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Changing How We Think About and Classify Seizures

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THE NEUROLOGICAL CHOICE

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Background

- Seizure is “a transient occurrence of signs and/or symptoms due to abnormal, excessive or synchronous neuronal activity in the brain.”
- A large number of brain cells (neurons) are activated abnormally at the same time

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Background

Why classify seizures?

- Classification of a seizure begins with historical elicitation or observation of certain symptoms or signs (semiology)
- Classifying seizure types helps guide further testing, treatment and prognosis
- Provides a common language and aids in communication and research among clinicians and medical community
- Provides common words for people with epilepsy and the general public to describe seizures

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History

- For decades terms such as “grand mal” and “petit mal” used
- In 1981 the ILAE (International League Against Epilepsy) devised a classification system
  - Divided seizures into partial and generalized onset
  - Further classified partial as simple or complex

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History

- Several drawback to this system:
  - Several important seizure types were not included
  - Lack of knowledge of the onset (the part or network of brain involved in generating the seizure) makes a seizure unclassifiable
  - Some terms used to classify seizures lack community acceptance or public understanding. (“psychic”, “complex partial”, “simple partial”, “dyscognitive”).

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- In 2017 ILAE revised classification system to specifically address these issues
- The 2017 seizure classification system presents BASIC and EXPANDED versions

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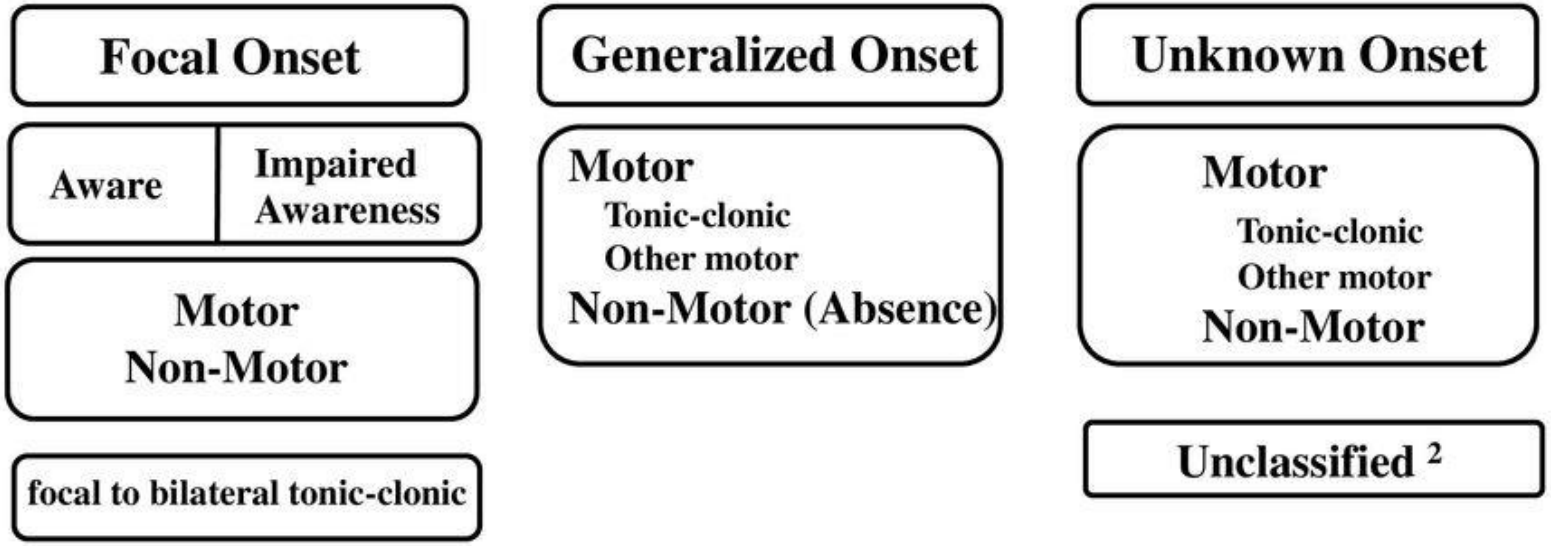
ILAE 2017 Classification of Seizure Types Basic Version [1]



[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*

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Basic Classification

- Seizures first classified by type of onset
  - Focal: originating within networks limited to one hemisphere
  - Generalized : originating at some point within and rapidly engaging bilaterally distributed networks
  - Unknown: Insufficient data to confidently classify seizure and can be classified later

Classifying seizure onset is crucial to determine treatment (medication choices, surgical candidacy?, outlook, and possible causes)

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Basic classification

- Secondly, seizures are classified by level of awareness defined as knowledge or self and environment during seizure (person may be unable to speak or respond). Does not refer to if person has awareness that a seizure occurred.
- Focal aware (replaces term simple partial)
- Focal with impaired awareness: If awareness is impaired at any point during a seizure (replaces term simple complex)

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Basic Classification

- Focal

  Aware vs impaired awareness

  motor vs nonmotor

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Basic classification

- A focal seizure can spread to both sides of the brain resulting in the classification “focal to bilateral tonic clonic seizure”. This term replaces secondarily generalized tonic clonic. “Generalized” now used to only classify seizures with generalized onset.

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Basic Classification

- Generalized onset seizures divided into:
  - –Motor
  - –Nonmotor (Absence)

  Level of awareness is not used in generalized onset seizures since the large majority of generalized seizures are associated with impaired awareness

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Basic Classification

- Unknown
  - –Motor
  - –Nonmotor

  Unclassified: seizures with patterns that do not fit into the other categories or seizures presenting insufficient information to allow for categorization

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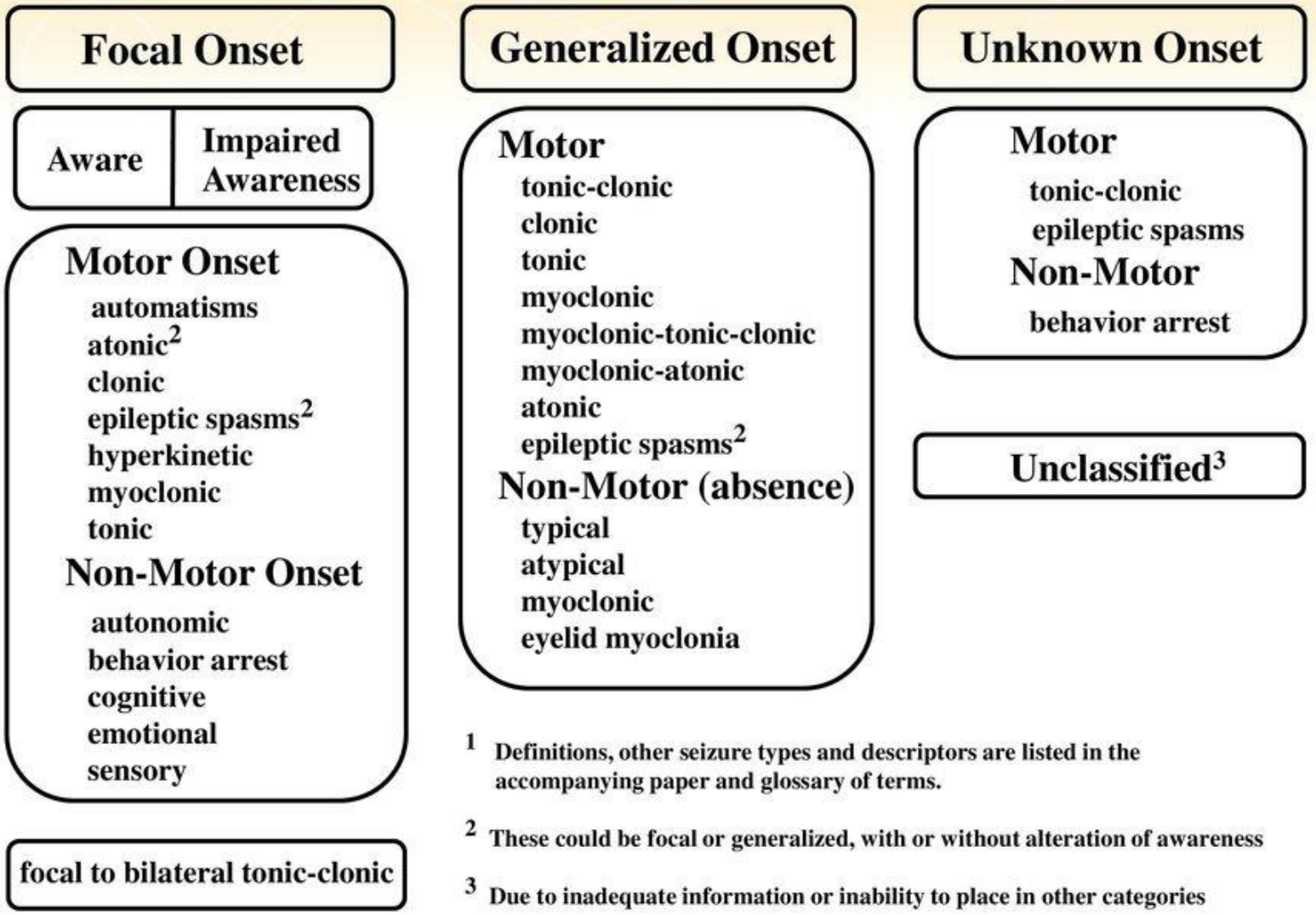
ILAE 2017 Classification of Seizure Types Expanded Version[1]



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

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Expanded classification

- Keeps the framework of the basic classification but with added seizure names as subgroups

- Classify by first most prominent motor or nonmotor symptom

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Expanded Classification

- Focal motor (aware/impaired awareness)
  - Automatisms: coordinated activity, lip smacking, hand fumbling, picking at objects, walking in circles, unbuttoning
  - Atonic: sudden loss of muscle tone “limp”, can involve head, trunk, limb, or jaw
  - Clonic: rhythmic, repetitive, jerking of one part of the body or face
  - Epileptic spasms: sudden flexion or bending of the trunk and extension or flexion of the proximal limbs, lasting seconds and may be occur in clusters.
  - Hyperkinetic: involving predominantly proximal limb or axial muscles in irregular sequential complex movements-pedaling, thrashing, rocking movements
  - Myoclonic: sudden, brief (<100 msec) involuntary single or repetitive contractions of muscle or muscle groups
  - Tonic: A sustained contraction (stiffening) of or arm, leg, face, neck lasting seconds to minutes

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Expanded Classification

- Focal Nonmotor (aware/impaired awareness)
  - Autonomic: A seizure whose primary effect is on autonomic nervous system functions, such as heart rate, blood pressure, sweating, skin color, hair standing on end (piloerection), and gastrointestinal sensations.
  - Behavior arrest: Arrest (pause/freeze) of activities. Use this IF this is the main feature throughout the seizure
  - Cognitive: This type of seizure refers to impaired cognition (thinking) during a seizure. The impairment might affect language, spatial perception, ability to calculate math, or other cognitive functions. Do not count loss of awareness or memory (unless only memory is impaired) as a focal cognitive seizure, because awareness is used to describe other seizure types.
  - Emotional: present with an emotion or perception of having an emotion as an early prominent sign. Fear, anxiety, euphoria/joy, laughing (gelastic) or crying (dycrystic)
  - Sensory: a perceptual experience not caused by appropriate stimuli (tingling, numbness, sounds, smells, vertigo, hot/cold feelings)

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Expanded Classification

- Focal to bilateral tonic-clonic

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Expanded Classification

- Generalized Motor
  - Tonic-clonic: Immediate loss of awareness, with stiffening of all limbs (tonic phase), followed by sustained rhythmic jerking of limbs and face (clonic phase, can be asymmetric).
  - Clonic: Rhythmical sustained jerking of limbs and/or head with no tonic stiffening phase, less common, occur typically in young children
  - Tonic: Bilateral limb stiffening or elevation often with neck stiffening
  - Myoclonic: Irregular, nonsustained jerking of limbs, face, eyes, or eyelids. The jerking of generalized myoclonus may not always be left-right synchronous, but it occurs on both sides.
  - Myoclonic-tonic-clonic: This seizure is like a tonic-clonic seizure, but it is preceded by a few myoclonic jerks on both sides of the body. Such seizures are commonly seen in people with the syndrome of juvenile myoclonic epilepsy
  - Myoclonic-atonic: This seizure presents with a few myoclonic jerks, followed by a limp drop. These seizures previously called myoclonic-astatic
  - Atonic: This is an epileptic drop attack, with sudden loss of muscle tone and strength and a fall to the ground or a slump in a chair. Atonic seizures usually last only seconds.
  - Epileptic spasm: Brief seizures with flexion at the trunk and flexion or extension of the limbs. Video-EEG recording may be required to determine focal versus generalized onset.

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Expanded Classification

- Generalized nonmotor (absence)
  - Typical absence: Sudden onset when activity stops with a brief pause and staring, sometimes with eye fluttering and head nodding or other automatic behaviors. If it lasts for more than several seconds, awareness and memory are impaired. Recovery is immediate. The EEG during these seizures always shows generalized spike-waves
  - Atypical absence: Like typical absence seizures, but may have slower onset and recovery and more pronounced changes in tone. Atypical absence seizures can be difficult to distinguish from focal impaired awareness seizures, but absence seizures usually recover more quickly and the EEG patterns are different.
  - Myoclonic absence: 3 per seconds myoclonic movements leading to pregessive arm elevation, EEG with 3 Hz generalized spike and wave
  - Eyelid myoclonia: Eyelid myoclonia represents jerks of the eyelids and upward deviation of the eyes, often precipitated by closing the eyes or by light

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Expanded Classification

- Unknown
  - Motor
    - tonic clonic
    - spasm
  - Nonmotor
    - behavior arrest

Identify a seizure as focal or generalized onset if there is about an 80% confidence level about the type of onset.

Seizures without enough confidence about onset are labeled of unknown onset.

If a seizure onset becomes clarified at a later date, the type will change.

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Descriptors

- There are many behaviors or sensations during a seizure that are too diverse to classify, thus the ILAE Task Force listed common descriptors that can be added to the classification.

Cognitive

- Aphasia
- Déjà vu/Jamais vu
- Dissociation
- Hallucinations
- Illusions
- Memory impairment

Emotional

- Agitation
- Anger
- Anxiety
- Crying
- Laughing
- Fear
- Paranoia
- Pleasure

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Descriptors

- Autonomic
  - Bradycardia
  - Flushing
  - Nausea/Vomiting
  - Pallor
  - Palpitations
  - Piloerection
  - Tachycardia
  - Respiratory changes
- Automatisms
  - Eye blinking
  - Head nodding
  - Oral facial
  - Manual
  - Eye blinking
  - undressing
  - Walking
  - Vocalizations/speech
- Motor
  - Dystonic
  - Figure of 4
  - Jacksonian
  - Paralysis
  - Paresis
  - Versive
- Sensory
  - Auditory
  - Gustatory
  - Olfactory
  - Visual
  - Somatosensory
  - Hot/cold sensations

Laterality: left, right, bilateral

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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.

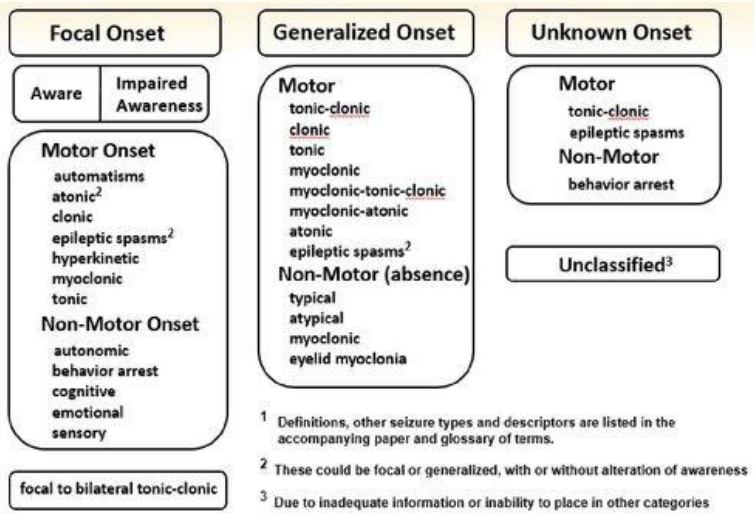
Impaired awareness at any point: A focal seizure is a *focal impaired awareness seizure* if awareness is impaired at any point during the seizure.

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.

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Examples

- 1. Tonic–clonic: 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 magnetic resonance imaging (MRI) findings are normal. This seizure is classified as unknown onset 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 with no further qualifiers.

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Examples

- Focal onset bilateral tonic–clonic: 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 absence 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.

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Examples

- Tonic: A child has brief seizures with stiffening of the right arm and leg, during which responsiveness and awareness are retained. This seizure is a focal aware tonic seizure (the words “motor onset” can be assumed). In the old system, the seizure would have been called tonic, with a perhaps incorrect assumption of generalized onset.

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Examples

- Focal impaired awareness: A 25-year-old woman describes seizures beginning with 30 s 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 seizure would be classified as focal impaired awareness . Even though the patient is able to interact with her environment, she cannot interpret her environment, and is mildly confused. Prior classification would have been complex partial seizure

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Examples

- Sequential seizure manifestations: A seizure begins with tingling in the right arm of a 75-year-old man. Patient says that it then progresses to rhythmic jerking of the right arm lasting about 30 s. He retains awareness and memory for the event. This seizure is a focal (non-motor-onset) sensory seizure. Additional description would be useful, namely focal sensory seizure with somatosensory features progressing to right arm clonic activity.

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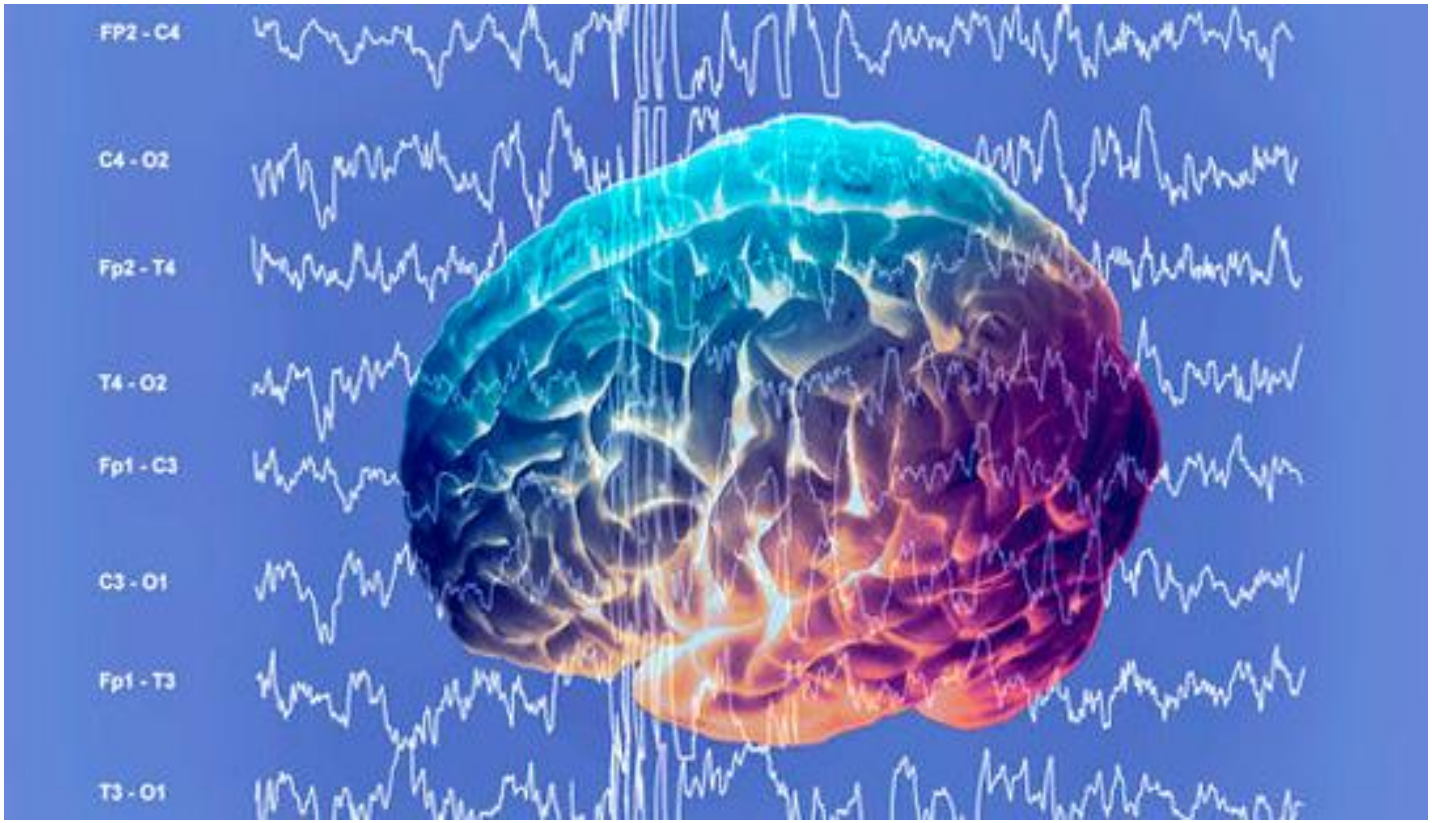
Questions?



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