Types of Seizures and Common Epilepsy Syndromes in Children

February 26, 2019
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

Proposal for Revised Classification of Epilepsies and Epileptic Syndromes

Commission on Classification and Terminology of the International League Against Epilepsy
<table>
<thead>
<tr>
<th>Idiopathic Focal</th>
<th>Symtomatic Focal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal Seizures</td>
<td>Focal seizures</td>
</tr>
<tr>
<td>Normal intellect/PE</td>
<td>Gross/subtle cognitive and/or exam abnormalities</td>
</tr>
<tr>
<td>Normal EEG background</td>
<td>Abnormal neuroimaging</td>
</tr>
<tr>
<td>Normal neuroimaging</td>
<td><strong>Variable prognosis</strong></td>
</tr>
<tr>
<td><strong>Good Prognosis</strong></td>
<td>Ex: Temporal lobe epilepsy, Post-traumatic epilepsy</td>
</tr>
<tr>
<td>Ex: Benign Rolandic Epilepsy</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Idiopathic Generalized</th>
<th>Symtomatic Generalized</th>
</tr>
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<tbody>
<tr>
<td>Generalized Seizures</td>
<td>Generalized seizures (many types)</td>
</tr>
<tr>
<td>Normal intellect/PE</td>
<td>Subnormal intellect/Abnormal exam</td>
</tr>
<tr>
<td>Normal EEG background</td>
<td>Abnormal EEG background</td>
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<tr>
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<td><strong>Poor Prognosis</strong></td>
</tr>
<tr>
<td>Ex: Childhood Absence, Juvenile Myoclonic Epilepsy</td>
<td>Ex: Lennox-Gastaut syndrome</td>
</tr>
</tbody>
</table>

Source: Joshi and Shellhaas 2014
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 in 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

*‡Anne T. Berg, †Samuel F. Berkovic, §Martin J. Brodie, †Jeffrey Buchhalter, ‡**J. Helen Cross, ††Walter van Emde Boas, ‡‡Jerome Engel, §§Jacqueline French, ¶¶Tracy A. Glauser, ##Gary W. Mathern, ###Solomon L. Moshé, †Douglas Nordli, †††Perrine Plouin, and ‡Ingrid E. Scheffer
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)
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
Some Common Childhood Epilepsy Syndromes

- Childhood Absence Epilepsy
- Benign Focal Epilepsies of Childhood
- Juvenile Myoclonic Epilepsy
- Infantile Spasms/West Syndrome
- Lennox Gastaut Syndrome
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
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.
Diagnosis?
**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
EEG in Absence Epilepsy
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
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.
Diagnosis?
Juvenile Myoclonic Epilepsy

- Onset: 12-18 years
- Myoclonus
  - 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 recommended
- Treatment:
  - Valproic acid used to be the drug of choice
  - Recently levetiracetam, lamotrigine, zonisamide
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.
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.
Diagnosis?
Benign Rolandic Epilepsy
(Benign Childhood Epilepsy with Central-Temporal Spikes)

- Most common focal epilepsy in 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
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
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.
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.
Diagnosis?
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
West Syndrome

- Described by Dr. West in his own child
- Triad of:
  - Infantile spasms
  - Hypasarc[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
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)
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)
PNEE

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

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### ILAE 2017 Classification of Seizure Types Basic Version

<table>
<thead>
<tr>
<th>Focal Onset</th>
<th>Generalized Onset</th>
<th>Unknown Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aware</strong></td>
<td><strong>Motor</strong></td>
<td><strong>Motor</strong></td>
</tr>
<tr>
<td><strong>Impaired Awareness</strong></td>
<td><strong>Tonic-clonic</strong></td>
<td><strong>Tonic-clonic</strong></td>
</tr>
<tr>
<td><strong>Motor</strong></td>
<td><strong>Other motor</strong></td>
<td><strong>Other motor</strong></td>
</tr>
<tr>
<td><strong>Non-Motor</strong></td>
<td><strong>Non-Motor (Absence)</strong></td>
<td><strong>Non-Motor</strong></td>
</tr>
<tr>
<td>focal to bilateral tonic-clonic</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Unclassified</strong></td>
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</tbody>
</table>

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.

ILAE 2017 Classification of Seizure Types Expanded Version

- **Focal Onset**
  - **Aware**
  - **Impaired Awareness**
  - **Motor Onset**
    - automatisms
    - atonic
    - clonic
    - epileptic spasms
    - hyperkinetic
    - myoclonic
    - tonic
  - **Non-Motor Onset**
    - autonomic
    - behavior arrest
    - cognitive
    - emotional
    - sensory

- **Generalized Onset**
  - **Motor**
    - tonic-clonic
    - clonic
    - tonic
    - myoclonic
    - myoclonic-tonic-clonic
    - myoclonic-atonic
    - tonic
    - epileptic spasms
  - **Non-Motor (absence)**
    - typical
    - atypical
    - myoclonic
    - eyelid myoclonia
  - **focal to bilateral tonic-clonic**

- **Unknown Onset**
  - **Motor**
    - tonic-clonic
    - epileptic spasms
  - **Non-Motor**
    - behavior arrest
  - **Unclassified**

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.

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

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**
But this is currently impossible with our limited understanding

**Anatomy**
- Temporal
- Frontal
- Parietal
- Occipital
- Diencephalic
- Brainstem

**Networks**
- Neocortical
- Limbic
- Thalamo-Cortical
- Brainstem

**Practical, by:**
- AED response
- Surgical target
- Disabling
- EEG pattern
- Many others

**Modify Existing**
- 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

<table>
<thead>
<tr>
<th>Symptoms + Signs</th>
<th>Seizure Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Automatisms</td>
<td>Focal impaired awareness seizure</td>
</tr>
<tr>
<td>Autonomic</td>
<td>Absence seizure</td>
</tr>
</tbody>
</table>

Operators:
- One-to-many
- Many-to-one
### Key Seizure Signs and Symptoms?

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

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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
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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

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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
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New Seizure Types

New Focal Seizures

Motor
- atonic
- automatisms
- clonic
- epileptic spasms
- hyperkinetic
- myoclonic
- tonic

Non-Motor
- behavior arrest
  - (autonomic)
  - (cognitive)
- emotional
  - (sensory)

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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- **Update word usage for greater public clarity**
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- Standardize common descriptors to describe seizures
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## Wording Changes

<table>
<thead>
<tr>
<th>OLD TERM</th>
<th>NEW TERM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unconscious (still used, not in name)</td>
<td>Impaired awareness (surrogate)</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 (word discontinued)</td>
<td>Focal impaired awareness</td>
</tr>
<tr>
<td>Psychic</td>
<td>Cognitive</td>
</tr>
<tr>
<td>Secondarily generalized tonic-clonic</td>
<td>Focal to bilateral tonic-clonic</td>
</tr>
<tr>
<td>Arrest, freeze, pause, interruption</td>
<td>Behavior arrest</td>
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- Update the 2001 glossary of seizure terms
- Standardize common descriptors to describe seizures
- Map old to new terms
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

- 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
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

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- Classify seizures of unknown onset
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<table>
<thead>
<tr>
<th>WORD</th>
<th>DEFINITION</th>
<th>SOURCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>absence, typical</td>
<td>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, which also can be encountered with focal onset seizures.</td>
<td>Adapted from 11</td>
</tr>
<tr>
<td>absence, atypical</td>
<td>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</td>
<td>Adapted from Dreifuss 1</td>
</tr>
<tr>
<td>arrest</td>
<td>see behavioral arrest</td>
<td>new</td>
</tr>
<tr>
<td>atonic</td>
<td>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.</td>
<td>11</td>
</tr>
<tr>
<td>automatism</td>
<td>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.</td>
<td>11</td>
</tr>
</tbody>
</table>
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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- 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.

<table>
<thead>
<tr>
<th>Cognitive</th>
<th>Automatisms</th>
</tr>
</thead>
<tbody>
<tr>
<td>acalculia</td>
<td>aggression</td>
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<td>versive</td>
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<td>gustatory</td>
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<td>erection</td>
<td>hot-cold sensations</td>
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<td>olfactory</td>
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<td>somatosensory</td>
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<td>hyper/hypoventilation</td>
<td>vestibular</td>
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<td>nausea or vomiting</td>
<td>visual</td>
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<td>pallor</td>
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<td>palpitations</td>
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<td>piloerection</td>
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<td>tachycardia</td>
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<tr>
<td>right</td>
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<tr>
<td>bilateral</td>
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<th>Old Term for Seizure</th>
<th>New Term for Seizure [choice] (optional)</th>
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</thead>
<tbody>
<tr>
<td>absence</td>
<td>(generalized) absence</td>
</tr>
<tr>
<td>absence, atypical</td>
<td>(generalized) absence, atypical</td>
</tr>
<tr>
<td>absence, typical</td>
<td>(generalized) absence, typical</td>
</tr>
<tr>
<td>akinetic</td>
<td>[focal/generalized] atonic</td>
</tr>
<tr>
<td>astatic</td>
<td>[focal/generalized] atonic</td>
</tr>
<tr>
<td>atonic</td>
<td>[focal/generalized] atonic</td>
</tr>
<tr>
<td>aura</td>
<td>focal aware</td>
</tr>
<tr>
<td>clonic</td>
<td>[focal/generalized] clonic</td>
</tr>
<tr>
<td>complex partial</td>
<td>focal impaired awareness</td>
</tr>
<tr>
<td>convulsion</td>
<td>[focal/generalized] motor [tonic-clonic, tonic, clonic], focal to bilateral tonic-clonic</td>
</tr>
</tbody>
</table>
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*. 
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 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.
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.
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.
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.

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

Old = atypical absence seizures
New = atypical absence 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.
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.
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.”
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.

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

**Old** = simple partial autonomic seizures

**New** = focal aware 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.
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.

Old = myoclonic astatic seizures
New = myoclonic-atonic seizures
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.
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.
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**

Old = infantile spasms (focality not specified)
New = focal epileptic spasms
10. A 75 year-old man reports an internal sense of body trembling. No other information is available.
“Words, words, words, I’m so sick of words!”
Eliza Doolittle, *My Fair Lady*
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. Moshe, 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

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. Moshe, ‡‡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

Focal Onset
- Aware
  - Impaired Awareness
- Motor Onset
  - Nonmotor Onset
    - focal to bilateral tonic-clonic

Generalized Onset
- Motor
  - Tonic-clonic
  - Other motor
- Nonmotor (Absence)

Unknown Onset
- Motor
  - Tonic-clonic
  - Other motor
- Nonmotor
  - Unclassified
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

**Focal Onset**
- **Aware**
- **Impaired Awareness**

  **Motor Onset**
  - automatisms
  - atonic
  - clonic
  - epileptic spasms
  - 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

<table>
<thead>
<tr>
<th>Focal Onset</th>
<th>Generalized Onset</th>
<th>Unknown Onset</th>
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<tbody>
<tr>
<td><strong>Aware</strong></td>
<td><strong>Impaired</strong></td>
<td><strong>Motor</strong></td>
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<tr>
<td>focal to bilateral tonic-clonic</td>
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<td><strong>Unclassified</strong></td>
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</tbody>
</table>
Note
When a seizure type begins with “focal, generalized or absence” then the word “onset” can be presumed
Terms no longer in use

- Complex partial
- Simple partial
- Partial
- Psychic
- Dyscognitive
- Secondarily generalized tonic-clonic
Instruction manual for the ILAE 2017 operational classification of seizure types


Epilepsia, **(*):1–12, 2017
doi: 10.1111/epi.13671
Table 1. Common descriptors of behaviors during and after seizures (alphabetically)

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</tr>
<tr>
<td>Pleasure</td>
<td>Sensory</td>
</tr>
<tr>
<td>Autonomic</td>
<td>Auditory</td>
</tr>
<tr>
<td>Asystole</td>
<td>Gustatory</td>
</tr>
<tr>
<td>Bradycardia</td>
<td>Hot-cold sensations</td>
</tr>
<tr>
<td>Erection</td>
<td>Olfactory</td>
</tr>
<tr>
<td>Flushing</td>
<td>Somatosensory</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Vestibular</td>
</tr>
<tr>
<td>Hyper/hypoventilation</td>
<td>Visual</td>
</tr>
<tr>
<td>Nausea or vomiting</td>
<td></td>
</tr>
<tr>
<td>Pallor</td>
<td></td>
</tr>
<tr>
<td>Palpitations</td>
<td>Laterality</td>
</tr>
<tr>
<td>Piloerection</td>
<td>Left</td>
</tr>
<tr>
<td>Respiratory changes</td>
<td>Right</td>
</tr>
<tr>
<td>Tachycardia</td>
<td>Bilateral</td>
</tr>
</tbody>
</table>

*Note*
Clarify features of seizures but do not define unique seizure types

Free text descriptors encouraged
Table 2. Glossary of terms

<table>
<thead>
<tr>
<th>Word</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absence, typical</td>
<td>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 seizure of generalized onset. The word is not synonymous with a blank stare.</td>
</tr>
</tbody>
</table>

Table 3. Summary of rules for classification

1. Onset: Decide whether the onset is generalized or focal.
2. Awareness: Decide whether awareness is preserved or lost.
3. Impaired consciousness: Decide whether consciousness is impaired or not.
4. Onset: Decide whether the onset is focal or generalized.
5. Behavior: Decide whether the behavior shows an orofacial or axial pattern.
6. Motor/no motor: Decide whether the motor component is present or absent.

Table 4. Abbreviations for the most important seizure types

<table>
<thead>
<tr>
<th>Seizure type</th>
<th>Abbreviations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal aware seizure</td>
<td>FAS</td>
</tr>
<tr>
<td>Focal impaired awareness seizure</td>
<td>FLAS</td>
</tr>
<tr>
<td>Focal motor seizure</td>
<td>FMS</td>
</tr>
<tr>
<td>Focal nonmotor seizure</td>
<td>FNMS</td>
</tr>
<tr>
<td>Focal epileptic spasm</td>
<td>FES</td>
</tr>
<tr>
<td>Focal to bilateral tonic–clonic seizure</td>
<td>FBTCS</td>
</tr>
<tr>
<td>Generalized tonic–clonic seizure</td>
<td>GTCS</td>
</tr>
<tr>
<td>Generalized absence seizure</td>
<td>GAS</td>
</tr>
<tr>
<td>Generalized motor seizure</td>
<td>GMS</td>
</tr>
<tr>
<td>Generalized epileptic spasm</td>
<td>GES</td>
</tr>
<tr>
<td>Unknown onset tonic–clonic seizure</td>
<td>UTCS</td>
</tr>
</tbody>
</table>

Dacrycistic
Dialectic
Drop attack
Fencer's posture
(Focal) [aware or impaired awareness]
(Focal) [generalized] tonic
(Focal) [generalized] atonic,
(figure-of-4)

(left) may occur. Grimacing, head nodding, or sudden eye closure may occur in clusters. Infantile spasms are the best known

the following conditions: (1) At least two unprovoked seizures; (2) one unprovoked (or reflex) seizure and a 10-year general occurrence rate (at least 60%). Often two or more at once, (3) diagnosis of epilepsy is resolved for individuals who had an age-dependent
Etiology

- Tuberous Sclerosis
- GLUT1 deficiency

Seizure types

- Focal onset
- Generalized onset
- Unknown onset

Etiology

- Structural
- Genetic
- Infectious
- Metabolic
- Immune
- Unknown
• 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
Epilepsy types

- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Seizure types

- Focal onset
- Generalized onset
- Unknown onset

Etiology

- Structural
- Genetic
- Infectious
- Metabolic
- Immune
- Unknown
Old term
‘Idiopathic Generalized Epilepsies’

Idiopathic Generalized Epilepsies

- Childhood Absence Epilepsy
- Juvenile Absence Epilepsy
- Juvenile Myoclonic Epilepsy
- Generalized Tonic-Clonic Seizures Alone
Genetic *versus* idiopathic

- ‘Idiopathic’ = presumed hereditary predisposition

- **Genetic ≠ 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

Epilepsy types
- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Epilepsy Syndromes

Etiology
- Structural
- Genetic
- Infectious
- Metabolic
- Immune
- Unknown

Co-morbidities
Epilepsy syndromes

• There are no approved ILAE epilepsy syndromes
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 correlates, 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

https://www.epilepsydiagnosis.org
ABSENCE - TYPICAL

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 automatms 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 5 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.
Typical Absence Seizure Video 1

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

Typical Absence Seizure Video 2

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 3

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.
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. Impaired upper airways obstruction
   h. Orthostatic intolerance
      i. Long QT and cardiac syncope
   j. Hypercyanotic spells

3. BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS
   a. Daydreaming / inattention
   b. Intimacy gratification
   c. Eliciting 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
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 2010
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, e.g. cerebral palsy, autism spectrum disorder, intellectual disability.
- Outcome poor even though seizures stop, *e.g.* 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

Symptomatic Generalized Epilepsies

Developmental and/or Epileptic Encephalopathies

(Static) Encephalopathies
ILAE Classification of the Epilepsies

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

- New classification framework will
  - Change the approach to diagnosis in the clinic
  - Be applied to patients and guide management

- Updates terminology to reflect current thinking
  - Scientific advances
Torbjörn Tomson, Emilio Perucca, Ingrid Scheffer, Jackie French, Yue-Hua Zhang, Satish Jain, Gary Mathern, Sam Wiebe, Edouard Hirsch, Sameer Zuberi, Nico Moshe