SEIZURES AND EPILEPSY IN CHILDREN
Epidemiology of Seizures and Epilepsy in Children

- 4-6% incidence of a single seizure
- 1% incidence of epilepsy (≥ 2 unprovoked seizures)
- 70-80% achieve remission (“outgrow” seizures)
- HISTORY is the most important tool in differentiating a seizure from a non-seizure look-alike
- EEG is an adjunctive test to clinical history
- after 1st unprovoked seizure:
  - If EEG normal, 40% recurrence risk
  - If EEG abnormal, 80% recurrence risk
- 50% of 2nd unprovoked seizures occur within 6 months of 1st seizure
Epilepsy

- Definition: Two or more unprovoked seizures that occur at interval greater than 24 hours (i.e. recurrent seizures)
Causes

1. **Idiopathic (primary) in 80%** of cases
   - Genetic basis exist for many epileptic syndromes

2. **Organic (secondary) in 20%** of cases
   - Congenital cerebral malformation.
   - Degenerative brain diseases.
   - Post-traumatic.
   - Post-hemorrhagic.
   - Post-infection.
   - Post-toxic.
   - Post-anoxic.
ILAE 2017 Classification of Seizure Types Expanded Version

**Focal Onset**
- Aware
- Impaired Awareness

**Motor Onset**
- automatisms
- atonic
- clonic
- epileptic spasms
- hyperkinetic
- myoclonic
- tonic

**Non-Motor Onset**
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

**Generalized Onset**

**Motor**
- tonic-clonic
- clonic
- tonic
- myoclonic
- myoclonic-tonic-clonic
- myoclonic-atonic
- atonic
- epileptic spasms

**Non-Motor (absence)**
- typical
- atypical
- myoclonic
- eyelid myoclonia

**Unknown Onset**
- Motor
  - tonic-clonic
  - epileptic spasms
- Non-Motor
  - behavior arrest

**Unclassified**

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1. Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms.
2. These could be focal or generalized, with or without alteration of awareness.
3. Due to inadequate information or inability to place in other categories.

<table>
<thead>
<tr>
<th>Seizure Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Absence seizures</td>
<td>Transient loss of consciousness, with an abrupt onset and termination, unaccompanied by motor phenomena except for some flickering of the eyelids and minor alteration in muscle tone. Absences may be typical (petit mal) or atypical and can often be precipitated by hyperventilation.</td>
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<tr>
<td>Myoclonic seizures</td>
<td>Brief, often repetitive, jerking movements of the limbs, neck or trunk. Non-epileptic myoclonic movements are also seen physiologically in hiccoughs (myoclonus of the diaphragm) or on passing through stage II sleep (sleep myoclonus).</td>
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<td>Tonic seizures</td>
<td>Generalised increase in tone</td>
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<tr>
<td>Tonic–clonic seizures</td>
<td>Rhythmical contraction of muscle groups following the tonic phase. In the rigid tonic phase, children may fall to the ground, sometimes injuring themselves. They do not breathe and become cyanosed. This is followed by the clonic phase, with jerking of the limbs. Breathing is irregular, cyanosis persists and saliva may accumulate in the mouth. There may be biting of the tongue and incontinence of urine. The seizure usually lasts from a few seconds to minutes, followed by unconsciousness or deep sleep for up to several hours.</td>
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<tr>
<td>Atonic seizures</td>
<td>Often combined with a myoclonic jerk, followed by a transient loss of muscle tone causing a sudden fall to the floor or drop of the head.</td>
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</table>
2- Generalized tonic clonic seizures (Grand mal).

* The commonest form; pass in 3 phases.

Aura (pre ictal phase)
A warning signs before the attack may exist suggesting a focal origin of the epileptiform discharge: e.g. localized muscle spasm or paraesthesia.

Attack (ictal phase)
- Sudden loss of consciousness
- Tonic phase: tonic contraction of whole body → rigid posture, apnea, cyanosis, rolling of eyes & drolling of saliva.
- Clonic phase: rhythmic contraction & relaxation of all muscles groups → tongue biting & loss of sphincter control.

Post ictal phase
- Semiconscious for 30 min-2hr.
- Headache
- Sleep.
Absence (Petit Mal) Epilepsy

- Sudden onset of staring, interrupting speech or activity
- Occurs multiple times per day
- Short duration (seconds)
- Occurs in school aged children ~ 4-12 years, otherwise normal
Childhood Absence Epilepsy

- Typical onset between 4-8 years of age
- Staring and behavioral arrest
- Can have eye blinking or eye flutter
- Usually will last a few seconds with rapid return to baseline
- Typically occur daily, multiple times per day
- During brief episodes there is memory lapse → academic decline
- Often can be provoked with 2-3 minutes of hyperventilation
Childhood

- Frequency 8%
- Genetic predisposition- strong 20%
- Female preponderance 75%
- EEG: bilateral, synchronous, symmetrical 3 / sec spike wave, normal background.
- Seizures: Very frequent simple absences.
- Rx: VPA or ESM with control in 70 - 80%.
- Evolution: Remission- 95%.
- Rare persistence of absences only- 6%
Absence (Petit Mal) Epilepsy

EEG findings characteristic:
- bilateral generalized 3 Hz spike-and-wave discharges
- provoked by hyperventilation and photic stimulation
EPILEPSY SYNDROMES
Infantile Spasms

- Typical age of onset is between 4-7 months
- Rapid flexion of the trunk, neck and extremities (self hugging)
  - contraction can last for 5-10 seconds and relax 0.2-2 sec.
- Occur in clusters
  - Last < 1 min to 10-15 min
- The infant often cry (spasm)
- More common before sleep or upon awakening and all day
- Common associations
  - Tuberous sclerosis
  - HIE
Infantile Spasms (West Syndrome) – a severe epilepsy

Clinical spasms (1-2 secs)
- a subtle momentary flexion or extension of the body
- occur in clusters when drowsy (waking or falling asleep)

Severely abnormal EEG pattern: disorganized, discontinuous, high amplitude, multifocal spikes called **HYPSARRHYTHMIA**

Treatment: **ACTH**
Infantile Spasms

- **EEG – Hypsarrhythmia**
  - Triad of: high amplitude, disorganized background, multifocal discharges
- **Treatment**
  - Adrenocorticotropic Hormone (ACTH)
  - Vigabatrin if the spasms are symptomatic in a patient with Tuberous Sclerosis Complex
- **Prognosis**
  - If not treated early or effectively → increased risk of developmental delay and intellectual disability
Infantile spasms

- may be mistaken for colic, reflux, hiccups, or a startle!
- called **symptomatic** if etiology identified:
  - brain insult at birth (e.g., hypoxia-ischemia, meningitis)
  - brain malformation
  - neurocutaneous disorder (Tuberous Sclerosis)
  - metabolic disorder

- called **cryptogenic** if NO identifiable cause
- **prognosis best** (10% good outcome) if **idiopathic**
  - normal development at onset of infantile spasms
  - extensive etiology testing negative
- **prognosis poor** for:
  - seizure control (infantile spasms and future seizures)
  - future neurocognitive and developmental abilities
Lennox-Gastaut Syndrome – a severe epilepsy

- Often evolves from **infantile spasms**
- Neurodevelopmentally impaired children
- Syndrome defined by a **TRIAD** of:
  - 1. **mixed seizure types**: atonic, atypical absence, myoclonic, tonic-clonic, partial
  - 2. developmental delay
  - 3. abnormal EEG pattern: slow (< 2.5 Hz) spike wave discharges
- Symptomatic or cryptogenic etiologies (like IS)
- Prognosis poor
Juvenile Myoclonic Epilepsy (JME)

Seizure types:
- myoclonic in AM
- “grand mal”
- absence

EEG: bilateral generalized 4-6 Hz spike-wave or polyspike-wave activity
Juvenile Myoclonic Epilepsy

• **Keyword**
  - New onset seizure during *adolescence* (12-18 y)

• **Clinical Presentation**
  - Generalized tonic-clonic seizure
  - Jerking movements of the upper extremities in the morning

• **Risk factors**
  - Sleep deprivation (sleep over), stress, alcohol use
Juvenile Myoclonic Epilepsy

- EEG
  - Generalized polyspike-and-wave discharges at 4-6Hz

- Treatment
  - Valproic Acid

- Prognosis
  - Life-long risk of seizures
Benign Rolandic Epilepsy

Benign Focal Epilepsy of Childhood with Centrotemporal Spikes

EEG has characteristic pattern:

- bilateral
- independent
- centrotemporal spikes
Benign Rolandic Epilepsy

- **Treatment** recommended only if:
  - Seizures frequent (which is unusual)
  - Socially stigmatizing if occur in wakefulness
  - Anxiety provoking for parents if occur in sleep

- **Effective treatments:**
  - **Avoidance of sleep deprivation**
  - **Medications:** carbamazepine, oxcarbazepine
  - **Time** (outgrown by adolescence)
Other Epilepsy Syndromes

- Rett Syndrome
  - Occurs only in girls (X-linked lethal mutation)
  - Initial normal development → dev regression / autistic (loss of motor / language / social skills)
  - Acquired microcephaly (deceleration of head growth)
  - Hand wringing / alternating hand movements
  - Apnea / hyperpnea / breathholding
  - Seizures
Partial (Focal) Epilepsy

- Onset of seizure begins in one area of one cerebral hemisphere (apparent clinically or via the EEG)
- **AWARE:** no impairment of consciousness
- **IMPAIRED AWARENESS:** impairment of consciousness (staring)
- **FOCAL TO BILATERAL TONIC CLONIC:** partial seizure that ends in a generalized convulsion
Anatomic Onset of Focal Epilepsies

- Most frequently involved brain regions:
  - Temporal Lobe (80 %) >
    Frontal Lobe >>
    Parietal or Occipital
  - MRI or CT:
    - Normal or Abnormal
  - Neurologic exam:
    - Normal or Abnormal

Mesiotemporal sclerosis
Spells that mimic seizures

- Apnea / ALTE
- GER
- Sleep disorders (nocturnal myoclonus, night terrors, narcolepsy/cataplexy)
- Migraine variants (esp. aura)
- Benign breathholding spells
  - No neuro consult / lab / EEG / CT, Fe for cyanotic type
- Syncope
- Movement Disorders (tics, tremor, dystonia)
- ADD
- Behavioral Stereotypies (PDD)
- Pseudoseizures (psychogenic seizures)
  - Strange posturing, back arching, writhing
  - Alternating L and R limb shaking during same seizure
  - Psychosocial stressor
Breath-holding attacks

Temper

Occur in some toddlers when they are upset. The child cries, holds his breath and goes blue. Sometimes children will briefly lose consciousness but rapidly recover fully. Drug therapy is unhelpful. Attacks resolve spontaneously, but behaviour modification therapy, with distraction, may help.

Reflex anoxic seizures

Head trauma
Cold food
Fright
Fever

Occur in infants or toddlers. Many have a first-degree relative with a history of faints. Commonest triggers are pain or discomfort, particularly from minor head trauma, cold food (such as ice-cream or cold drinks), fright or fever. Some children with febrile seizures may have experienced this phenomenon. After the triggering event, the child becomes very pale and falls to the floor. The hypoxia may induce a generalised tonic-clonic seizure. The episodes are due to cardiac asystole from vagal inhibition. The seizure is brief and the child rapidly recovers. Ocular compression under controlled conditions often leads to asystole and paroxysmal slow-wave discharge on the EEG.
Treatment of epileptic seizures

- often not until after the second unprovoked seizure
- choice of AED based on maximum efficacy for that particular seizure type and minimal side effects
- 70% become seizure free on monotherapy
- an additional 15% become seizure free on polypharmacy
- 15% remain intractable
- Discontinue AED after 2 years seizure free EXCEPT for JME

Alternate treatments:
- Ketogenic diet (high fat diet)
- Vagal nerve stimulator – FDA approved for partial seizures in 12 years+
- Epilepsy surgery
Classic side effects of AEDs

- **valproic acid (Depakote):** hepatotoxicity, weight gain, acute pancreatitis
- **lamotrigine (Lamictal):** Stevens-Johnson syndrome
- **phenytoin (Dilantin):** gingival hypertrophy, acute ataxia, osteoporosis
- **phenobarbital:** adverse behavior / hyperactivity
- **carbamazepine (Tegretol):** agranulocytosis, aplastic anemia
- **oxcarbazepine (Trileptal):** hyponatremia
- **ethosuximide (Zarontin):** lupus-like reaction
- **topiramate (Topamax):** weight loss, acidosis, renal stones
- **felbamate (Felbatol):** aplastic anemia
- **gabapentin (Neurontin):** behavioral changes
Status Epilepticus

- Def: any type of seizure lasting > 5 minutes or repeated seizures without recovery between seizures.
- Seizures > 1 hour are associated with neuronal injury due to glutamate excitotoxicity.
- Evaluation and acute treatment for seizure > 5 minutes:
  - ABC’s (RR, HR, BP)
  - Check temp, glucose, electrolytes, CBC, renal and hepatic function, AED levels
  - Benzodiazepine □ phenytoin □ phenobarbital
Febrile Seizures

- 2-4 % of children age ~ 6 months – 6 years
- Provoked by a sudden spike in temp usually with URI, Acute OM, AGE (genetic predisposition)
- “Simple”
  - Generalized convulsion (whole body shaking)
  - Brief (< 15 minutes)
  - Only one in the course of an illness
  - Future risk of epilepsy (1%) SAME AS other children
- “Complex”
  - focal seizure (one side of body shaking, staring)
  - prolonged (> 15 minutes)
  - multiple in 24 hours
- Complex febrile seizures hint at an increased risk of future epilepsy
Febrile Seizures

- Indications for lumbar puncture:
  - Infants < 12 months
  - Prolonged complex febrile seizures
  - Febrile status epilepticus
  - Children who are partially treated with antibiotics
Treatment of Febrile Seizures (not epilepsy)

- **Considered benign** not warranting daily anti-seizure medication

- **Rectal Diastat** (valium gel) may be used to:
  - abort prolonged complex febrile seizure
  - prevent complex febrile seizure clusters (if child known to cluster)
  - prevent febrile seizure recurrence during a febrile illness

- **Anti-pyretics** have **NOT** been proven to decrease the risk of recurrent febrile seizures
Febrile seizures

- Affect 3% of children; have a genetic predisposition
- Occur between 6 months and 6 years of age
- Are usually brief, generalised tonic-clonic seizures occurring with a rapid rise in fever
- If a bacterial infection, especially meningitis, is present, it needs to be identified and treated
- Advise family about management of seizures, consider rescue therapy
- If simple – does not affect intellectual performance or risk of developing epilepsy
- If complex, 4–12% risk of subsequent epilepsy.