

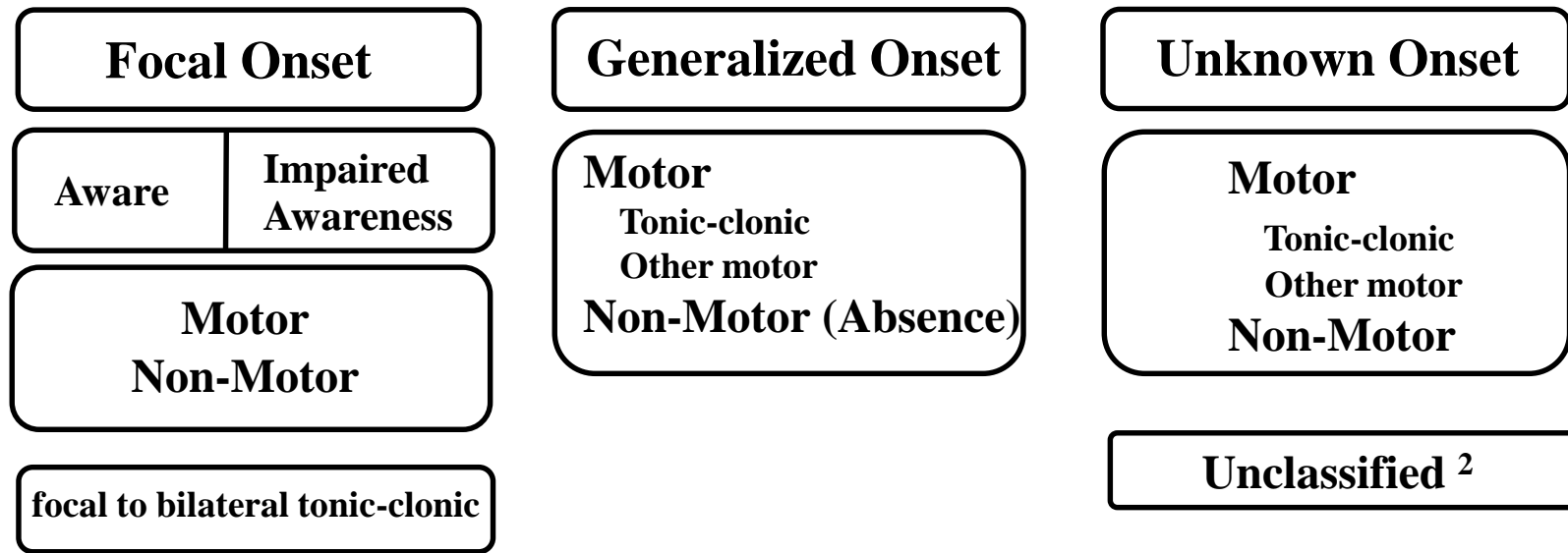
The 2017 ILAE Classification of Seizures

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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 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 Version¹

Focal Onset

Aware

Impaired
Awareness

Motor Onset

automatisms
atonic²
clonic
epileptic spasms²
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 spasms²

Non-Motor (absence)

typical
atypical
myoclonic
eyelid myoclonia

Unknown Onset

Motor

tonic-clonic
epileptic spasms
Non-Motor
behavior arrest

Unclassified³

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

INTERNATIONAL CLASSIFICATION OF SEIZURES 1981

Partial Seizures (start in one place)

Simple (no loss of consciousness or 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

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

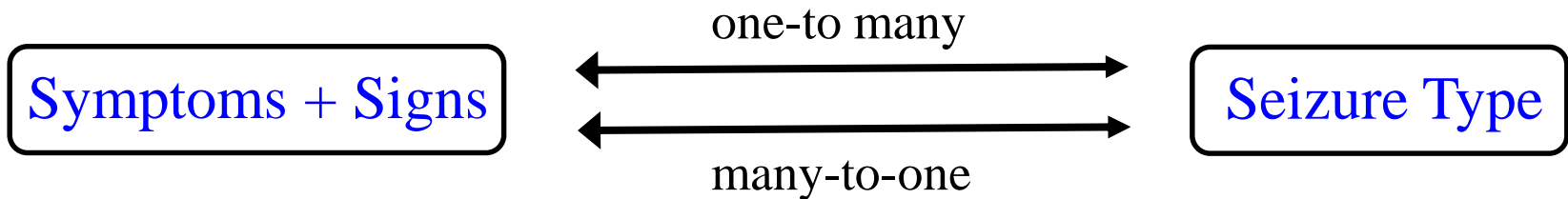
Modify Existing

1981 ILAE System
2010 ILAE update

- In the absence of fundamental knowledge, ILAE chose to extend the existing classification
- This 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

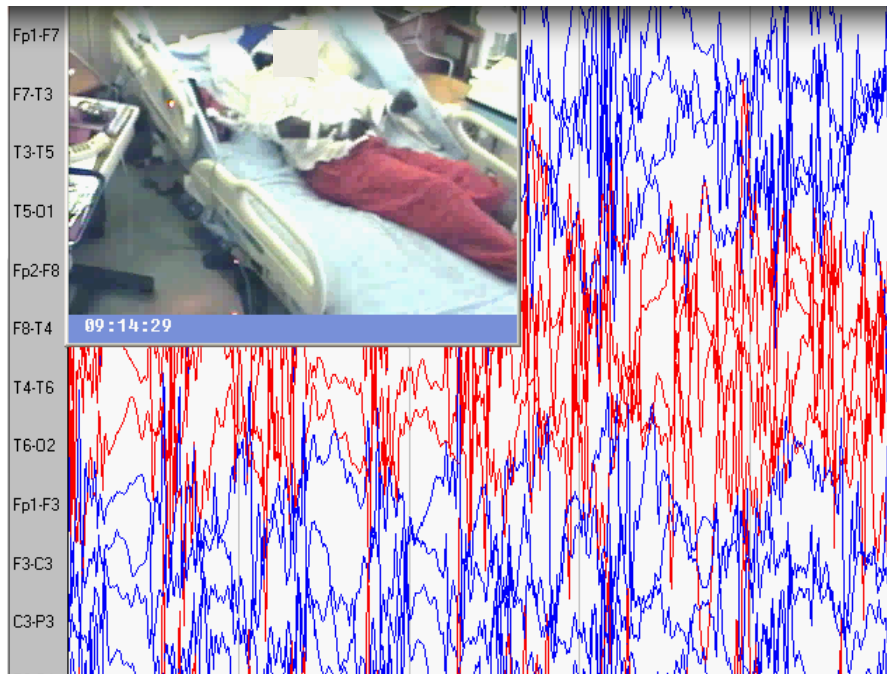


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Seizures of Unknown Onset

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

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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**

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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., Dose)
myoclonic-tonic-clonic (e.g., JME)

New combined seizures

(focal to bilateral tonic-clonic)

(parentheses) indicates prior existence, but renaming

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Wording Changes

OLD TERM	NEW TERM
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

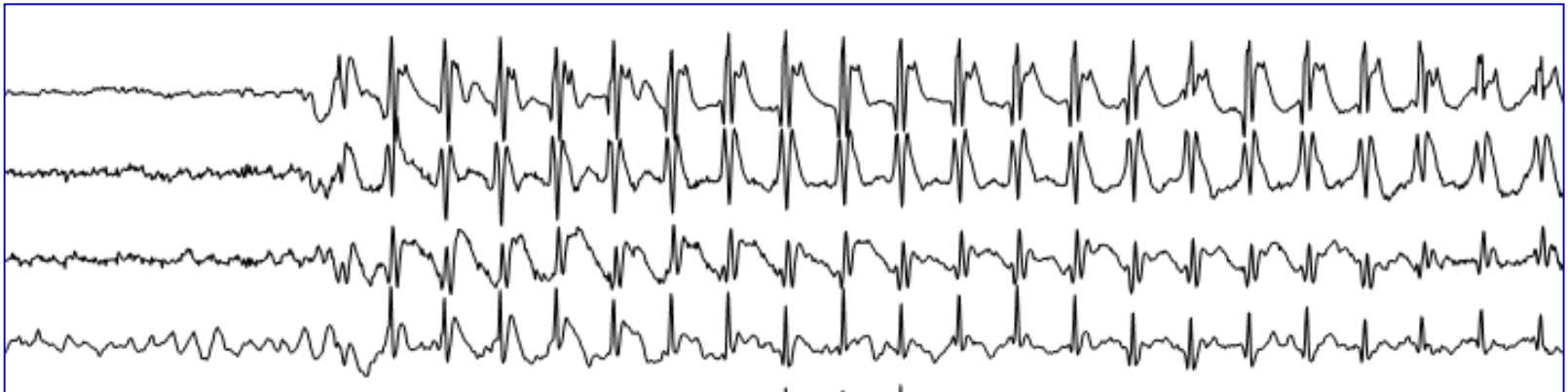
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Supportive Information

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**



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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

▶ G40 Epilepsy and recurrent seizures

- ▶ G40.0 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes
 - ▶ G40.00 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes
 - ▶ G40.001 with status epilepticus
 - ▶ G40.009 without status epilepticus
 - ▶ G40.01 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes
 - ▶ G40.011 with status epilepticus
 - ▶ G40.019 without status epilepticus
- ▶ G40.1 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes
 - ▶ G40.10 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes

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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

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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**
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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
<u>akinetic</u>	[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.

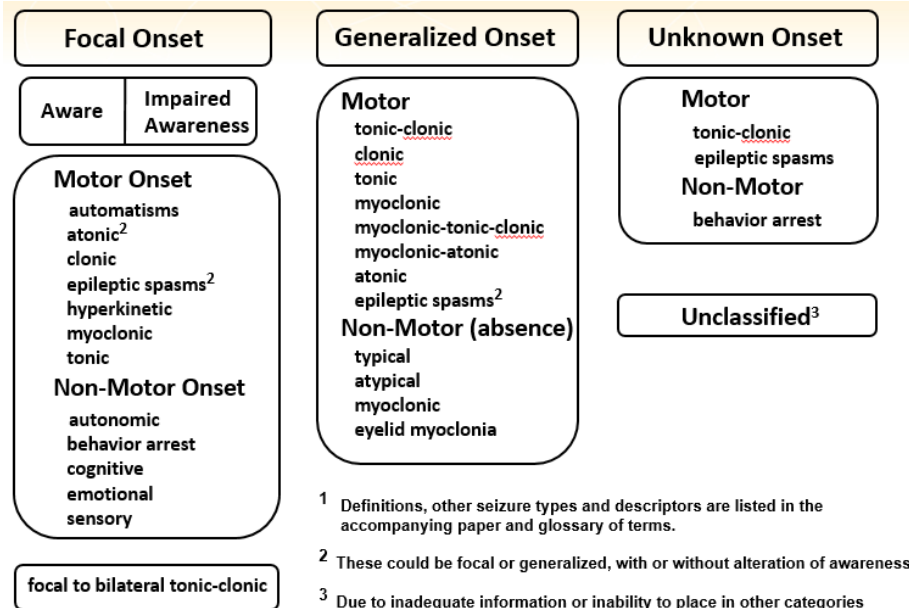
Impaired awareness at any point: A focal seizure is a *focal impaired awareness seizure* if awareness is impaired at any point during the seizure.

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)

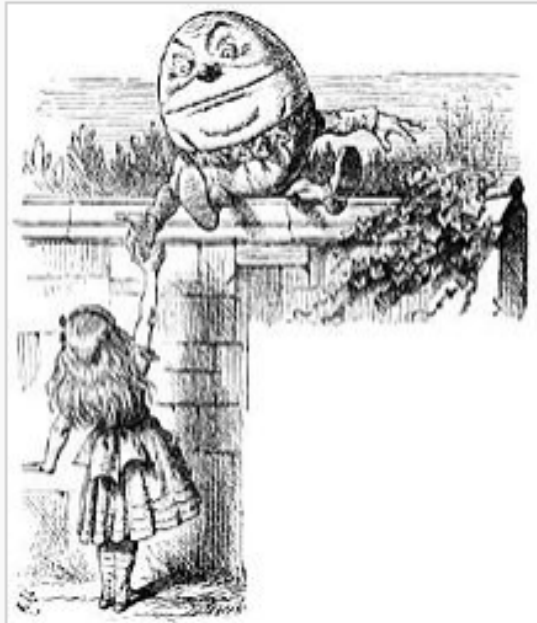
New = focal epileptic spasms

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*