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new ILAE Classification of seizures and epilepsy

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- seizure and epilepsy classification systems have been used in clinical practice and research since the 1970s. Over the years, multiple revisions have been implemented.

- the most recent of which is the 2017 International League Against Epilepsy (ILAE) operational epilepsy classification system.

- This system aims to better define seizures and epilepsies by classifying them using key clinical features, EEG findings, imaging, and genetics.

- Seizure and epilepsy classification has evolved over the years. Prior to the first modern seizure classification by Gastaut in 1969, seizures and epilepsy types were not distinctly recognized.

- Although initially met with resistance, this system gained international recognition after 1970 and was widely used.

- In 1981 the ILAE, informed by advances in technology—notably video recording with simultaneous EEG—published a classification of seizures followed by a proposal of epilepsy classification in 1985,which was then revised in 1989, wherein the concept of an epilepsy syndrome was introduced.

- The **1981 ILAE** seizure classification system dichotomized seizures into either partial or generalized seizures.
- **Partial seizures** were defined as an epileptic seizure in which "the first clinical and EEG changes indicate initial activation of a system of neurons limited to part of one cerebral hemisphere."
- Partial seizures were **subdivided by level of consciousness**:

✓ simple partial seizures
were associated with no impairment of awareness, and

✓ complex partial seizures
were associated with impairment of awareness.

➢ **third partial seizure type** included seizures that evolved to a secondary generalized convulsion.

- **generalized seizure** was defined as a seizure "in which the first clinical changes indicate initial involvement of both hemispheres."Six generalized seizure types were identified: absence, myoclonic, clonic, tonic, tonic-clonic, and atonic.

- The **1985 epilepsy classification** was also a dichotomized system dividing epilepsies into either idiopathic or symptomatic.
- The term idiopathic derives from the Greek idios, meaning self, own, and personal. Idiopathic epilepsies and syndromes were described as disorders "not preceded or occasioned by another."
- In these disorders, no underlying cause is present other than a **possible hereditary predisposition.**
- Notable **idiopathic epilepsies** included juvenile myoclonic epilepsy and childhood absence epilepsy
- **Symptomatic epilepsies** occurred because of a known disorder or lesion.

- A revision of the epilepsy classification in 1989 added **cryptogenic epilepsies**, which were likely symptomatic, but a cause was not identified.

- The current classification systems build on newly formulated definitions of seizures and epilepsy.
- The **2005 updated seizure definition** is a "transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain."

- Further, in **2014 the ILAE** redefined **epilepsy a disease** and not a disorder to emphasize the importance and impact of epilepsy
- Epilepsy occurs when an individual has an epileptic seizure and his or her "brain…demonstrates a pathologic and enduring tendency to have recurrent seizures.

- Previously, **epilepsy was diagnosed** when an individual had **at least two** unprovoked or reflex seizures more than 24 hours apart.

- Although the **new definition of epilepsy** includes this presentation, epilepsy is also diagnosed if a person has one unprovoked or reflex seizure and has a probability of at least 60% of having another seizure within the next 10 years.

- The **probability of at least 60%** was chosen for this definition because this is the lower limit of the confidence interval for someone with two unprovoked seizures having another seizure within 10 years

- **Epilepsy is considered resolved when …**

❖ a patient with an age-dependent epilepsy syndrome is older than the age in which this syndrome was active

or

❖ when a patient has been seizure free for 10 or more years and has been off all antiepileptic drugs for 5 or more years.

- The prior and current classification systems aim to :

- group seizures according to clinical presentation and brain region onset and to
- group epilepsies according to seizure type, age of onset, probability of remission, EEG findings, radiologic findings, and genetics.

- Classification of seizures, epilepsies, and epilepsy syndromes creates a framework for patients, their families, clinicians, and researchers.

- For patients, the universal common language of the classification system provides a name and diagnosis, which improves understanding and recognition of the disease.

- Moreover, this common language enhances communication between patients, their families, and providers.

- Similarly, clinicians can use the language to better communicate with patients and colleagues.

- The classification systems allow clinicians to consider history and include data from new technologies when making a diagnosis, choosing treatment, and assessing prognosis.

UPDATED SEIZURE CLASSIFICATION

- The new classification addresses the limitations of the 1981 seizure classification, which include the following:

(1) some seizure types can have either focal or generalized onset

(2) lack of knowledge about seizure onset makes a seizure unclassifiable and difficult to place within the 1981 system

(3) retrospective seizure descriptions often do not include the level of consciousness

(4) terms used in the 1981 seizure classification such as complex partial or simple partial are difficult to understand and

(5) some seizure types are not included in the 198 1classification.

- To address needs of different clinicians and researchers, both basic
- (FIGURE 1-1) and expanded (FIGURE 1-2) versions of seizure classification were created.

- The basic version of the seizure classification is a contracted form of the expanded classification and is intended to be more useful for pediatricians,non-neurologists, general neurologists, physicians in general practice, nurses,and health care workers.

- The expanded version is more detailed and will likely be used more by epileptologists/neurophysiologists and researchers.

Basic version of seizure classification

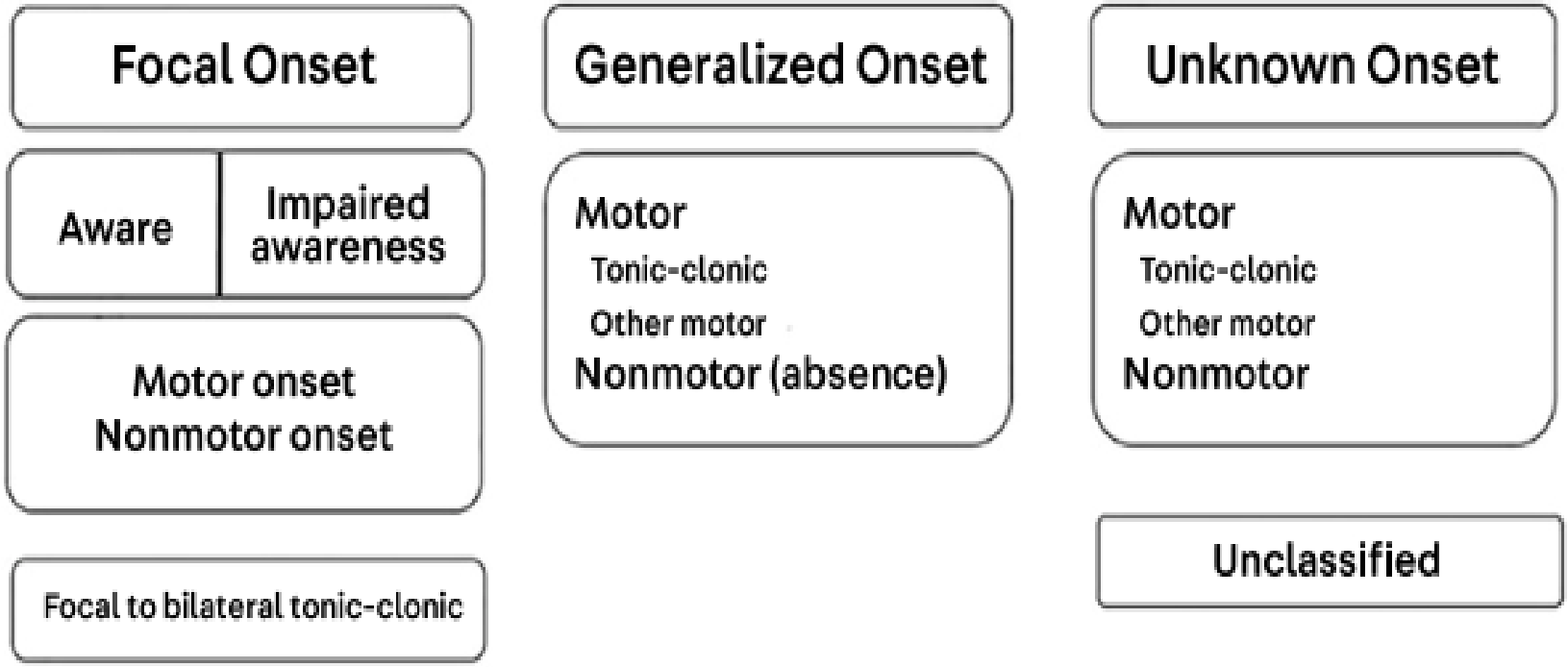

FIGURE 1-1

Basic version of 2017 International League Against Epilepsy seizure type classification.

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Expanded version of seizure classification

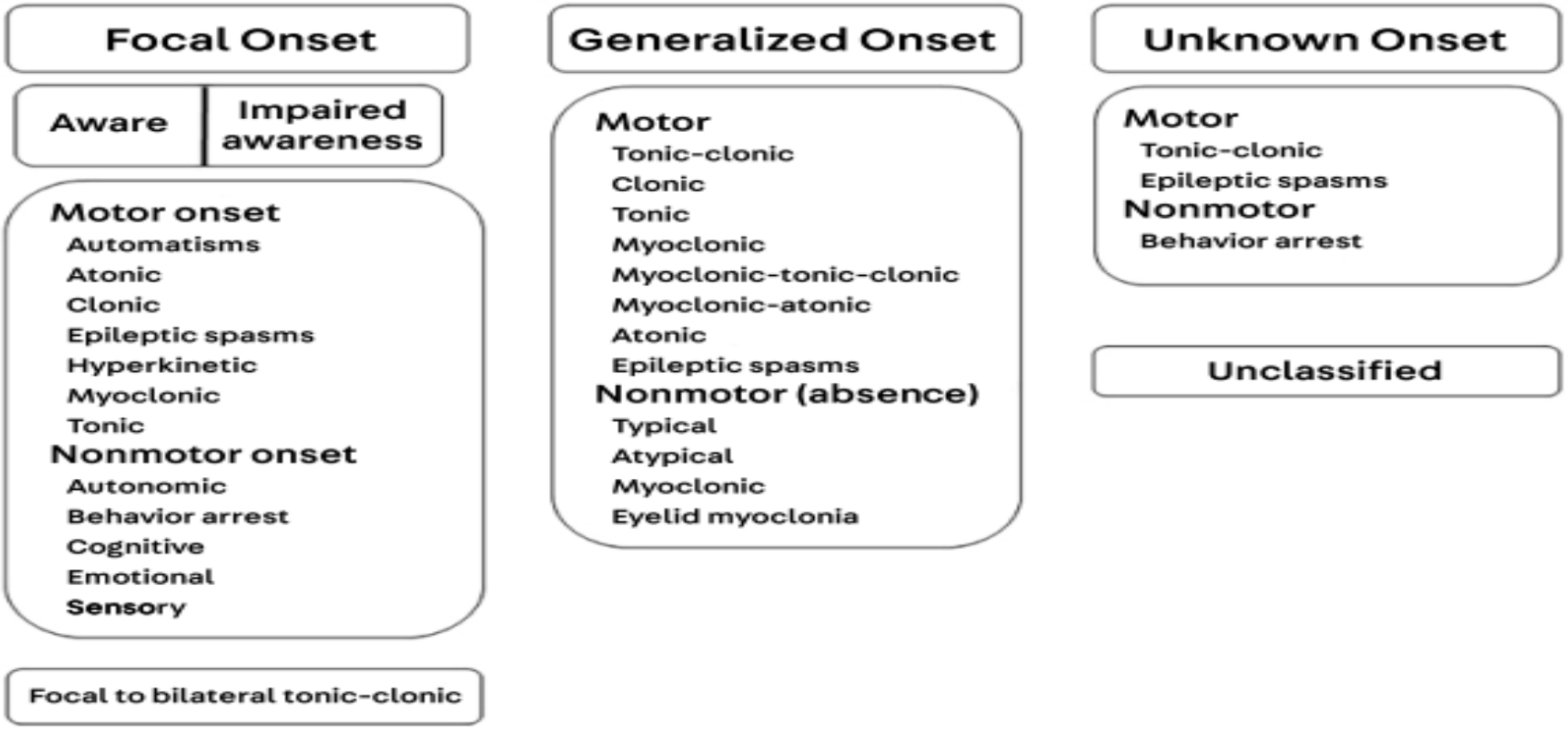

FIGURE 1-2
Expanded version of 2017 International League Against Epilepsy seizure type classification.
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Seizure classification starts with

- whether the initial manifestations of the seizure **are focal or generalized**.

- **Focal seizures** originate within a neuronal network limited to one hemisphere that may be discretely localized or more widely distributed, whereas
- **generalized seizures** originate at some point within the brain and rapidly engage bilateral distributed networks.

- If the onset of the seizure is missed or is unclear, the seizure is of **unknown onset.**

➢ Focal Seizures

- Focal seizures are classified according to;

✓ the patient's level of awareness

✓ first most prominent motor or nonmotor features of the seizure.

- These early prominent features are important to consider when localizing the seizure onset or epileptogenic zone.
- The final feature used in classification of focal seizures is:

✓ whether the focal seizure evolves to a bilateral tonic-clonic seizure

- secondary generalized tonic-clonic seizure is no longer used ✘ because the term focal seizure more completely differentiates this type from generalized seizures.

- **Awareness** is defined as knowledge and understanding that something is happening or exists.

- When a person is having a focal seizure, his or her awareness is determined by whether the person knows who they are and what is going on in his or her surroundings during the seizure;

it does not refer to awareness of the seizure occurring.

- Awareness is also distinct from responsiveness.
- If awareness is impaired for any portion of the seizure, then the seizure is classified as a focal seizure with impaired awareness.

- Awareness may be considered a surrogate for consciousness.

- Impaired awareness or consciousness during a seizure is likely secondary to depressed subcortical arousal systems, leading to deep sleep activity in widespread neocortical regions, hence the involvement of both subcortical and cortical structures.
- A focal aware seizure replaces the previously termed simple partial seizure
- a focal impaired awareness seizure replaces the term complex partial seizure.
- If unknown, the level of awareness does not need to be included.

- Patients with ***Focal aware seizures*** can interact normally with the environment during seizure except for limitations imposed by the seizure on specific localized brain functions, such as aphasia during a dominant temporal lobe seizure, which could be confused for lack of awareness due to the patient's inability to speak.

- ***Focal impaired awareness seizures***, on the other hand, are defined by impaired consciousness and imply further spread of the seizure discharge at least to basal forebrain and limbic areas.
  Postictally,patients are confused and disoriented for several minutes, and determining the transition from ictal to postictal state may be difficult without simultaneous EEG recording.

- Of focal impaired awareness seizures, 70% to 80% arise from the temporal lobe; foci in the frontal and occipital lobes account for most of the remainder.

- **Focal motor seizures** can be more specifically defined.
- Motor-onset manifestations include :

✓ Automatisms
✓ epileptic spasms
✓ atonic
✓ clonic
✓ hyperkinetic
✓ myoclonic
✓ tonic seizures

- Automatisms are coordinated, purposeless, repetitive motor activities that may appear normal in other circumstances. Examples include:
- oral automatisms such as lip smacking
- manual automatisms including repetitive hand movements such as patting

- **Focal atonic seizures** are characterized by loss of tone in one body part.
- **Clonic seizures** are repeated, regularly spaced stereotypical jerking movements.
- **Epileptic spasms** were previously only considered generalized seizures.

✓ Clinically, epileptic spasms present in young children with flexion of the waist and flexion or extension of the arms, usually in clusters.

✓ If epileptic spasms occur in infants or early in life, they can be referred to as infantile spasms.

✓ Differentiating focal epileptic spasms from generalized epileptic spasms may require careful observation of clinical and electrographic features.

- **Hyperkinetic** or excessive muscular movement seizures can have variable features clinically , including thrashing or pedaling.

- **Focal myoclonic seizures** present with jerking but, in contrast to clonic seizures, the jerking is irregular and not rhythmic.

- **Tonic seizures** refer to motor seizures with increased tone or stiffening of the limb or neck.

- **Focal seizures with nonmotor symptoms** as the first prominent feature include

✓ autonomic

✓ behavior arrest

✓ Cognitive

✓ emotional

✓ sensory seizures

❑ **Autonomic seizures** present with changes in heart rate, blood pressure, sweating , skin color, piloerection, or gastrointestinal sensations.

❑ **Behavioral arrest seizures**

are characterized by cessation of movement, which should be the dominant feature throughout the entire seizure and not just a brief part of the seizure;

- clinical symptoms include a blank stare and cessation from talking or moving.

- **Patients with nonmotor cognitive seizures** can experience changes in language function, thinking, or associated higher cortical functions; more specific examples include:
  - déjà vu (a familiar feeling) , jamais vu (a feeling of unfamiliarity), or olfactory hallucinations.

- **Emotional seizures** appear with clear emotional changes such as dread, fear, anxiety, or pleasure.

- **Focal sensory seizures** are classified according to changes in sensory phenomena such as taste, smell, hearing, vision, pain, numbness, or tingling.

- Focal seizures can be further classified as to whether they evolve to a **bilateral tonic-clonic seizure**.

- As discussed previously, this classification replaces secondary generalized tonic-clonic to avoid any confusion between generalized and focal seizures.

- These seizures start in one area of the brain (as with all focal seizures) and then spread to both sides of the brain. This spread is typically clearly seen on EEG.

Generalized Seizures

- Similar to focal seizures, generalized seizures are classified according to motor or nonmotor manifestations.
- Broadly, **motor seizures** are either tonic-clonic or other motor seizures.
- **Nonmotor generalized seizures** primarily refer to absence seizures.

- **Motor onset** more specifically includes tonic-clonic, clonic, tonic, myoclonic, myoclonic-tonic-clonic, myoclonic-atonic, atonic, or epileptic spasms.
- **Generalized tonic-clonic seizures** generally last 1 to 3 minutes and result in immediate loss of awareness or consciousness. The initial tonic phase is a stiffening of all limbs. The patient may groan or cry in the beginning as air is forced past the vocal cords.The tongue may also be bitten during this phase. The clonic phase occurs after the tonic phase and is characterized by sustained rhythmic jerking of the limbs. If the person has impaired breathing, he or she may look dusky. Incontinence of either bladder or bowel occurs when the body relaxes.

- **A generalized clonic seizure** is characterized by bilateral and sustained rhythmic jerking.

- A patient with **generalized tonic seizures** will have stiffening of all limbs.

❖ In contrast to generalized clonic seizures,

- **generalized myoclonic seizures** are associated with irregular and not necessarily synchronous bilateral jerking of limbs, face, eyes, or eyelids.

- **Myoclonic-tonic-clonic seizures** are a new seizure designation and begin with irregular jerking on both sides followed by a tonic-clonic seizure. Myoclonic-tonic-clonic seizures are common in juvenile myoclonic epilepsy

- **Myoclonic-atonic seizures** are also a new seizure designation and are characterized by an initial irregular jerking followed by loss of tone on both sides. These seizures are common in epilepsy with myoclonic-atonic seizures **(Doose syndrome).**

- **Atonic seizures** ***also called drop attacks*** The term *astatic seizure* is now no longer in use
  are brief and occur when there is bilateral loss of tone and the muscles become limp. may be fragmentary (eg,head drop) or generalized
  If the person is standing when the seizure occurs, he or she will fall, often resulting in injury.
- **Epileptic spasms** are also brief and typically occur in clusters with flexion at the trunk and flexion or extension of the limbs. As with focal epileptic spasms, an EEG may be needed to distinguish whether the seizure is generalized.

- **Nonmotor or absence seizures** include typical, atypical, myoclonic, or eyelid myoclonia.

- **Typical absence seizures** present with a sudden cessation of activity sometimes with eye fluttering, head nodding, or other automatisms followed by an immediate recovery. EEG always reveals generalized spike-wave activity during the seizure.
- **Atypical absence seizures** are similar to absence seizures but have other features including slower onset, prolonged recovery, and more pronounced changes in tone. *EEG correlate of slow spike and wave (less than 3 Hz frequency). Atypical absences are seen most often in children with epilepsy who are developmentally delayed or in epileptic encephalopathies, such as the Lennox-Gastaut syndrome*

- **A myoclonic absence seizure** begins with a few irregular jerks followed by an absence seizure.
- **Eyelid myoclonia** is defined by jerks of the eyelids and upward deviation of the eyes. **Light** and **closing the eyes** can precipitate these generalized seizures.
- Eyelid myoclonia with absence seizures is seen in **Jeavons syndrome**.

Unknown Seizures

- Seizures of unknown onset can be classified by

✓ motor (tonic-clonic, epileptic spasms) or

✓ nonmotor (behavior arrest) presentations.

- If information is inadequate or if the seizure cannot be categorized, then the seizure is considered **unclassified.**

UPDATED EPILEPSY CLASSIFICATION

- The second level of classification is the **epilepsy type**.
- This classification assumes the patient has epilepsy as defined by the previously discussed updated definition.
- The epilepsy type is predominantly determined clinically;
- characteristic EEG findings provide supportive evidence.
- Similar to seizure classification, the epilepsies are classified as generalized or focal.
- The new classification system additionally recognizes two new categories: combined generalized and focal epilepsy and unknown epilepsy

- ❑ Patients with **generalized epilepsy** have one or more of the generalized seizure types, and their EEGs typically display generalized spike-wave activity.
- For individuals who have generalized seizure types and a normal EEG, other data are needed to determine whether the epilepsy is generalized.

Having myoclonic jerks or a pertinent family history supports the diagnosi of a generalized epilepsy type.

- ❑ Clinically, patients with one or more focal seizure types have **focal epilepsy**. These epilepsies can be either unifocal or multifocal.
- Although not always seen, focal EEG findings such as focal slowing or epileptiform discharges support the diagnosis of focal epilepsy.
- Concordant focal MRI findings are also supportive.

- ❑ Designation of **combined generalized and focal epilepsy** is for patients with both focal and generalized seizures. EEG may reveal both focal and generalized electrographic findings. Examples of combined generalized and focal epilepsy include **Dravet syndrome** and **Lennox-Gastaut syndrome**.

- When the patient has epilepsy as defined by the ILAE but it remains undetermined whether the patient has focal or generalized epilepsy, the classification of **unknown epilepsy type** is used.
- Patients with this classification may not have an available EEG or the EEG may be indeterminate.
- Other supporting studies such as MRI and family history are also either not available or do not clarify the epilepsy classification.

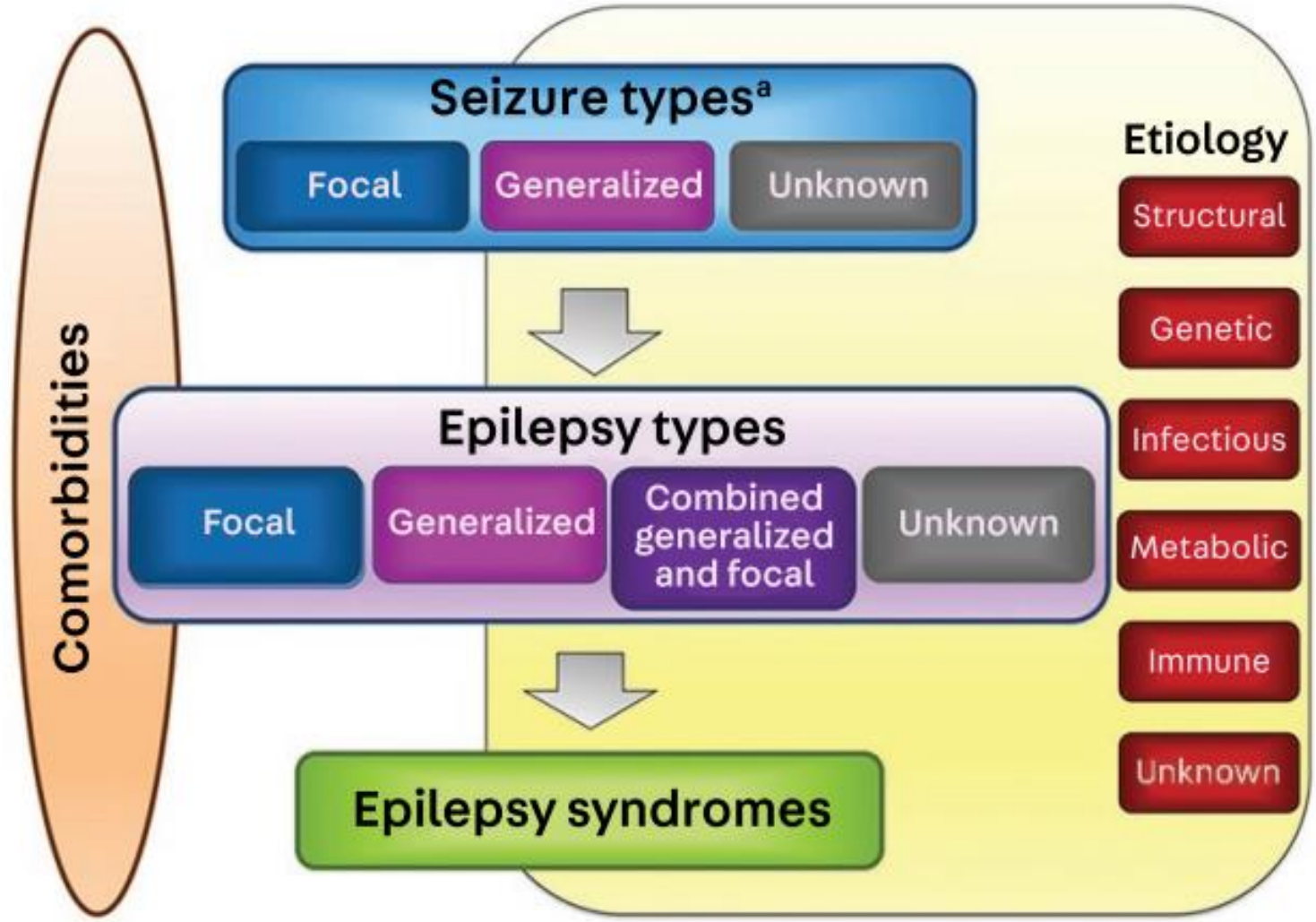


FIGURE 2-1
Classifications of the epilepsies.

Focal Epilepsy

Temporal Lobe Epilepsy

- most common epilepsy seen in adults
- epileptogenic region involves mesial temporal lobe structures, especially the hippocampus, amygdala, and parahippocampal gyrus.
- Seizures usually begin in late childhood or adolescence,
- and a ***history of febrile seizures*** is common.
- Virtually all patients have focal impaired awareness seizures, some of which evolve to bilateral tonic-clonic activity.
- ***Auras are frequent***; visceral sensations are particularly common.
- Other typical behavioral features include a motionless stare, loss of awareness that may be gradual, and oral-alimentary automatisms, such as lip smacking. Ipsilateral manual automatisms are also common

➢ focal temporal slowing and epileptiform sharp waves
When seizures persist, workup for surgical treatment should be undertaken as
up to 80% of selected patients will achieve ***complete seizure freedom***

Focal Epilepsy
Frontal Lobe Epilepsy

- The following features, when taken together, suggest frontal lobe epilepsy:

1. **Brief** seizures that begin and end abruptly with little, if any, postictal period

2. A tendency for seizures to cluster and to occur at night

3. Prominent, but often bizarre, motor manifestations, such as asynchronous thrashing or flailing of arms and legs; pedaling leg movements; pelvic thrusting; and loud, sometimes obscene, vocalizations, all of which may suggest psychogenic seizures

4. Minimal abnormality on scalp EEG recordings

5. A history of status epilepticus

NEW CLASSIFICATION OF EPILEPSY SYNDROME

- The epilepsy syndrome is a new addition to the current classification system and is defined as "a cluster of features incorporating seizure types, EEG, and imaging features that tend to occur together.

➢ Factors that contribute to epilepsy syndrome include age of onset, remission, triggers, diurnal variation, intellectual and psychiatric dysfunction, EEG findings, imaging studies, family history ,and genetics.

- The ILAE has never formally classified a list of epilepsy syndromes; however, well-known and accepted syndromes are described.

- Previously, the **term benign** was used to describe some of the epilepsy syndromes, but it is no longer used as it infers the epilepsy has minimal effect on the patient.

- It is now more clearly understood that any epilepsy can have social effects and can be associated with other comorbidities such as learning disorders or psychiatric conditions.

The term **self-limiting** is now used.

EPILEPSY SYNDROME

- **Idiopathic or Genetic Generalized Epilepsy Syndromes**
- **Reflex Epilepsy Syndromes**
- **Focal Epilepsy Syndromes**
- **Infantile Spasms (West Syndrome)**
- **Lennox-Gastaut Syndrome (A Type of Combined Focal and Generalized Epilepsy Syndrome)**

Idiopathic or Genetic Generalized Epilepsy Syndromes

- ✓ Idiopathic generalized epilepsies include :
- ✓ childhood absence epilepsy
- ✓ Juvenile absence epilepsy
- ✓ juvenile myoclonic epilepsy
- ✓ and generalized tonic-clonic seizures alone (TABLE 1-1).

- Controversy surrounds the use of the term idiopathic, and removing it from epilepsy classification has been advocated.
- Idiopathic was meant to refer to self or genetic. There is, however, concern that use of the word genetic infers inherited, and many patients with epilepsy have de novo mutations or have complex genetic syndromes that occur with or without environmental factors. Many in the epilepsy community want to continue using the term idiopathic generalized epilepsy, and the ILAE task force decided to use it to refer to the previously mentioned epilepsies. When the clinician determines that a clear genetic etiology is present, the term genetic generalized epilepsy may be used to refer to the epilepsy syndrome.

EEGs of the idiopathic generalized epilepsie reveal a

- normal electrographic background and
- characteristic generalized spike-wave findings

TABLE 1-1

Idiopathic or Genetic Epilepsy Syndromes

Epilepsy Syndrome	Seizure Types	Age of Onset	Self-limiting (Yes or No)	EEG Findings
Childhood absence epilepsy	Absence, generalized tonic-clonic (rare)	4 to 10 years	Yes	Normal background, occipital intermittent rhythmic delta activity, 3-3.5 Hz generalized spike-wave discharges
Juvenile absence epilepsy	Absence, generalized tonic-clonic, myoclonic (rare)	Adolescence to early adulthood	No	Normal background, polyspikes may be present, 3-3.5 Hz generalized spike-wave discharges
Juvenile myoclonic epilepsy	Myoclonic, generalized tonic-clonic, absence (rare)	10 years to mid-twenties	No	Normal background, 3-3.5 Hz generalized spike-wave discharges, >4 Hz generalized spike-wave discharges, high-amplitude polyspike-wave discharges with myoclonic seizures, photoparoxysmal response in up to 40% of patients
Epilepsy with generalized tonic-clonic seizures alone	Generalized tonic-clonic	Childhood to mid-adulthood	No	Normal background, generalized spike/polyspike-wave discharges

Childhood absence epilepsy

- affects neurologically **normal** girls more than boys
- typically self-limiting.
- **Onset** is usually between 4 and 10 years of age
- **remission** usually occurring in adolescence
- Patients present with absence seizures and occasionally with generalized tonic-clonic seizures **(30% to 50%)**

- Early occurrence of generalized tonic-clonic seizures is associated with a **poorer prognosis**.

Juvenile absence epilepsy

- **onset** in adolescence and early adulthood
- **peak onset** between 10 and 13 years of age
- Girls and boys are affected equally
- Absence seizures occur less frequently than in childhood absence epilepsy.
- Generalized tonic-clonic seizures occur early in the presentation
- myoclonic seizures, although rare, may also occur.
- In contrast to childhood absence epilepsy, this syndrome is **not self-limiting**

Juvenile myoclonic epilepsy

- one of the most typical epilepsy syndromes.
- **Onset** ranges from before age 10 through the mid-twenties and later in some cases.
- more commonly in **women**
- All patients have myoclonic seizures and commonly have generalized tonic-clonic seizures.
- Absence seizures rarely occur.
- Most patients do not have spontaneous remission and require lifelong treatment with antiepileptic medication.

Epilepsy with generalized tonic-clonic seizures alone

- characterized by presentation of generalized tonic-clonic seizures
- with an age range of childhood through mid-adulthood
- **peak onset in the second decade of life**
- **Previously** it was referred to as generalized tonic-clonic seizures **upon awakening** but was changed …

➢ after recognition that **seizures can occur at any time of day**.

- Similar to juvenile absence epilepsy and juvenile myoclonic epilepsy…

epilepsy with generalized tonic-clonic seizures alone is **not self-limiting**, and lifelong antiepileptic drug treatment is typically required.

Reflex Epilepsy Syndromes

- Reflex epilepsy syndromes are epilepsies in which seizures are provoked by a specific stimulus
- Seizures are typically generalized tonic-clonic seizures, but…
  - other generalized seizure types may also occur.
- **Rarely, focal** seizures may present as a reflex epilepsy.

- The **most common** reflex epilepsy syndrome is **photosensitive epilepsy.**

- Other reflex epilepsy syndromes include reading epilepsy and startle epilepsy

Focal Epilepsy Syndromes

- Well-described focal epilepsy syndromes include:

✓ **childhood epilepsy with centrotemporal spikes**

✓ **Panayiotopoulos syndrome**.

❑ **Childhood epilepsy with centrotemporal spikes**

✓ Previously was referred to as benign epilepsy with centrotemporal spikes. benign rolandic epilepsy

✓ is a self-limited epilepsy, 15% of all pediatric seizure disorders

✓ presents in the school years with brief focal motor hemifacial seizures and nocturnal focal motor seizures evolving to bilateral tonic-clonic seizures.

✓ **EEG background is normal** with **sleep-activated centrotemporal spikes**.

❑ Panayiotopoulos syndrome

- is also a self-limited epilepsy
- characterized by having focal autonomic seizures
- **often prolonged**
- Focal occipital high-amplitude sleep-activated spikes seen on EEG.
- Possible autonomic symptoms include;

✓ vomiting, pallor, mydriasis, cardiorespiratory, gastrointestinal, and thermoregulatory symptoms, incontinence, and hypersalivation.

- These seizures can be **mistaken for acute gastroenteritis**.
- **onset** of seizures is between **3 and 6 years of age**
- Seizures usually **resolve by ages 11 to 13 years**.
- **Birth and developmental history are typically normal**, as is the neurologic examination.

❑ *Gastaut-type idiopathic occipital epilepsy of childhood*

- Self limited benign focal epilepsy syndrome, is characterized by

✓ Elementary **visual hallucinations**, typically multicolored and circular, appearing in the periphery of a hemifield, which may be flashing, may move across the visual field, or remain static.

- They are **usually stereotyped**

- The EEG is characterized by **occipital paroxysms** that are typically **bilateral and synchronous** and
- **induced by eye closure** (eliminating visual fixation)

❑ *Lennox-Gastaut Syndrome (A Type of Combined Focal and Generalized Epilepsy Syndrome)*

heterogeneous group of childhood epileptic encephalopathies that are characterized by mental retardation, uncontrolled seizures, and a distinctive EEG pattern.

❑ *Infantile Spasms (West Syndrome)*

unique age-specific form of **generalized or focal epilepsy** that may be either **idiopathic or symptomatic**.

When all clinical data are considered, including results of imaging studies, **only about 15% of patients are now classified as idiopathic**.

Symptomatic cases result from diverse conditions, including cerebral dysgenesis, tuberous sclerosis, phenylketonuria, intrauterine infections, or hypoxic-ischemic injury.

❑ Autosomal dominant epilepsy with auditory features

- **auditory symptoms** with or without **receptive aphasia** as prominent ictal features.
- Most patients have **focal to bilateral tonic-clonic seizures**
- **Mutations in the *LGI1*** have been identified in **about 50% families**

❑ autosomal dominant nocturnal frontal lobe epilepsy

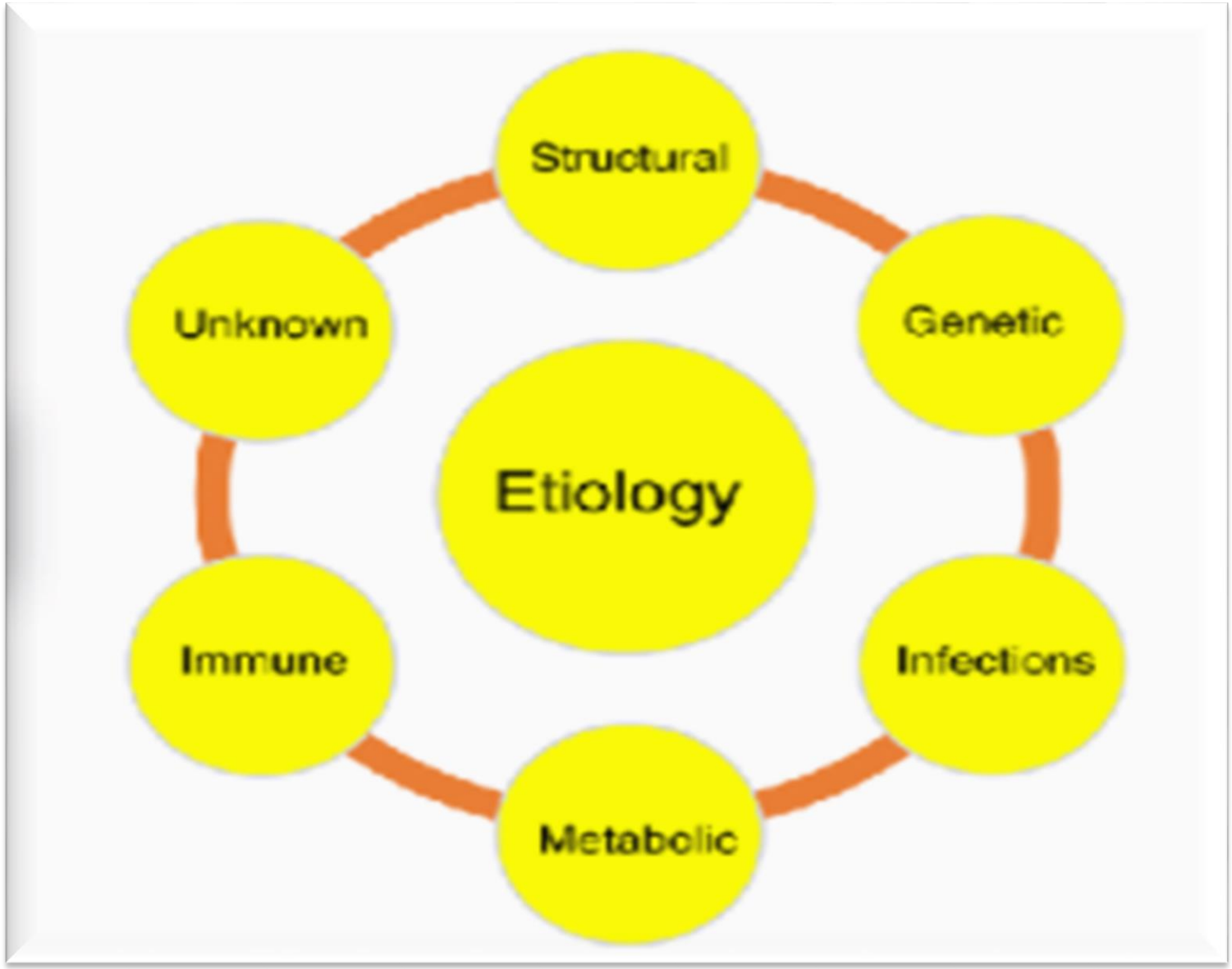
seizures usually occur **in clusters during sleep**

often **stereotyped** and **brief** and vary from **simple arousals to dramatic bizarre hyperkinetic movements.**

✓ Most patients respond well to medication, and
seizures do become **milder and less frequent with advancing age**.

EPILEPSY ETIOLOGY

- The etiology of seizures and epilepsies is emphasized in the new classification system.
- In the prior classification system of the 1980s, etiology was inferred when classifying the epilepsy.
- Idiopathic primarily referred to genetic causes
- symptomatic referred to the presence of a known disorder or lesion,
- cryptogenic referred to a presumed but unknown symptomatic cause.
- As discussed, the term idiopathic is now used to refer to four well-described epilepsy syndromes.
- The terms symptomatic and cryptogenic are no longer used. ✘


Structural
Genetic
Infections
Metabolic
Immune
Unknown
Etiology

structural etiology

- A structural etiology is determined when a structural abnormality is seen on neuroimaging and when the signs and symptoms of seizures, in combination with EEG data, suggest this abnormality is the probable cause of the seizures.

- If the clinical and EEG data are discordant with localization of the visible structural abnormality, then the imaging abnormality is not relevant to the patient's epilepsy.

- Structural abnormalities may be genetic, acquired, or both.

- Possible structural abnormalities include stroke, trauma, tumor, malformations of cortical development, and infection.

Genetic etiologies

- Genetic etiologies are determined if there is a known or presumed genetic mutation in which seizures are a core symptom of the disorder.
- Genetic epilepsies are diverse, and the list grows each year.
- **Importantly, genetic does not always mean inherited**.
- Although some epilepsies are inherited, many occur secondary to a de novo (new) mutation in the affected individual.

- In some cases, the genetic mutation is not identified, but the clinical presentation, EEG findings, and family history suggest a genetic etiology.

- In addition, the genetic etiology for some epilepsy syndromes such as juvenile myoclonic epilepsy is inferred from research studies including twin and familial aggregation studies.

- Overall, genetic etiology is defined by having a known mutation, clinical presentation with supportive data and family history, or a syndrome with evidence from research studies to suggest a genetic etiology

Infectious etiologies

- Infectious etiologies are the most common worldwide etiology.
- An important distinguishing point is that the patient has epilepsy secondary to an infectious etiology and not seizures in the setting of an acute infectious illness.

- Prototype infectious etiologies include neurocysticercosis, HIV, cytomegalovirus, and cerebral toxoplasmosis.

- Epilepsy onset secondary to a prior infectious insult such as meningitis or encephalitis is also considered an infectious etiology.

metabolic etiology

- Epilepsies with a metabolic etiology occur secondary to a known or presumed metabolic disorder in which seizures are a core symptom of the disorder.

- Overlap with a genetic etiology may occur as many metabolic disorders have known genetic mutations.

- Of course, identifying a genetic etiology early in presentation is important because management interventions such as a change in diet or supplementation can affect its natural course.

Immune etiologies

- Immune etiologies are increasingly recognized as potential causes of epilepsy.
- As with the other etiologies, seizures are a core symptom of the immune disorder.

- In patients with identified immune etiologies, **immunotherapy** should be considered.

❑ Examples of immune etiologies for seizures include;

✓ **anti–N-methyl-D-aspartate (anti-NMDA) receptor encephalitis**

✓ anti–leucine-rich, glioma inactivated 1 **(anti-LGI1) encephalitis.**

Unknown etiologies

- The last etiologic category is unknown;
- **up to one-third of patients** with epilepsy have no clear etiology.

- A cause likely exists, but its identification may be limited by inadequate resources such as poor access to up-to-date brain
- imaging, immune antibody testing, or genetic testing.

- It is hoped that with improved access to care as well as continued research into understanding the epilepsies the percentage of patients who have an unknown etiology will diminish and that the structure of the new classification system will provide a framework to better understand the epilepsies.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