EPILEPSY AND SEIZURES

ILAE Commission for Classification and Terminology

What is seizure?

A seizure represents the clinical expression of abnormal, excessive or synchronous discharges of neurons residing primarily in the cerebral cortex. This abnormal paroxysmal activity is intermittent and usually self-limited, lasting seconds to a few minutes.

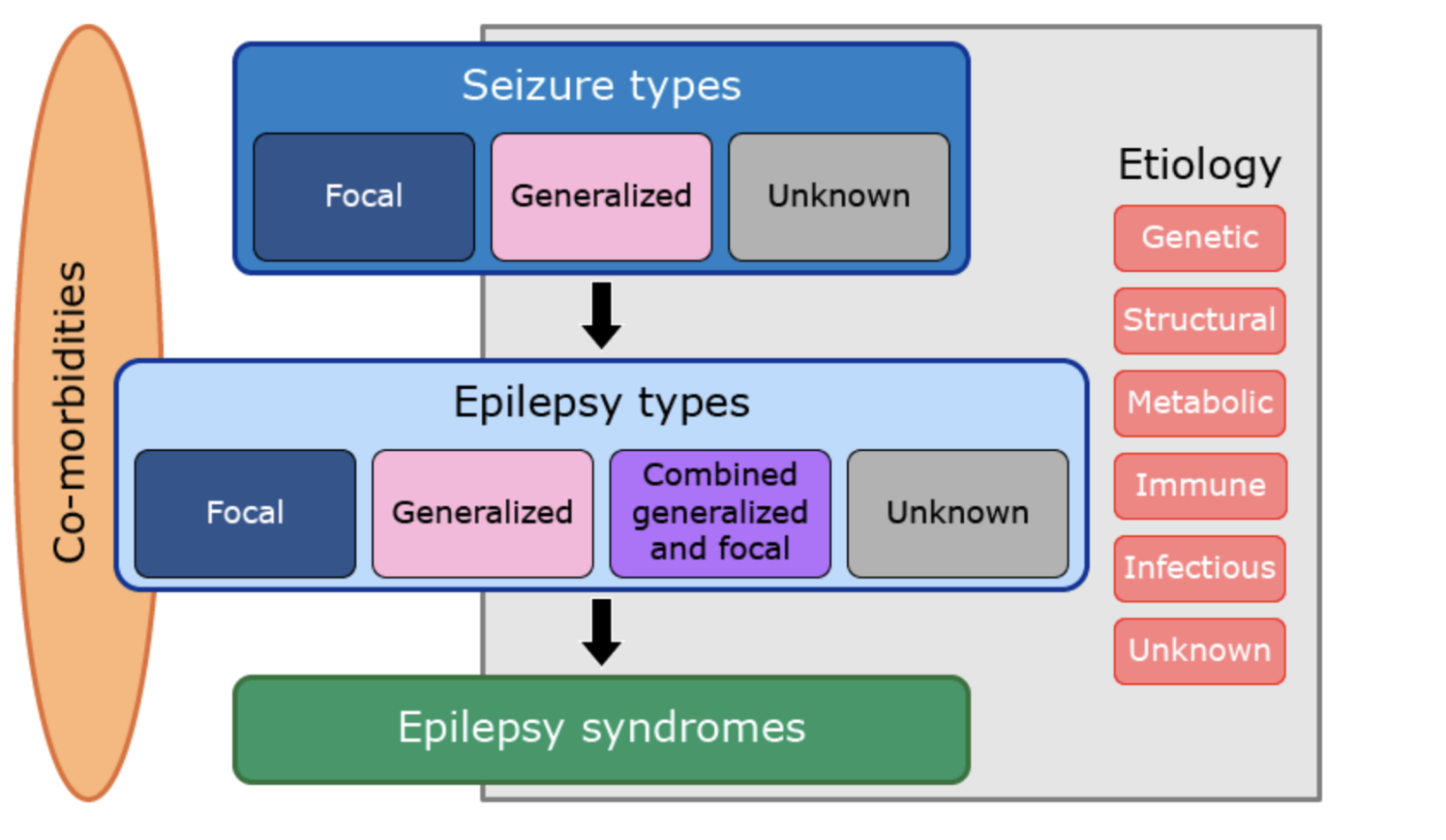
Provoked (symptomatic) seizure

- A seizure that occurs in close temporal association with an acute systemic illness or brain insult. Examples include hyponatremia, hypocalcemia, high fever, toxic exposure, intracranial bleeding, or bacterial meningitis.
- This is not epilepsy.

What is Epilepsy

- Epilepsy is a state of an enduring predisposition to recurrent epileptic seizures . An individual is considered to have epilepsy when any of the following exists :
- ●At least two unprovoked seizures occurring more than 24 hours apart.
- ●One unprovoked seizure and a probability of further seizures that is similar to the general recurrence risk after two unprovoked seizures (eg, ≥60 percent), occurring over the next 10 years. This may be the case with remote structural lesions such as stroke, central nervous system infection, or certain types of traumatic brain injury.

ILAE framework for classification of epilepsy



Purpose of the International Classification of Seizures and Epilepsies

- To provide a common international terminology and classification
- Largely for clinical (treatment) purposes
- Purpose of classification: to organize items according to their fundamental relationships

Seizure types

Self limited seizure types

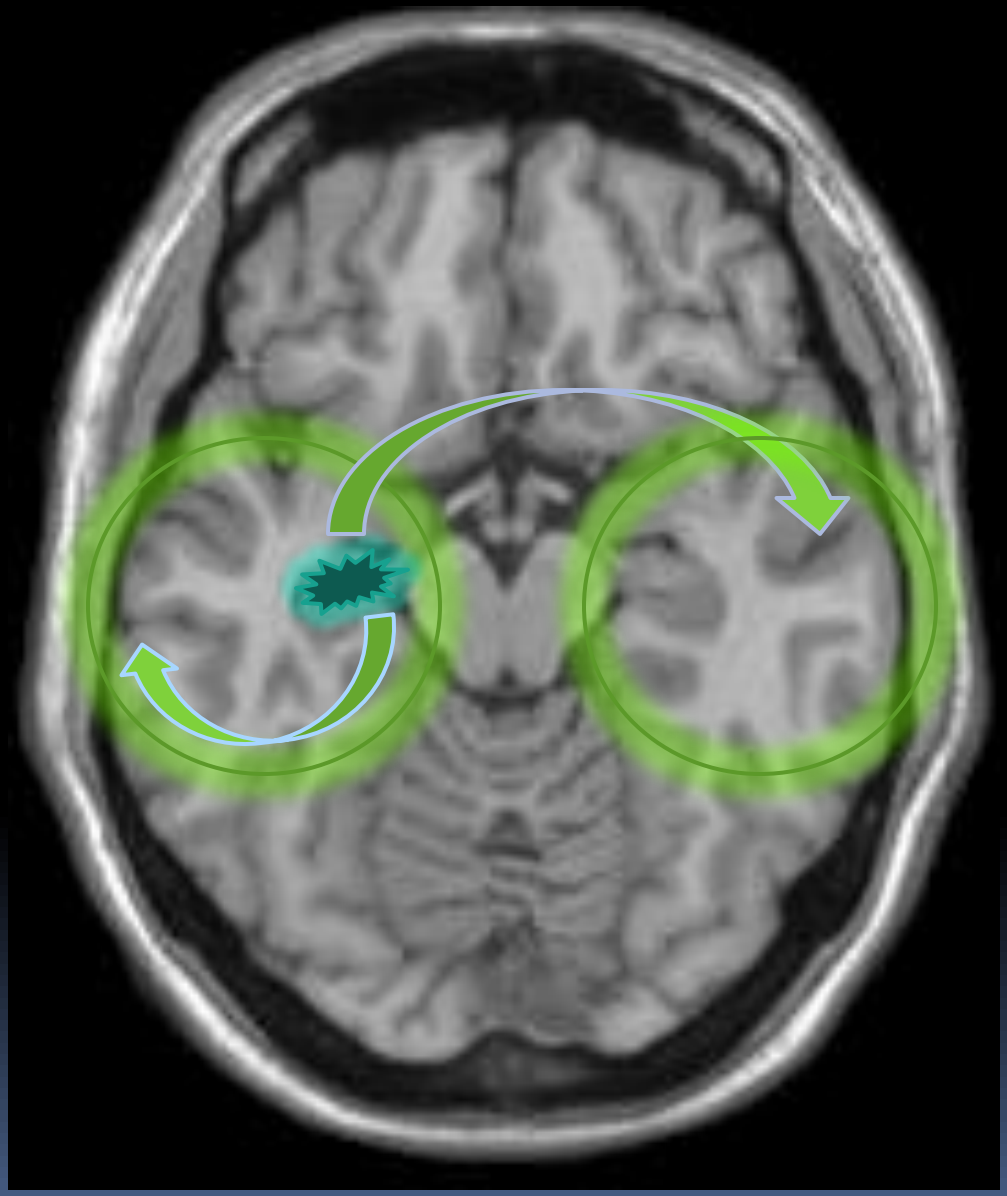
- Generalized seizures
- Focal seizures

Continuous seizure types

- Generalized status epilepticus
- Focal status epilepticus

Focal reconceptualized

- For seizures:
  - *Focal epileptic seizures are conceptualized as originating within networks limited to one hemisphere. These may be discretely localized or more widely distributed….*



FOCAL SEIZURES

- Focal sensory seizures
- Focal motor seizures
- Autonomic seizures

FOCAL SEIZURES
Focal sensory seizures

- manifestations may include paresthesia, feelings of distortion of an extremity, vertigo, gustatory sensations, olfactory symptoms, auditory symptoms, and visual phenomena such as flashing lights.
- Focal sensory seizures with elementary sensory symptoms (e.g. occipital and parietal seizures)
- Focal sensory seizures with experiential sensory symptoms (e.g. temporo parieto occipital junction seizures).

FOCAL SEIZURES
Focal motor seizures

These may manifest as focal motor activity, sometimes with an anatomic spread or march of activity (Jacksonian), versive movement (turning of the eyes, head and/or trunk), vocalization, or arrest of speech.

Focal seizures autonomic

- The features of autonomic seizures may include an epigastric "rising" sensation (a common aura with medial temporal lobe epilepsy), sweating, piloerection, and pupillary changes.

Focal seizures

Blume et al, Epilepsia 2001

- Without impairment of consciousness or awareness
  - Previous term: simple partial
  - With observable motor or autonomic components
    - eg. focal clonic, autonomic, hemiconvulsive
  - With subjective sensory or psychic phenomena
    - Aura - specific types
- Where alteration of cognition is major feature
  - Previous term: complex partial
  - Dyscognitive

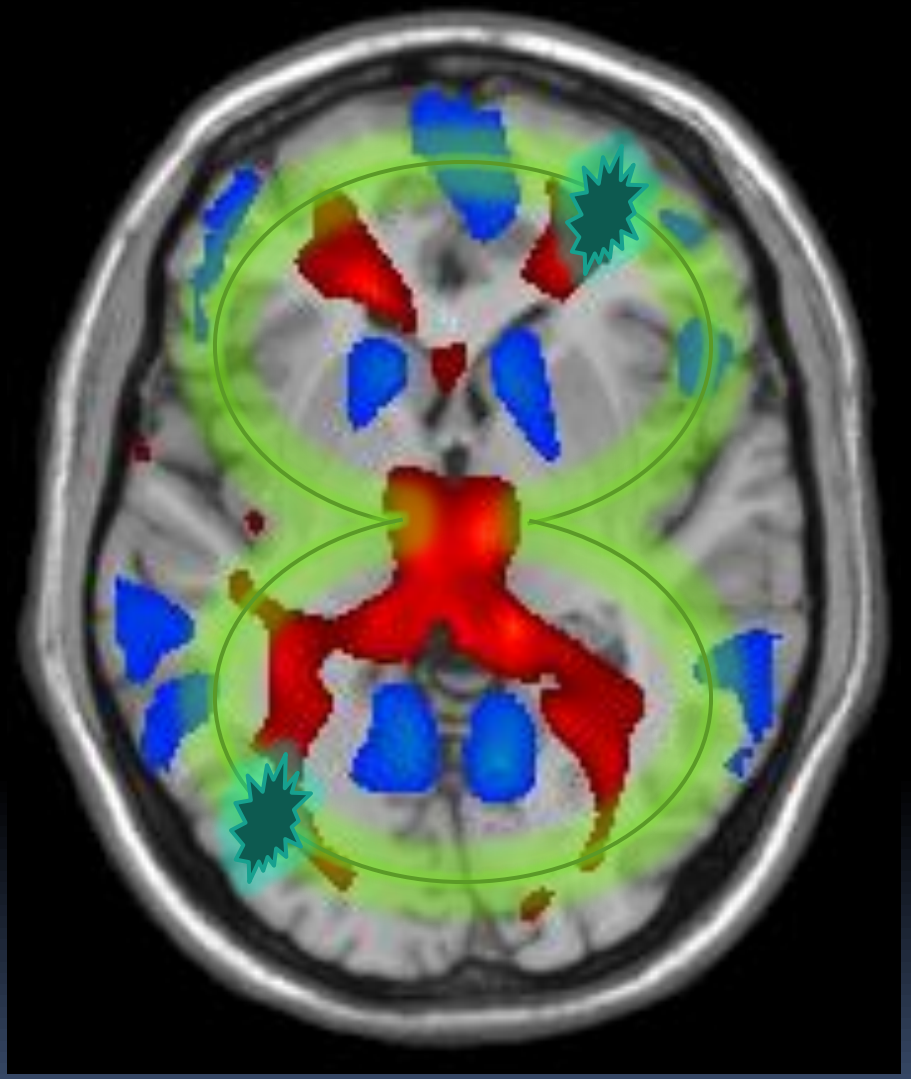
Focal seizures

Blume et al, Epilepsia 2001

- Evolving to bilateral, convulsive seizure
  - Previous terms: partial seizure secondarily generalized; secondarily generalized tonic-clonic seizure
  - With tonic, clonic or tonic and clonic components

Generalized - reconceptualized

- For seizures
  - *Generalized epileptic seizures are conceptualized as originating at some point within, and rapidly engaging, bilaterally distributed networks. …can include cortical and subcortical structures, but not necessarily include the entire cortex.*



Generalized seizures

- Tonic and clonic
- Absence
- Myoclonic
- Atonic
- Other types

Generalized seizures tonic and clonic

- Tonic and clonic (includes variations beginning with a clonic or myoclonic phase.
- Clonic seizures with tonic features and without tonic features.
- Tonic seizures

Generalized seizures

Absence

- Typical absence seizures
- Atpical absence seizures
- Myoclonic absence seizures

Absence seizure

- YouTube: Absence Seizure

GTC seizure

- YouTube: Generalized Tonic Clinic Seizure

Focal seizure with impaired awareness

- YouTube: Focal Seizure Involving face with Preserved Awareness
- YouTube: Focal Onset Impared Awareness

Generalized seizures
Myoclonic

- Myoclonic seizures
- Massive bilateral myoclonus
- Eylid myoclonus : with absence and without absence
- Myoclonic atonic seizures
- Negative myoclonic

Generalized seizures
Other types

- Reflex seizures in generalized epilepsy syndromes
- Seizures of the posterior neocortex
- Neocortical temporal lobe seizures

Generalized Seizures

Tonic-clonic (in any combination)

Absence
- Typical
- Atypical
- Absence with special features
  - Myoclonic absence
  - Eyelid myoclonia

Myoclonic
- Myoclonic
- Myoclonic atonic
- Myoclonic tonic

Clonic

Tonic

Atonic

Seizure types thought to occur within and result from rapid engagement of bilaterally distributed systems

Unknown

- With some types of epilepsy, the onset cannot be clearly determined as generalized or focal . Epileptic spasms are key example. Epileptic spasms, which include infantile spasms, are seizures that involve spasms of the muscles of the neck, trunk, and extremities

Recommended terminology for etiology

Use terms which mean what they say:

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

Genetic

- Concept: *the epilepsy is the direct result of a known or inferred genetic defect(s)*. Seizures *are the core symptom of the disorder.*
- Evidence: *Specific molecular genetic studies (well replicated) or evidence from appropriately designed family studies.*
- Genetic does *not* exclude the possibility of environmental factors contributing

Genetic syndrome

Syndrome group	Syndrome name	Abbreviation
Neonatal-infant	CDKL5-developmental and epileptic encephalopathy	CDKL5-DEE
	Dravet syndrome	DS
	Early infantile developmental and epileptic encephalopathy	EIDEE
	Epilepsy of infancy with migrating focal seizures	EIMFS
	Genetic epilepsy with febrile seizures plus	GEFS+
	Gelastic seizures with hypothalamic hamartoma	GS-HH
	Glucose transporter 1 deficiency syndrome	GLUT1DS
	Infantile epileptic spasms syndrome	IESS
	KCNQ2-developmental and epileptic encephalopathy	KCNQ2-DEE
	Myoclonic epilepsy in infancy	MEI
	Protocadherin 19 clustering epilepsy	PCDH19 clustering epilepsy
	Pyridoxine-dependent (ALDH7A1) developmental and epileptic encephalopathy	PD-DEE
	Pyridox(am)ine 5'-phosphate deficiency (PNPO) developmental and epileptic encephalopathy	P5PD-DEE
	Self-limited familial neonatal-infantile epilepsy	SeLFNIE
	Self-limited infantile epilepsy	SeLIE
	Self-limited neonatal epilepsy	SeLNE
	Sturge-Weber syndrome	SWS

Genetic syndromes

Child	Childhood occipital visual epilepsy	COVE
	Developmental and epileptic encephalopathy with spike-and-wave activation in sleep	DEE-SWAS
	Epileptic encephalopathy with spike-and-wave activation in sleep	EE-SWAS
	Epilepsy with eyelid myoclonia	EEM
	Epilepsy with myoclonic absences	EMA
	Epilepsy with myoclonic-atonic seizures	EMAtS
	Febrile infection-related epilepsy syndrome	FIRES
	Hemiconvulsion-hemiplegia epilepsy syndrome	HHE
	Lennox-Gastaut syndrome	LGS
	Photosensitive occipital lobe epilepsy	POLE
	Self-limited epilepsy with autonomic seizures	SeLEAS
	Self-limited epilepsy with centrotemporal spikes	SeLECTS
Idiopathic generalized epilepsies	Childhood absence epilepsy	CAE
	Epilepsy with generalized tonic-clonic seizures alone	GTCA
	Juvenile absence epilepsy	JAE
	Juvenile myoclonic epilepsy	JME

Genetic syndromes

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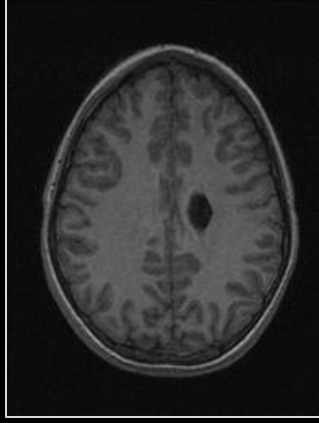
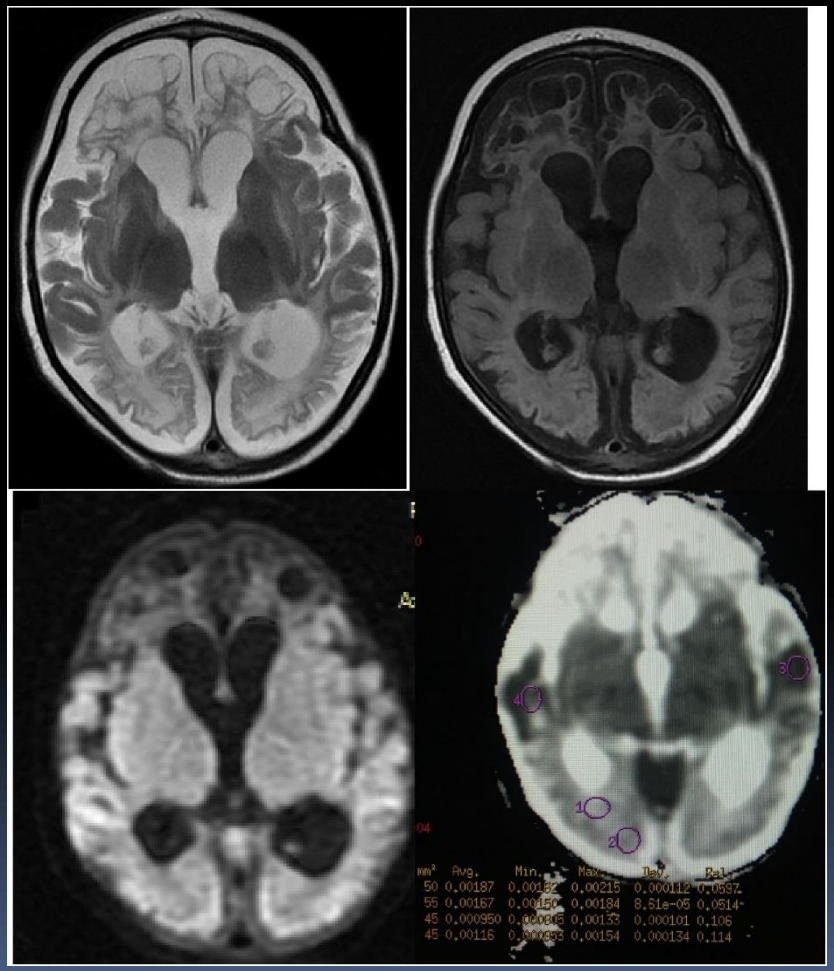
Structural



- . Virtually any insult to the cerebral cortex can cause a seizure.
- Acute: after minor head trauma (concussion), ischemia, or bleeding (intraparenchymal or subarachnoid hemorrhage).
- chronic disturbance of neuronal function caused by a remote event such as perinatal asphyxia or in utero stroke or the expression of a progressive neurologic disorder, such as a tumor or a neurodegenerative or neurometabolic disease.

HIE


mm² Avg. Min. Max. Dev. Rel.
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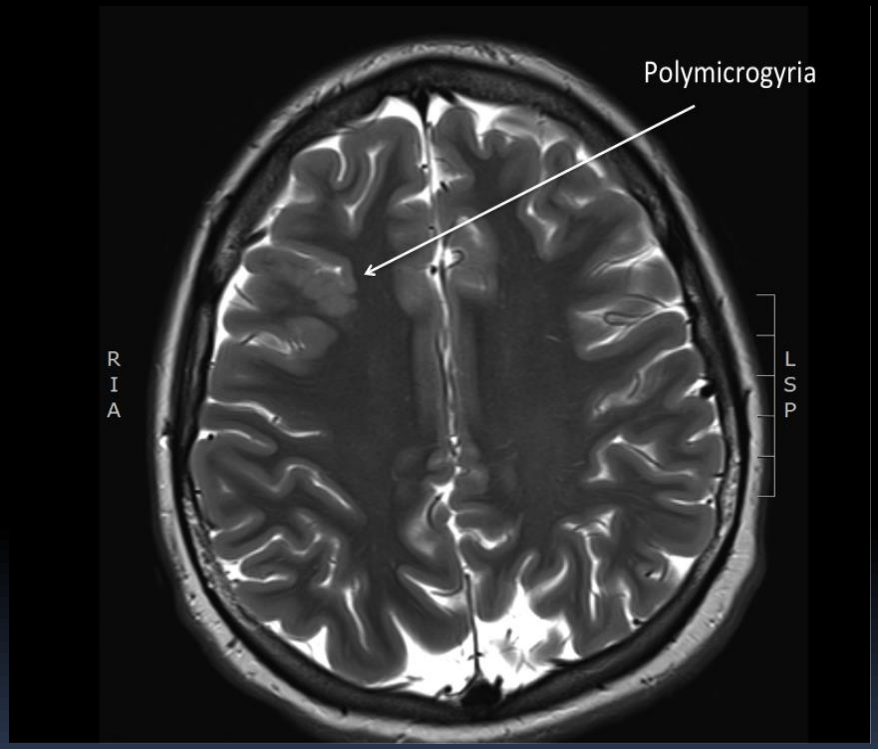
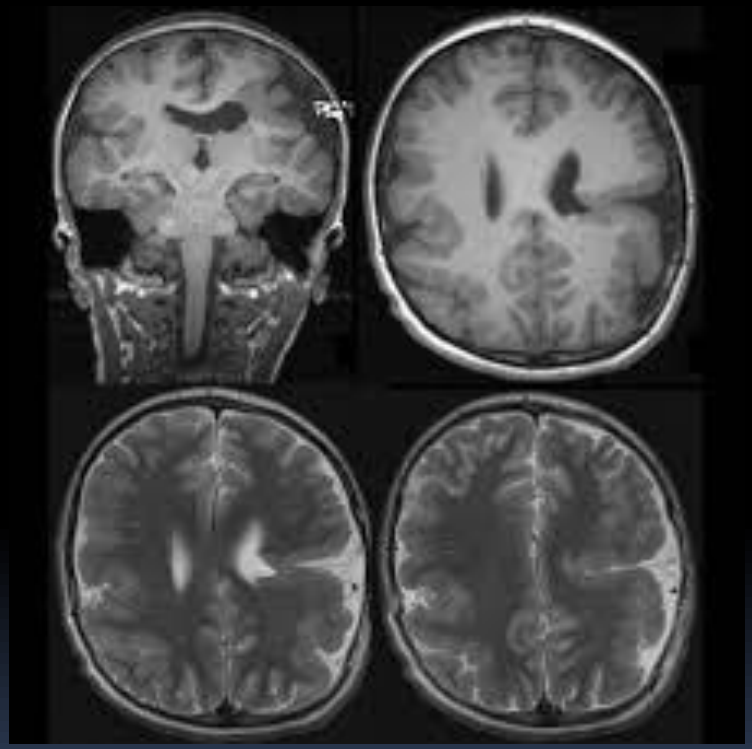
55 0.00167 0.00150 0.00184 8.51e-05 0.0514
45 0.000950 0.00076 0.00133 0.000101 0.106
45 0.00116 0.000993 0.00154 0.000134 0.114

Brain malformation





Metabolic

- Many metabolic conditions are associated with a substantially increased risk of developing epilepsy. Examples include glucose transporter deficiency, creatine deficiency syndromes, and mitochondrial cytopathies.
- Acute symptomatic seizures may be triggered by a transient disruption of cortical neuronal function such as a disturbed metabolic state (eg, high fever, hypocalcemia, hyponatremia).

Metabolic

- Immune-mediated central nervous system inflammation may cause epilepsy; examples include Rasmussen encephalitis and anti-N-methyl-D-aspartate (NMDA) receptor encephalitis.

Infectious

- Infections are the most common cause of epilepsy worldwide. Examples include HIV, neurocysticercosis, onchocerciasis, malaria, tuberculosis, and sequelae of prior meningitis or encephalitis.
- Acute symptomatic seizures can occur in the setting of chemical/inflammatory excitation caused by infection (eg, meningitis, encephalitis, sepsis).
- These acute seizures are often self-limited and do not persist beyond the infectious illness. However, some patients may develop epilepsy because of brain injury from the infection. It may take weeks, months, or even years for the seizures to reappear as epilepsy.
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Unknown

- Concept: T*he nature of the underlying cause is as yet unknown.*

International League Against Epilepsy: Updated classification of epielptic seizures (2025)