CONFLICTS OF INTEREST

• None
HISTORY OF THE ILAE CLASSIFICATION SYSTEM

Early 1900
• In 1909, the ILAE was founded.
• In the 1960s, a classification system was purposed by Herni Gastaut.

1980s
• 1981: Anatomy based classification.
  • Partial versus generalized
  • Simple versus complex
• The landmark 1985 “Classification of Epilepsies and Epileptic Syndromes” published.

2017
• In 2013, a draft document was submitted to the public for revision recommendations.
• In 2017, the ILAE classification system was published.
WHY WERE THE ILAE CRITERIA REVISED?

• Over the past 30 years, our understanding of the pathophysiology and etiology of seizure has expanded.

• It allows for more precise communication for both physicians and researchers.

• The 2017 classification can also help guide prognosis and potential therapies.

• Provides words for the patient’s to describe their disease.

• Clarify awareness during a seizure.
2017 ILAE CLASSIFICATION

• Multitiered classification system based on the knowledge available. The levels are as follows:
  • First level: Seizure type
  • Second level: Epilepsy type
  • Third level: Epilepsy syndrome

• At all levels, it reminds the practitioner to question the etiology.

• It is not meant for non-epileptic events.
  • Ex) syncope, movement disorders, psychogenic non-epileptic attacks, arrhythmias, etc.
OVERVIEW

Seizure Type

Epilepsy Type

Epilepsy Syndrome

Co-morbidities

Etiologies:
- Genetic
- Structural
- Infectious
- Immune
- Metabolic
- Unknown
LEVEL 1: SEIZURE TYPE

• Classified into categories:
  • Focal
  • Generalized
  • Unknown

• This may be the maximal level if the patient has only had a single seizure.

• If known, discuss whether or not awareness was impaired.
SEIZURE TYPE

**Focal**
- Aware versus Impaired Awareness
- Motor versus Non-Motor
- Focal to bilateral tonic-clonic

**Generalized**
- Motor
  - Ex) Tonic-clonic
- Non-Motor
  - Ex) Absence

**Unknown**
- Motor
- Non-motor

**Unclassified***
MOTOR
- Automatism
- Atonic
- Clonic
- Epileptic spasms
- Hyperkinetic
- Myoclonic
- Tonic
- Tonic-clonic
- Myoclonic-tonic-clonic
- Myoclonic-atonic

NON-MOTOR
- Behavioral arrest
- Autonomic
- Cognitive
- Emotional
- Sensory

Use the dominant classifier to describe the seizure.
If a person has a sensation of déjà vu, what term would be best used to describe this sensation?

A. Cognitive
B. Automatism
C. Emotional
D. Hyperkinetic
E. Behavioral Arrest
COGNITIVE

• Cognitive replaces the terms “psychic” and “dyscognitive”.

  • Refers to cognitive impairments such as aphasia
  • Refers to positive phenomena such as déjà vu, illusions, hallucinations and jamais vu
<table>
<thead>
<tr>
<th>Old Term</th>
<th>New Term</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unconscious</td>
<td>Impaired awareness</td>
</tr>
<tr>
<td>Partial</td>
<td>Focal</td>
</tr>
<tr>
<td>Simple partial</td>
<td>Focal aware</td>
</tr>
<tr>
<td>Complex partial</td>
<td>Focal impaired awareness</td>
</tr>
<tr>
<td>Dyscognitive</td>
<td>Focal impaired awareness</td>
</tr>
<tr>
<td>Psychic</td>
<td>Cognitive</td>
</tr>
<tr>
<td>Secondary generalization</td>
<td>Focal to bilateral tonic-clonic</td>
</tr>
</tbody>
</table>
LEVEL 2: EPILEPSY TYPE

- **Focal Epilepsy**
  - Unifocal
  - Multifocal
    - Impact on awareness should be noted

- **Generalized Epilepsy**
  - Absence, myoclonic, tonic, tonic-clonic and atonic

- **Combined Focal and Generalized**
  - On EEG, need to see independent generalized and focal discharges
    - Ex) Dravet syndrome and epilepsy of the Lennox-Gastaut type

- **Unknown**
  - Ex) Patient with several generalized convulsions per history but normal EEG recordings
A 25-year-old man developed seizures that can be described as clonic right face twitching that lasts 10-30 seconds. He has maintained awareness, able to communicate appropriately and perform motor tasks during these events.

This was previously called a simple partial seizure.

Now, these paroxysmal events are called **focal motor seizure with retained awareness**.
A 30-year-old woman has seizures described as starting with the sudden inability to understand language followed by confusion and left arm clonic jerking.

What is this called with the 2017 ILAE Classification?

**Focal impaired awareness cognitive seizure** (progressing to left arm clonic)
LEVEL 3: EPILEPSY SYNDROMES

• Incorporate seizure type, EEG and imaging features that tend to cluster together
  • Age-dependent features
  • Prognosis
  • Seizure triggers
  • Diurnal variation
  • Any distinctive intellectual dysfunction
  • Associated psychiatric co-morbidities
  • Treatment implications
ETIOLOGIES

- Genetic
- Structural
- Infectious
- Metabolic
- Immune
- Unknown
GENETIC

- Monogenetic
  - Inherited
  - De novo

- Complex (polygenetic)
  - May require an environmental factor to trigger

- Formerly called Idiopathic and Primary

- The well-known Generalized Genetic Epilepsies are:
  - Childhood Absence Epilepsy
  - Juvenile Absence Epilepsy
  - Juvenile Myoclonic Epilepsy
  - Generalized Tonic-Clonic Seizures Alone
• Visual abnormality seen on neuroimaging that is suspected to be causing the patient’s seizures

• Caused by many etiologies: strokes, tumors, trauma, infections, genetic (Ex -vascular malformations), cortical dysplasia of development
• Most common worldwide etiology
  • Meningitis and encephalitis
  • Neurocysticercosis
  • Tuberculosis
  • HIV
  • Cerebral malaria
  • Zika virus
  • And many other causes
METABOLIC

• Epilepsy that is a direct result of a known or presumed metabolic disorder

• Examples:
  • Porphyria
  • Uremia
  • Aminoacidopathies
  • Cerebral Folate Deficiency
  • Many more

• Epilepsy caused by metabolic etiologies require specific treatments to obtain seizure control.
IMMUNE

- Epilepsies caused by an underlying immune disorder that causes autoimmune-mediated central nervous system inflammation.
  - Examples:
    - NMDA (N-methyl-D-aspartate) receptor encephalitis
    - Anti-LGI1 encephalitis

- Treatment needs concurrent immunosuppression
UNKNOWN ETIOLOGY
ILAE SEIZURE CLASSIFICATION

Co-morbidities

Seizure Type

Epilepsy Type

Epilepsy Syndrome

Etiologies:
- Genetic
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OTHER REVISED TERMS

Benign

- Replace with either:
  - Self-limited
  - Spontaneous resolution
- Pharmaco-responsive
  - Easily controlled with the appropriate anti-epileptic drug

Removal:

- Malignant
- Catastrophic
CLINICAL PRACTICE: CASE SCENARIOS
Case # 1: 6-year-old boy with ~30 seconds of facial twitching at night. He has normal intelligence. MRI negative. EEG below.

What type of epilepsy does he have?
• Benign Rolandic Epilepsy also called Benign Epilepsy with Centro-Temporal Spikes (BECTS)

• What is this called according to the 2017 ILAE classification system? Most of the patient’s seizures occur in sleep. When these events occur during the daytime, there is no loss of awareness.

• Focal motor seizure without impaired awareness
Case #2: A 7-year-old child develops staring spells. Her school performance is slipping. Her MRI is normal. EEG obtained above while the patient was blowing into a pinwheel. What type of epilepsy does she have?
• Childhood Absence Epilepsy – which is one of the well-recognized Idiopathic Generalized Epilepsy (IGE); now referred to as Genetic Generalized Epilepsy (GGE).

• What is this type of epilepsy called according to the 2017 ILAE classification?

• Non-motor generalized epilepsy with impaired awareness

• Technically, the modifier of awareness is not needed in a generalized seizure.
ILAE NAME FOR THE “GRAND MAL” CONVULSION

• Generalized tonic-clonic seizure

• Because awareness is known to be impaired before or contemporaneously with the stiffening phase, the modifier of “loss of awareness” is not necessary.
Case # 3: A child was diagnosed with epilepsy of the Lennox-Gastaut type. EEG shows runs of slow 2-2.5Hz spike and wave complexes. Seizure types includes absence and others. What would this patient’s absence be called in the 2017 Classification?

Atypical absence seizures
The child with Lennox Gastaut later has a seizure described as whole body stiffening. What is this called in the 2017 Classification?

Generalized tonic seizure
CASE # 4

• A 24-year-old man with seizures described as “the hair on my arm stands on edge” with a feeling of being flushed. He is aware throughout the seizure. What is the new 2017 ILAE classification?

• Focal aware autonomic seizures
CASE # 5

- A 14-month-old girl has sudden flexion of her arms with head flexing forward for ~3 seconds. These repeat in clusters. EEG clips are shown below. What would you call the episodes of sudden flexion according to the 2017 ILAE Classification?
• Focal Epileptic Spasms

• Old term = infantile spasms


• Thank you to Dr. Alfred Frontera for use of imaging.