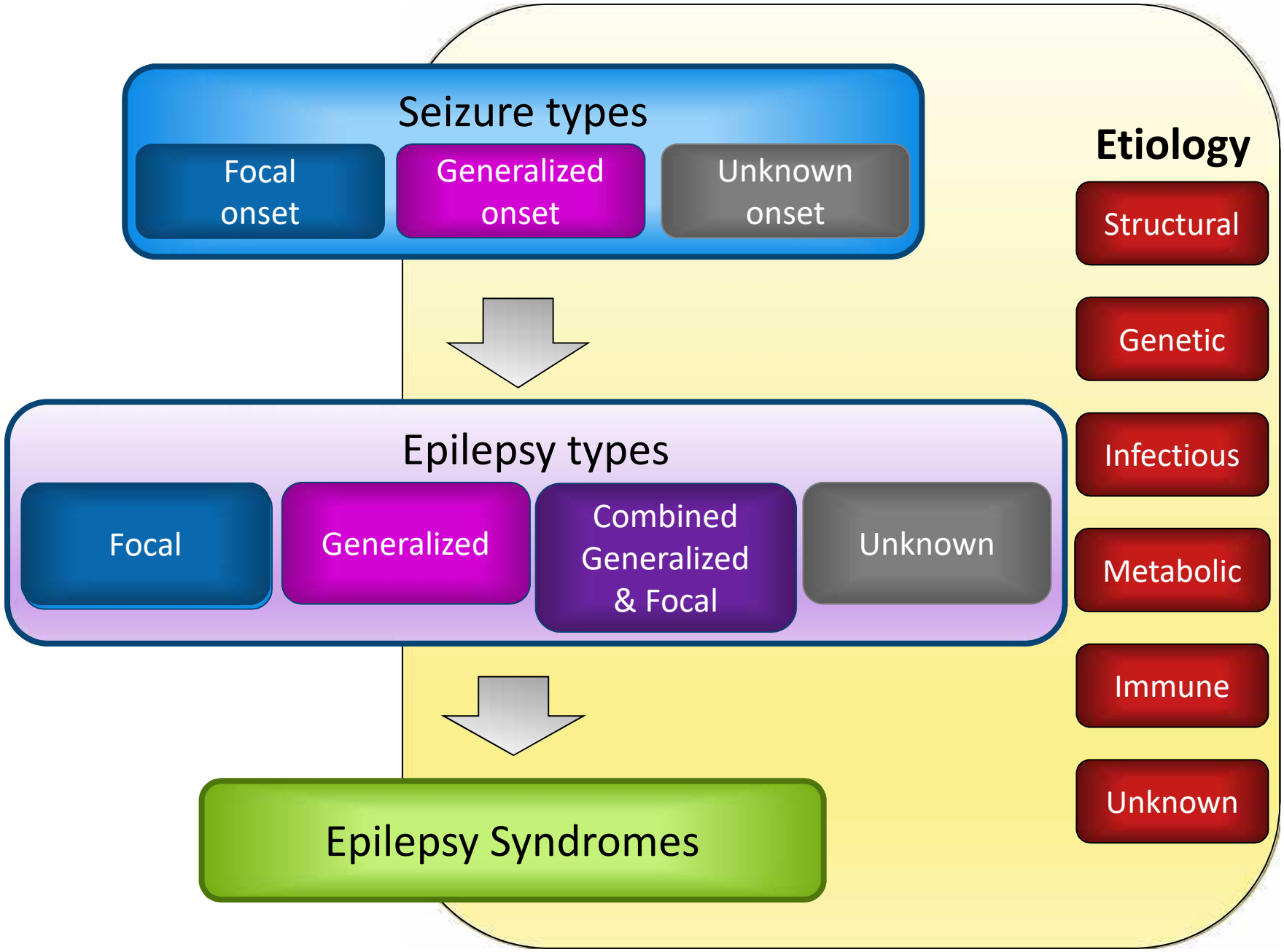
Generalized *and* Focal Epilepsies

- Combined focal and generalized epilepsies

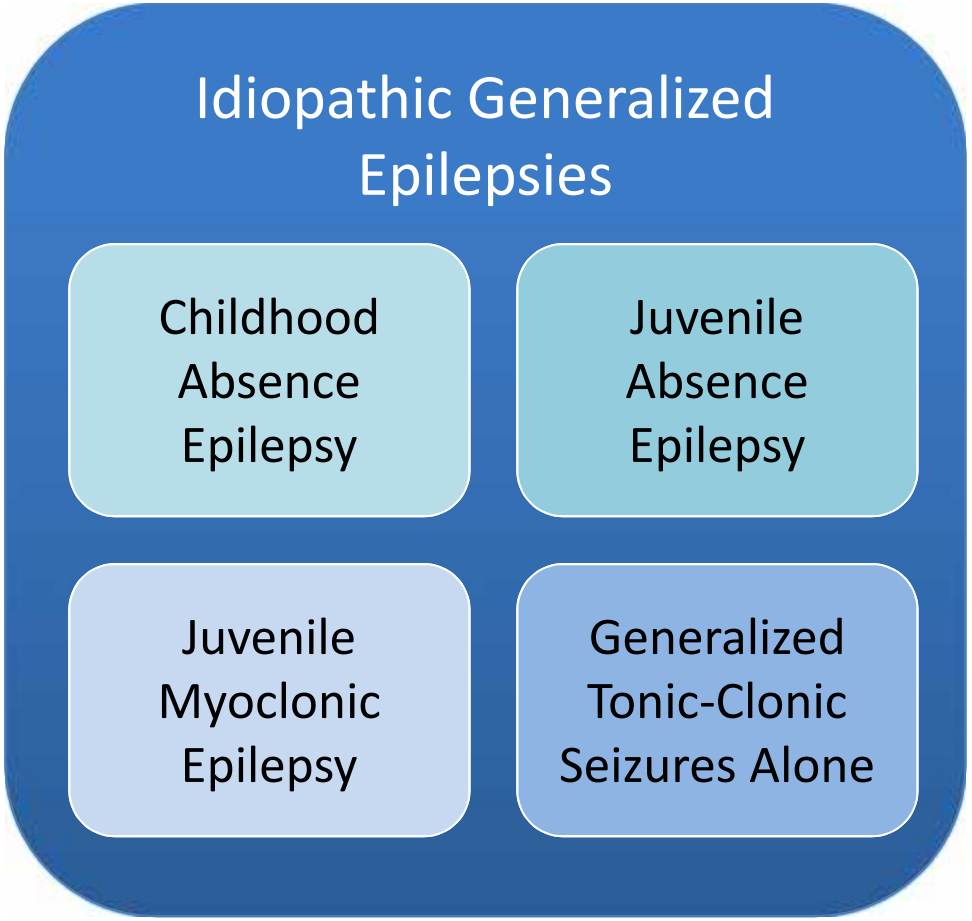
  Examples

  – 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

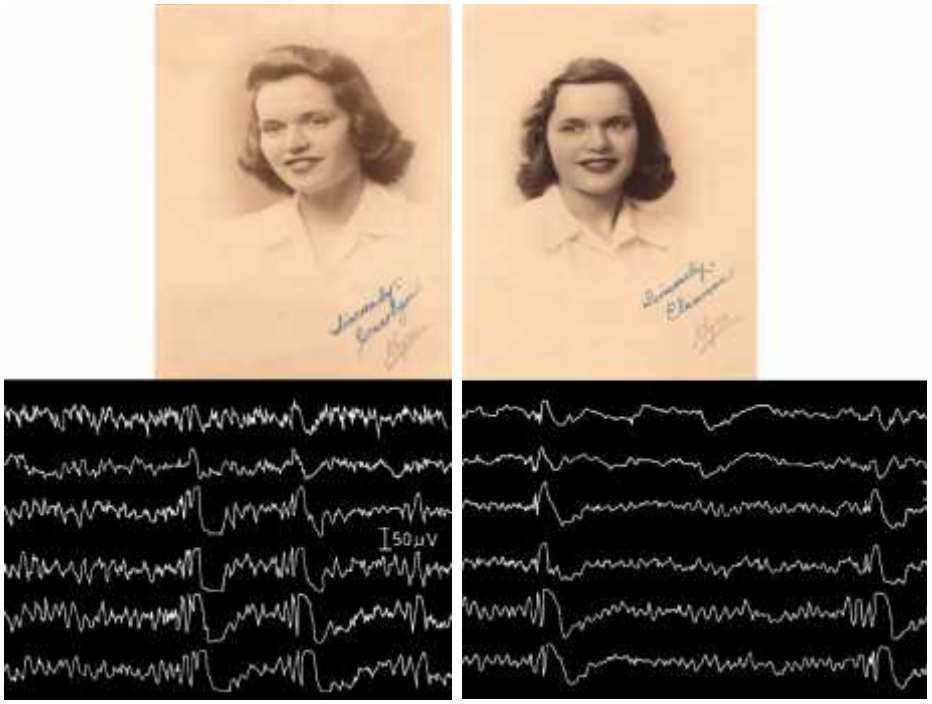
- 'Idiopathic' = presumed hereditary predisposition

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

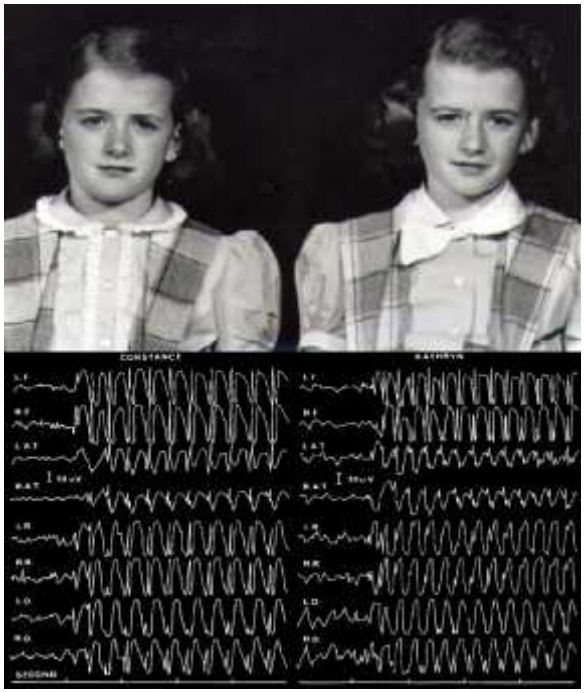
- Critical problem of stigma in some parts of the world

Genetic ≠ Gene testing

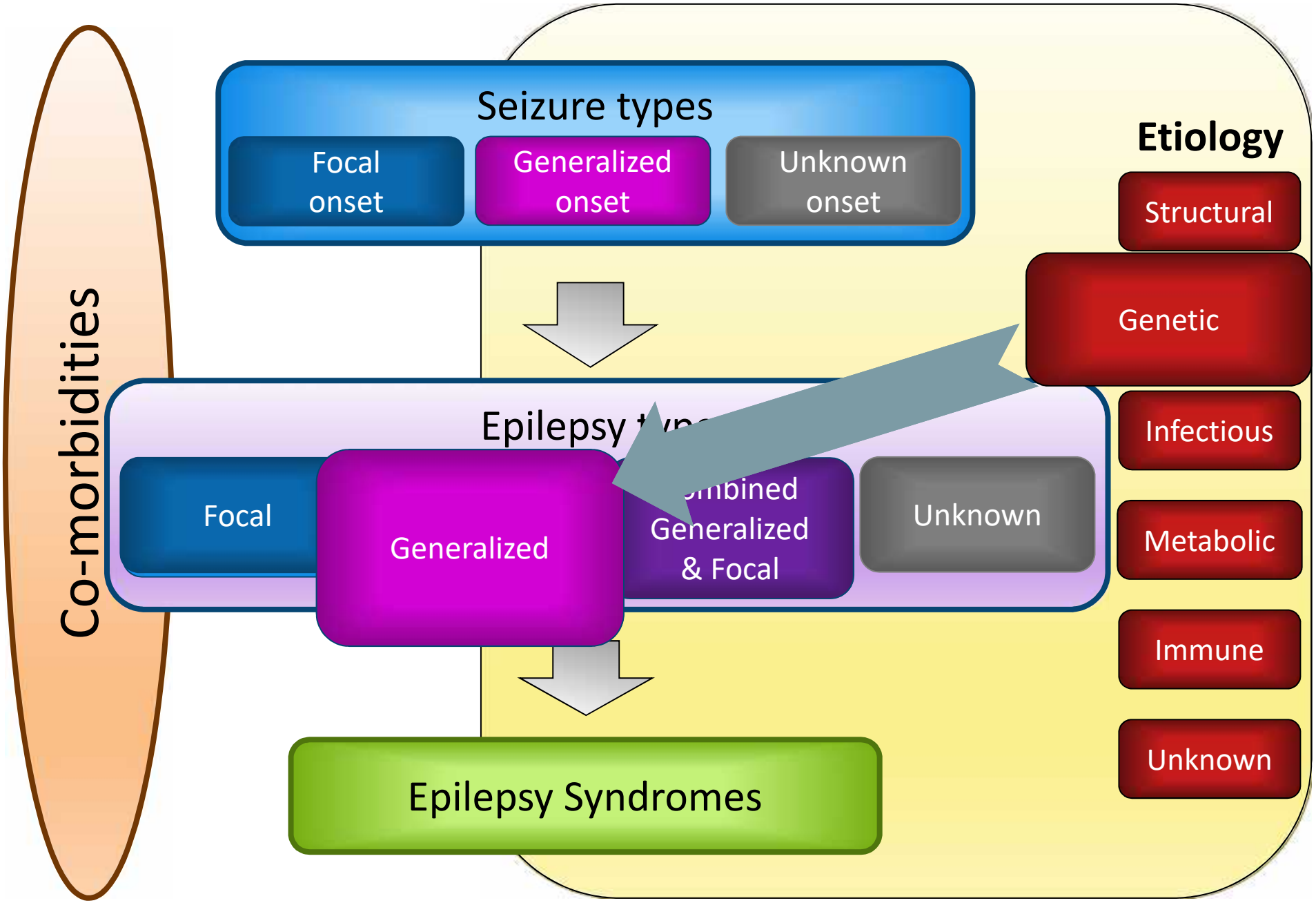
- Usually the mutation is *not* known
- Access to molecular genetic testing *not* necessary
- Diagnosed on clinical research eg. twin, family studies



JME pair; Lennox 1941



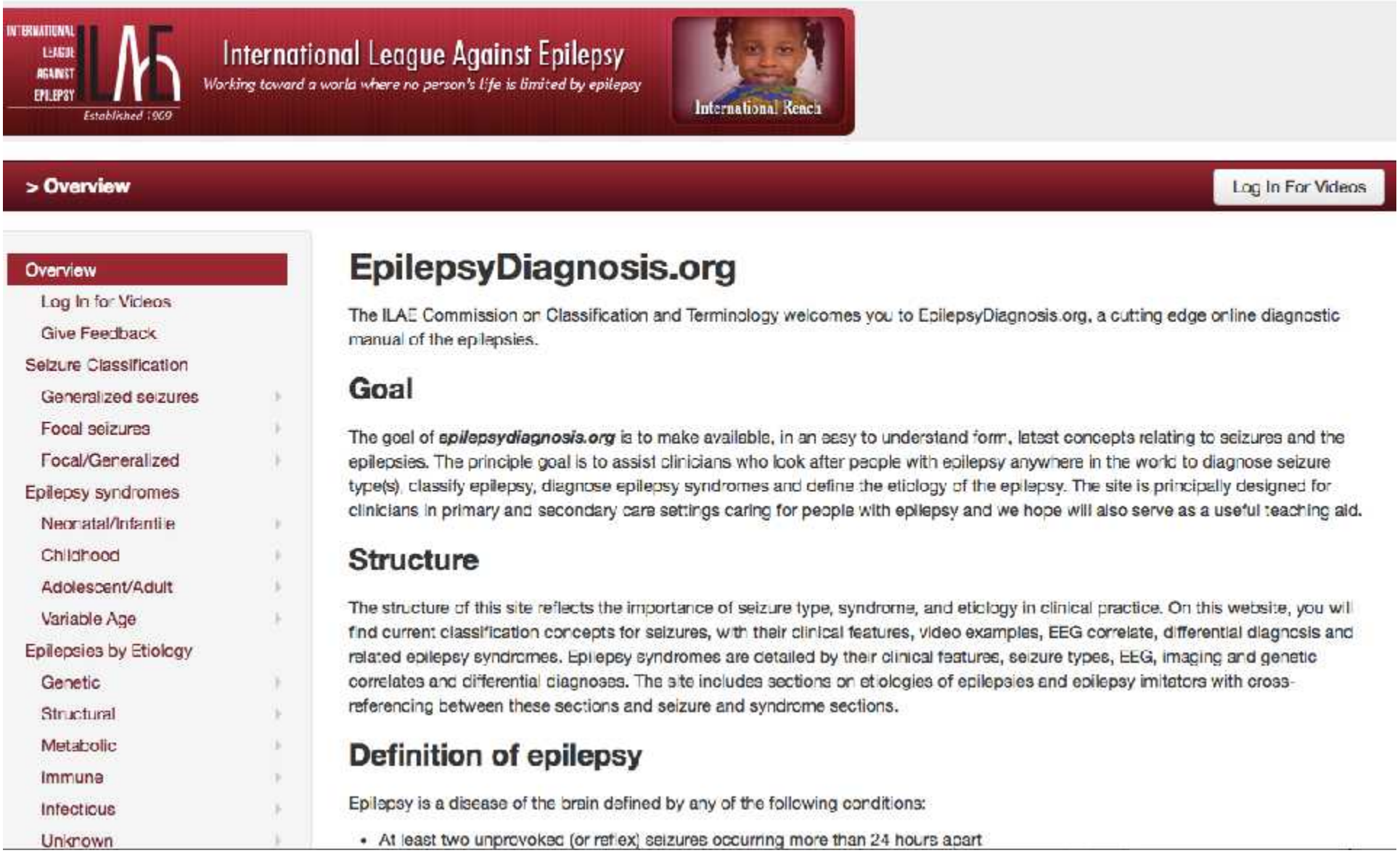
CAE pair; Lennox 1950


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



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.

ABSENCE - TYPICAL

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

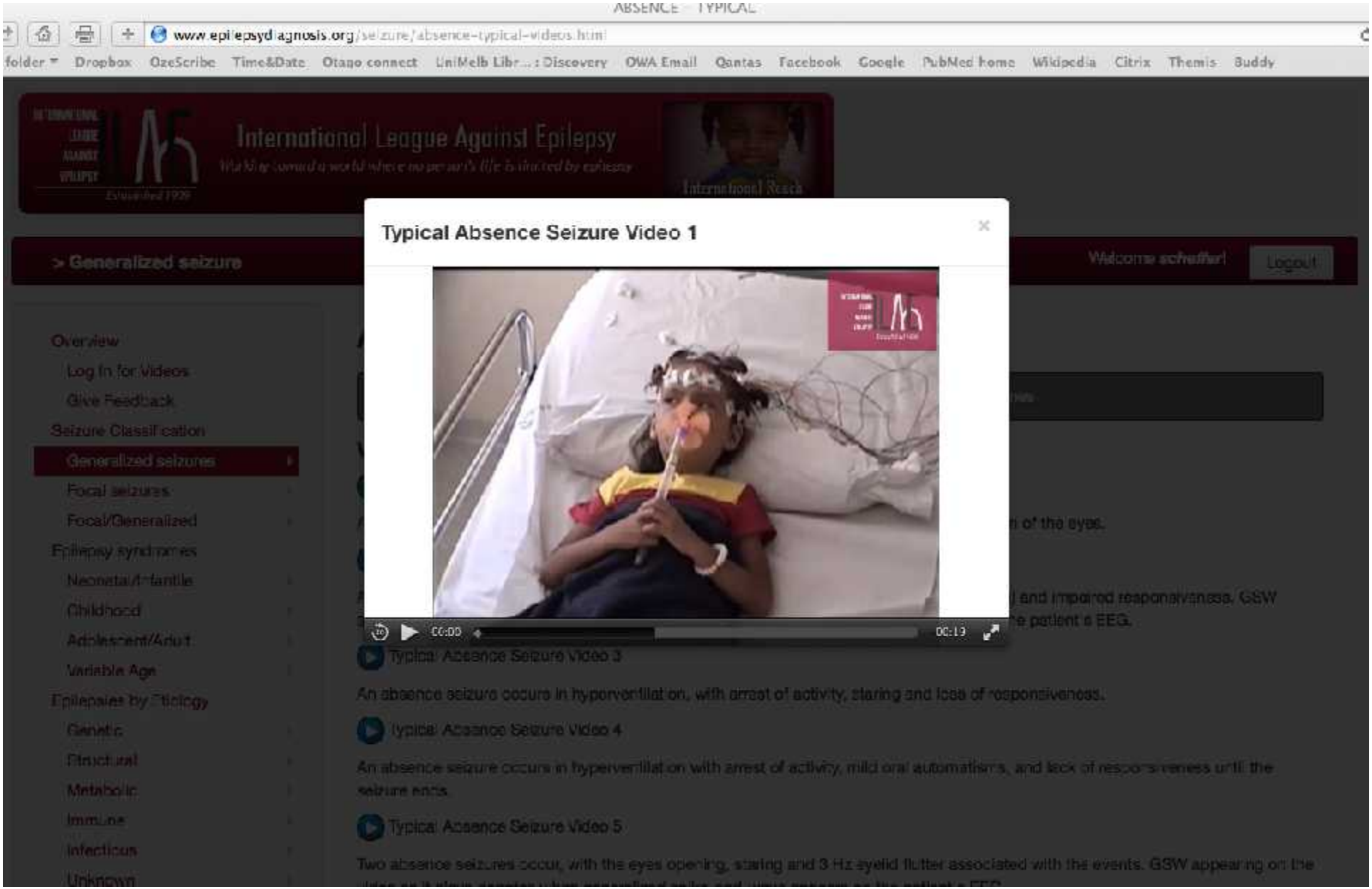
An absence seizure occurs in hyperventilation, with arrest of activity, staring and loss of responsiveness.

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.


ABSENCE - TYPICAL
www.epilepsydiagnosis.org/seizure/absence-typical-videos.html
International League Against Epilepsy
Typical Absence Seizure Video 1
00:00
00:13
> Generalized seizure
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
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

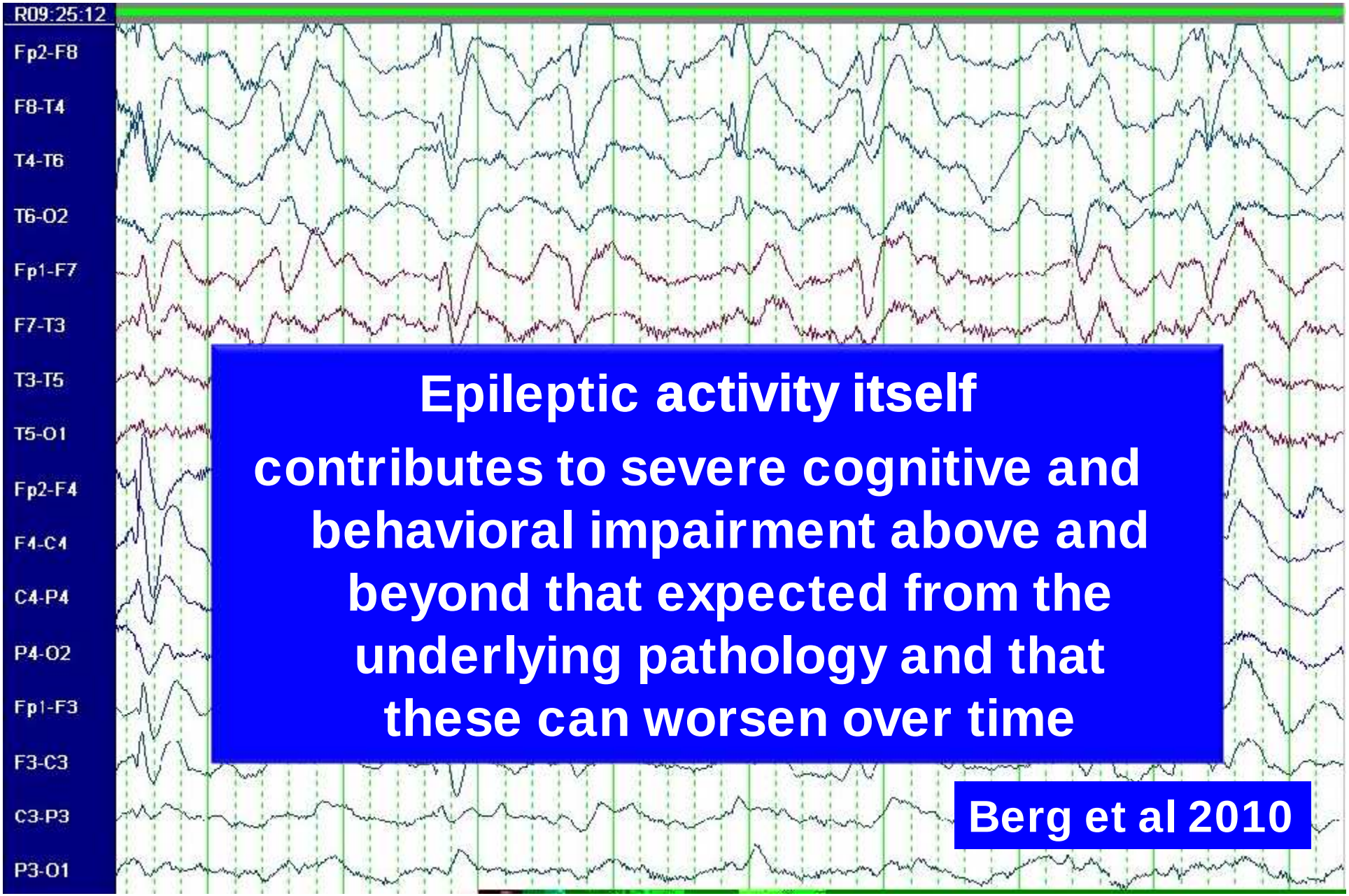
EPILEPSY IMITATORS

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



Developmental *and/or* Epileptic Encephalopathy

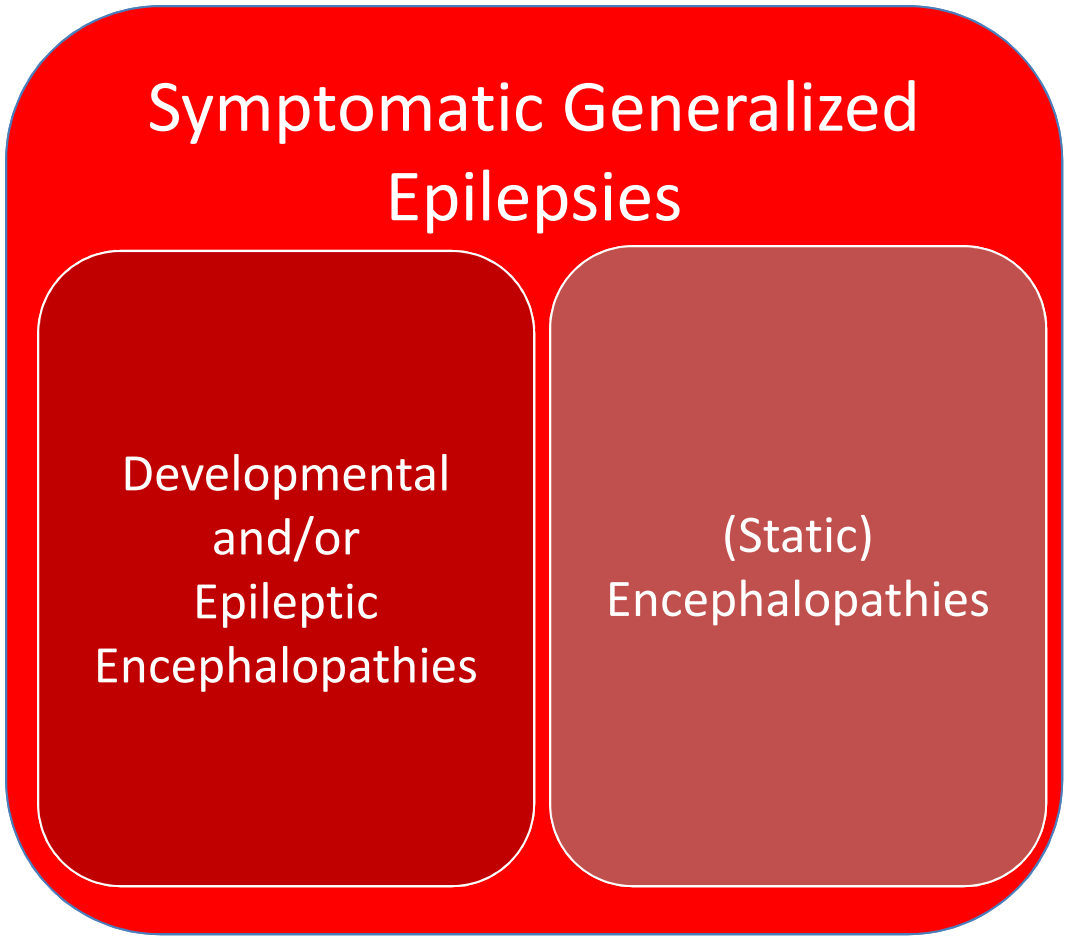
- For many encephalopathies, there is a developmental component *independent* of the epileptic encephalopathy
- Developmental delay may precede seizure onset
- Co-morbidities
  eg. cerebral palsy, autism spectrum disorder, intellectual disability
- Outcome poor even though seizures stop
  eg. KCNQ2,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