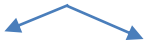
Pediatric Epilepsy

- **Epilepsy** is one of the **most common** chronic neurological disorders. It is characterized by **recurrent unprovoked seizures** or an **enduring predisposition** to generate epileptic seizures. If epilepsy begins in childhood, it is **often outgrown**. Seizures are **common** in childhood and adolescence. Approximately **3%** of children will experience a seizure.
- A seizure occurs when there is a **sudden change in behavior** or **sensation** caused by **abnormal** and **excessive electrical hypersynchronization** of neuronal networks in the cerebral cortex. Normal inhibition is overcome by excessive excitatory stimuli.
- If the cause of the seizures is known (for example: genetic, inborn errors of metabolism, metabolic (eg: low glucose, electrolyte abnormalities), structural (eg: malformations, tumors, bleeds, stroke, traumatic brain injury), infectious, inflammatory, or toxins) it is classified as **symptomatic**. If the cause is unknown, it is classified as **idiopathic**.

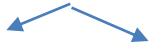
1. WHERE DID THE SEIZURE START? / WHAT KIND OF SEIZURE IS IT?

FOCAL ONSET	GENERALIZED ONSET	UNKNOWN ONSET
Seizure that **originates in a focal cortical area** with associated clinical features.	Seizure that involves **both sides of the brain** from the onset.	When it is **unclear** where the seizure starts.

2. IS AWARENESS IMPAIRED?



3. PROGRESSION TO BILATERAL?



Childhood Seizure Syndromes

- **Infantile Spasms (IS)**: epileptic spasms lasting seconds, that occur in clusters on awakening in infants < 1 year. West Syndrome Triad: IS, developmental regression/plateau, hypsarrhythmia on EEG. Care by neurologist required.
- **JME**: Age of onset can be as early as 9 – 10 years. Three seizure types – myoclonic, absence, GTCs. Myoclonic seizures are NOT the most common form of presentation. Most patients seek medical attention with the first GTC. GTCs are not uncommon. *Tx: valproic acid, lamotrigine, levetiracetam*
- **Rolandic**: variable frequency of seizures (not necessarily infrequent). Besides facial twitching, garbled or slurred speech is a common symptom. Often generalizes to a GTC/convulsive seizure. Age of onset 3 – 13 years (rare before age 3 years, and one would not think of Rolandic epilepsy if seizures started in the teens). Seizures may not require treatment if infrequent. *Tx: levetiracetam, oxcarbazepine/carbamazepine, clobazam*
- **Lennox-Gastaut Syndrome**: an epileptic encephalopathy, characterized by several different seizure types (tonic, atonic, generalized tonic-clonic, absence (atypical), etc.) that are often refractory. Seizure onset typically between 1-7 years but may be preceded by infantile spasms. Structural brain abnormalities and genetic etiologies are common. *Tx: lamotrigine and/or cannabidiol, valproic acid, clobazam, rufinamide*
- **Childhood Occipital Epilepsy**: Gastaut type: brief, frequent focal sensory visual seizures with onset from 18 months to 19 years. Remission in 50-60%. *Tx: carbamazepine*. Panayiotopoulos syndrome: infrequent focal prolonged autonomic seizures, onset in early childhood. Predominant symptom is prolonged episodes of nausea and vomiting. EEG: focal, high amplitude spikes over the occipital regions. Self-limiting. *Tx: oxcarbazepine/carbamazepine.*
- **Landau-Kleffner Syndrome**: rare disorder of acquired receptive aphasia and agnosia in children ≤ 6 years old. Abnormal EEG with continuous spike-waves during slow sleep (CSWS). Convulsive, non-convulsive seizures, or no seizures at all. *Tx: high dose diazepam.*
- **Reflex Epilepsy**: a specific stimulus (visual, auditory, etc.) or event triggers seizures (eg: photosensitive epilepsy in response to lights flickering). Absence of spontaneous seizures. *Tx: valproic acid, clonazepam, clobazam, lamotrigine.*

SEIZURE SEMIOLOGY (The terminology for seizure types is designed to be useful for communicating the key characteristics of seizures)

CLONIC: sustained rhythmical jerking movements.	**TONIC**: muscles stiffen or tense.	**ATONIC**: sudden loss of muscle tone, lasting seconds.	**MYOCLONUS**: sudden lighting-like jerk, may cluster.
EPILEPTIC SPASM: sudden flexion, extension, or flexion-extension of proximal and truncal muscles (1-2 seconds).	**AUTONOMIC**: eg: rising epigastric sensations, waves of hot or cold, rapid heart rate, etc.	**AUTOMATISMS:** Stereotyped, purposeless movements Eg: lip smacking, chewing, finger rolling, picking at clothes, etc.	**ABSENCE:** brief (≤ 10s), frequent (up to 100's) staring spells, associated with cessation of activity or impaired awareness.

Characteristic EEG Findings

- **Childhood Absence Epilepsy:** 3 Hz generalized spike-wave
- **JME:** 3-4 Hz generalized spike-wave and polyspike-wave
- **Rolandic:** Focal centro-temporal spike-wave activated by sleep

Adapted from:
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Updated by: Dr. Seema Adhami and Dr. Kristen Richard, June 2020

SEIZURE SAFETY

Educate patient/family about risks around water (swimming, bathing), fire/cooking, heights, and prolonged seizures.
No driving until cleared by an MD.

STATUS EPILEPTICUS

CAB – D stick, Get Access IV/IO, labs

IV Access:
LORAZEPAM (ATIVAN)
0.1mg/kg IV over 3 min repeat 15 min prn x1. Max single dose 4mg. Max total dose 6 mg.

No IV Access:
DIAZEPAM (VALIUM) PR
0.3-0.5 mg/kg(max 20mg)
Inject into lower 1/3 rectum
*high rate or recurrence due to rapid redistribution

LEVETIRACETAM (KEPPRA)
20mg/kg IV infusion, may repeat up to 2x to max 60mg/kg, dose not to exceed 1-3g

FOSPHENYTOIN
20mg PE/kg IV/IM Dosed in phenytoin equivalents (PE)
Infuse 3mg PE/kg/min, max 150mg PE/min

PHENOBARBITAL
20mg/kg IV Give over 15-20 min. Significant respiratory suppression likely with benzos and barbs together

If persistent,
PENTOBARBITAL coma
Loading 5mg/kg IV Infuse 1 to 3mg/kg/hr IV

ADDITIONAL CONSIDERATIONS

- **Labs:** d-stick, CBC, chem 12, VBG, b-HCG, tox screen, antiepileptic drug levels
- **Fever:** antipyretics. Empiric abx will not hinder dx meningitis
- **Hypoglycemia:** thiamine 100mg IV first if adult
- **Head trauma:** emergent head CT

DIAGNOSIS & MANAGEMENT

Medical history, including family history.
Seizure description: pre-ictal, ictal, and post-ictal.
Physical exam: thorough neurologic and mental status evaluation (eg: vital signs, response testing, memory recall testing, pupillary response, etc.).

DIAGNOSTIC TESTS:

- Electroencephalogram (EEG) – records the brain's electrical activity.
- Neuroimaging – CT or MRI to provide structural information.

COMMON ANTICONVULSANTS: oxcarbazepine carbamazepine, clobazam, clonazepam, ethosuximide, lamotrigine, levetiracetam, phenobarbital, phenytoin, valproic acid

Surgical evaluation is considered in **medically refractory** (failed 2 appropriately chosen and dosed medications) or **intractable** epilepsies.

Ketogenic diet: high in fats; low in protein and carbohydrates; may be considered in difficult to control epilepsy.

Medications

- Oxcarbazepine is now used far more often than Carbamazepine as it is easier to use – better tolerated, no need for monitoring levels (less prone to produce toxicity as CBZ) or blood counts. In fact, hardly any regular monitoring is needed