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, tumours, 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
Seizure that **originates in a focal cortical area** with associated clinical features.

GENERALIZED ONSET
Seizure that involves **both sides of the brain** from the onset.

UNKNOWN ONSET
When it is **unclear** where the seizure starts.

2. IS AWARENESS IMPAIRED? → YES / NO

3. PROGRESSION TO BILATERAL? → YES / NO

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.	**OTHERS:** change in cognition, emotions, or sensation. Behavior arrest.

CHILDHOOD EPILEPSY 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.

Juvenile Myoclonic Epilepsy: onset in puberty of myoclonic jerks, with early morning predominance. +/- generalized tonic-clonic and/or absence seizures. *Tx: valproic acid, lamotrigine, levetiracetam.*

Childhood Absence Epilepsy: brief (≤ 10 secs), frequent (up to 100's) staring spells with impaired awareness, provoked by hyperventilation (can test clinically). No post-ictal period. EEG pattern of 3 Hz. Peak age 5-6 years old, often outgrown. May be inherited. DDx: behavioral staring spells, inattentiveness, and focal seizures with impaired awareness. *Tx: ethosuximide, valproic acid, lamotrigine.*

Rolandic Epilepsy: infrequent, focal onset seizures arising from the Rolandic cortex, commonly occurring nocturnally or upon awakening. Episodes involve twitching or stiffness in the face. Seizures may not require treatment and generally cease by mid/late adolescence. *Tx: levetiracetam, 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: valproic acid, clobazam, stiripentol, 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: 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 SAFETY ⚠

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

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: ativan, 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.

March 2020
Katharine V. Jensen (Medical Student 2021, University of Alberta) &
Dr. Natarie Liu (Neurologist and Epileptologist, University of Alberta) for www.pedscases.com