Epilepsy in Children

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Huntsville Grand Rounds

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Learning Objectives

- Recognize some common paediatric epilepsy syndromes/presentations
- Recognize the difference between focal and generalized epilepsy
- Describe how to choose an initial anti-epileptic

Outline

- Background on seizures and epilepsy
- Case presentations
  - Teaching format
- Summary of interesting aspects and teaching points after each
- Time along the way and at the end of each case for questions
- All cases are real patients – please respect their privacy

Seizures & Epilepsy

Definitions

- **Seizure:** a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain (ILAE)
- **Epilepsy:**

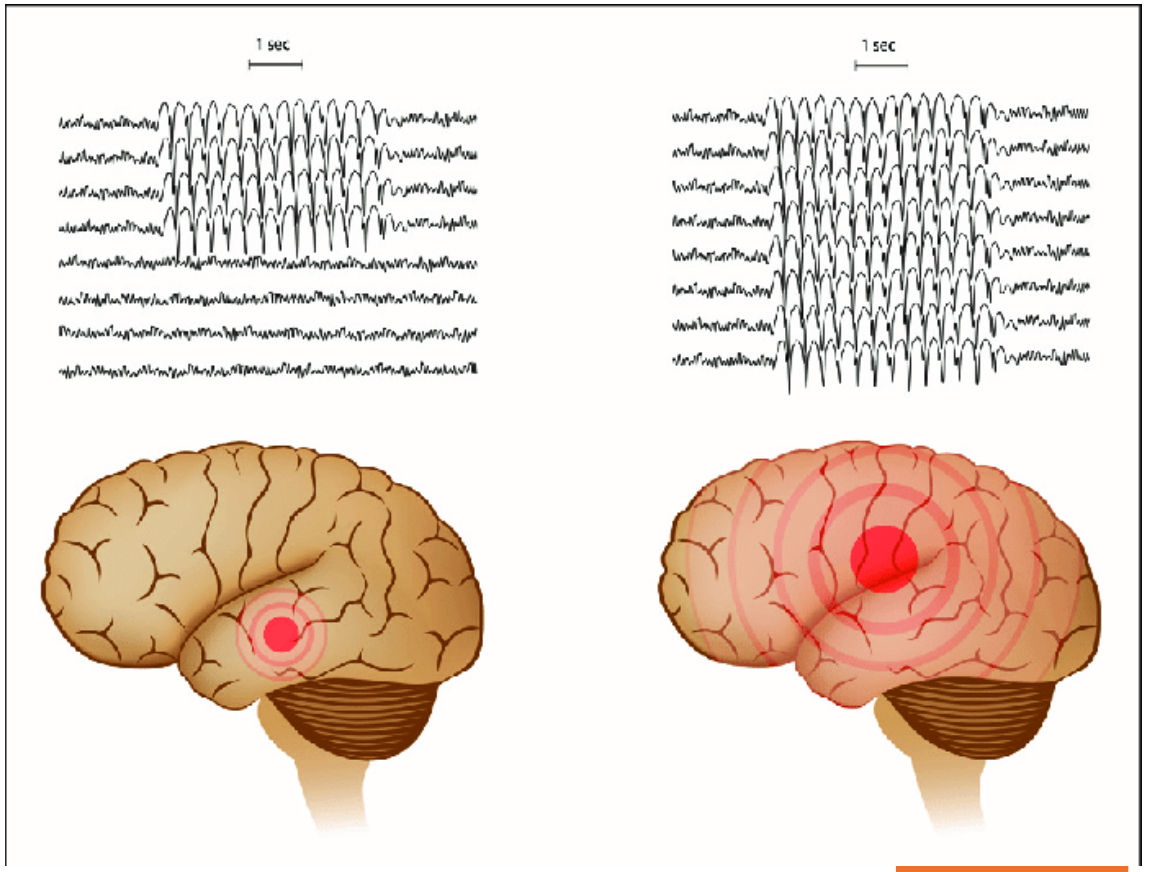
1. At least 2 unprovoked seizures on 2 separate occasions

2. 1 unprovoked seizure and a probability of further seizures (eg. abnormal EEG, brain malformation)

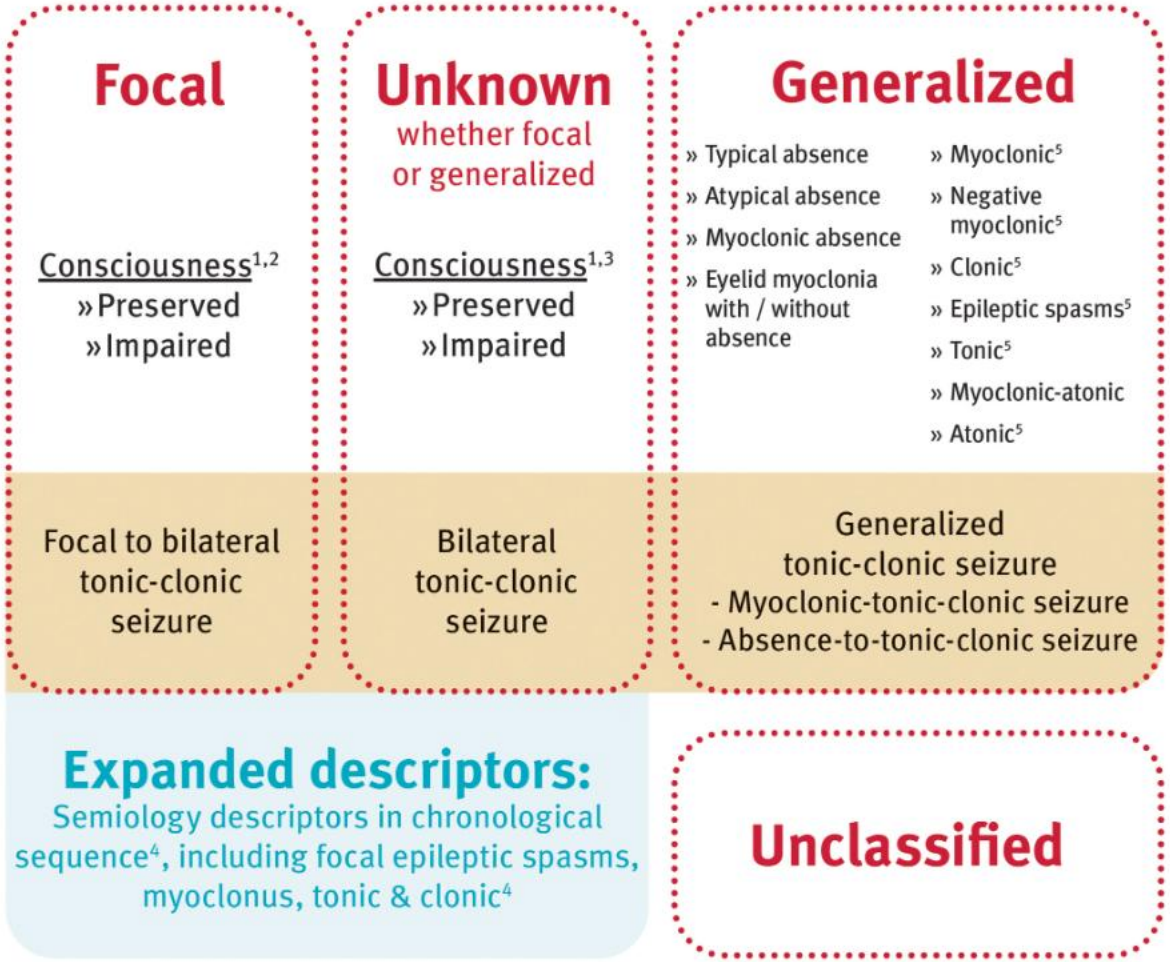
3. Diagnosis of an epilepsy syndrome

Focal vs. Generalized

- **Focal Seizure:** abnormal brain activity originates from one hemisphere of the brain
  - Often a more specific location is identifiable
- **Generalized Seizure:** abnormal brain activity originates from both hemispheres of the brain simultaneously



Seizure Classification



ILAE 2025

Why Classify?

- Helps identify etiology
- Helps identify epilepsy syndromes
- Guides further investigations
  - Focal: needs EEG, MRI
  - Generalized: maybe nothing
- Informs choice of anti-epileptics

Anti-Epileptic Drugs (AEDs)

Antiepileptic medications and seizure types

Seizure type	Antiepileptic drug
Broad spectrum: *all* seizure types (generalized from onset *and* focal onset seizures)	Clobazam, felbamate, lamotrigine, levetiracetam, rufinamide, topiramate, valproate, zonisamide
Narrow spectrum: focal with or without alteration in consciousness or awareness *and* focal evolving to bilateral convulsive seizure	Carbamazepine, eslicarbazepine, ezogabine, gabapentin, lacosamide, oxcarbazepine, perampanel, phenobarbital, phenytoin, pregabalin, primidone, tiagabine, vigabatrin
Absence seizure (a type of generalized seizure)	Ethosuximide

Case #1: LD

5 yo F

presenting with

staring spells

LD

Background

- Right-hand dominant
- **PHx:** healthy
- **Birth History:** term SVD, no complications
- **Development:** met all milestones
- **Medications:** None
- **Allergies:** None
- **Immunizations:** Up-to-date
- **Family history:** mother febrile seizures, maternal grandfather and great aunt epilepsy, brother NF1 with associated epilepsy

LD

Semiology

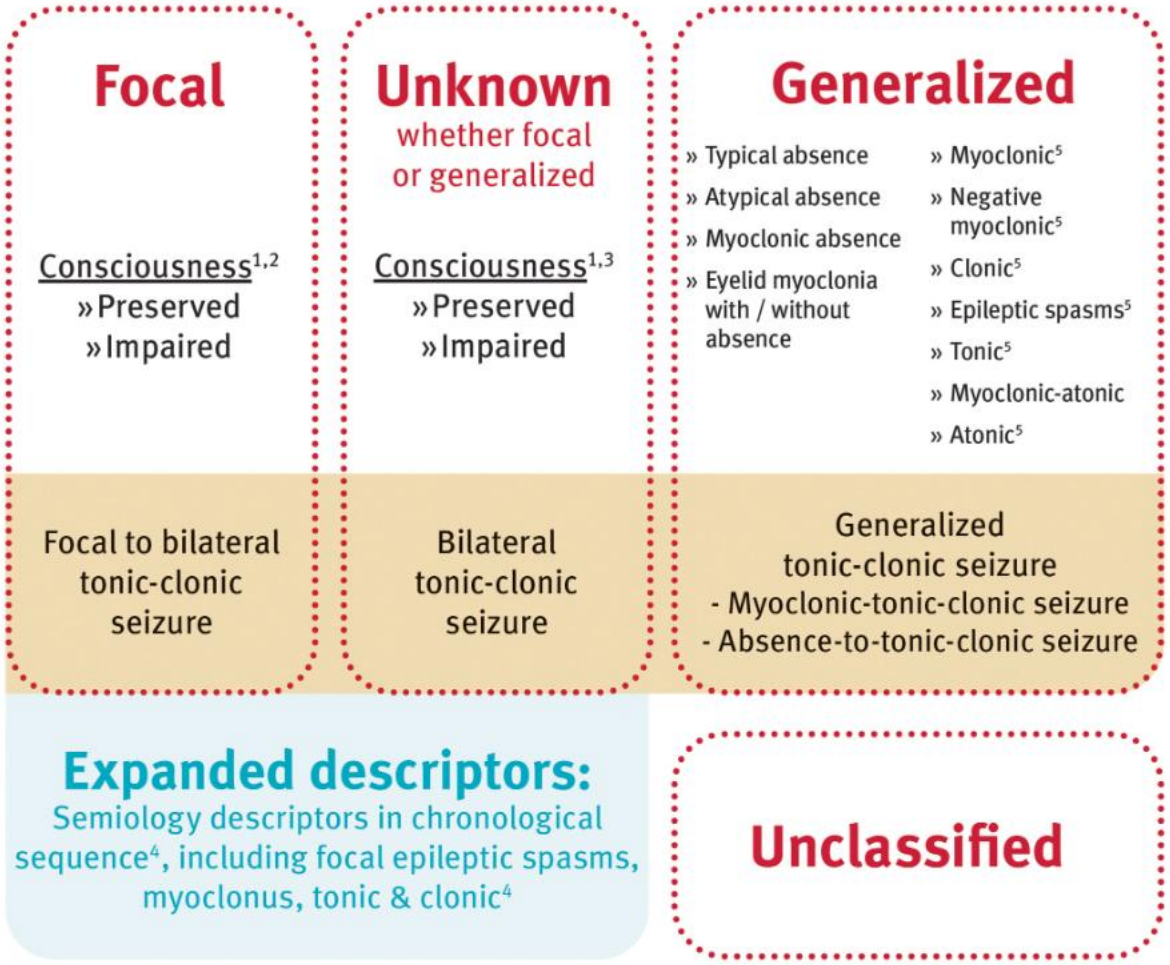
- Started at age 4
- Initial event was shaking of all 4 limbs, unresponsive, in context of fever
- Within 3 days developed episodes of "zoning out”
  - Complete behavioural arrest
  - Seems to be “mildly vibrating”
  - Will drop objects
  - Unresponsive to people calling her name
  - “snaps out of it”, looks confused for a second, then resumes what she was doing
  - Enuresis

LD

Additional History

- Events last no more than 30 seconds to a couple of minutes
- No warning in advance, no recollection after
- No post-ictal period
- Having at least 3-4 per day
- No triggers identified
- Sleep disturbance

Seizure Classification



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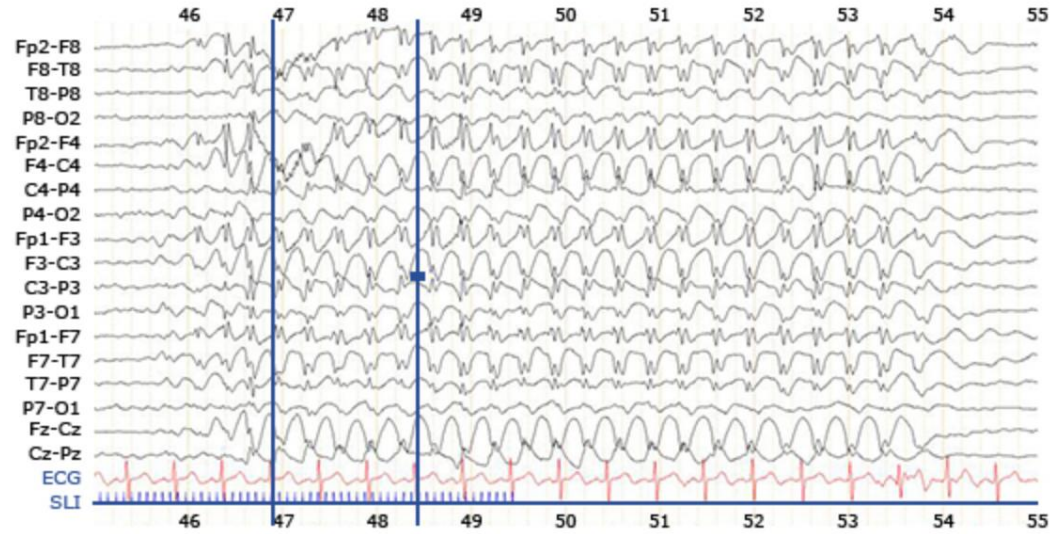
LD

Childhood Absence Epilepsy

- Generalized epilepsy
- Typical absence seizures
- Onset age 2-3 years (childhood) or 9-13 years (juvenile)
- Typical development – mild intellectual disability
- 3 Hz spike-and-wave EEG morphology
- 60-90% go into full remission
- Can develop other epilepsies (JME, IGE)
- Responds very well to certain AED



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LD

Treatment

- Started ethosuximide without waiting for EEG to confirm (family choice)
- No further seizures
- No further enuresis
- Sleep improved
- Had planned to up-titrate dose, but given excellent response family never increased from the initial prescribed dose

LD

Investigations

- EEG: Normal
  - On medication
  - Sensitivity of EEG?
- Epilepsy Genetics: normal

LD

Outcome

- Remained seizure-free on low-dose ethosuximide
- Stopped ethosuximide after 2-years seizure-free (family preference)
- Seizure-free ever since
- Doing great in all areas

LD

Takeaways

- Absence seizures are generalized seizures
- CAE is the most common generalized epilepsy syndrome in children
- Typical 3 Hz spike-and-wave EEG
- Treat with ethosuximide (or other AED targeting generalized seizures)
- Excellent prognosis, but lookout for other generalized epilepsies

Case #1: WL

9 yo M

presenting with

left-sided jaw-clenching and facial twitching

WL

Background

- Right-hand dominant
- **PHx:** healthy, ?dyslexia
- **Birth History:** unremarkable
- **Development:** met all milestones
- **Medications:** None
- **Allergies:** None
- **Immunizations:** Up-to-date
- **Family history:** no epilepsy, seizures, developmental delay, etc.

WL

Semiology

- Left jaw clenching and/or facial twitching
- Progressed face → arm → leg
- Some events are preceded by generally feeling odd and/or like he had a feeling an event was going to happen
- Completely aware during events and able to recount them after, though not able to speak during them
- Events typically lasted no more than 20 seconds
- Fatigued afterward for a few minutes up to 1 hour

WL

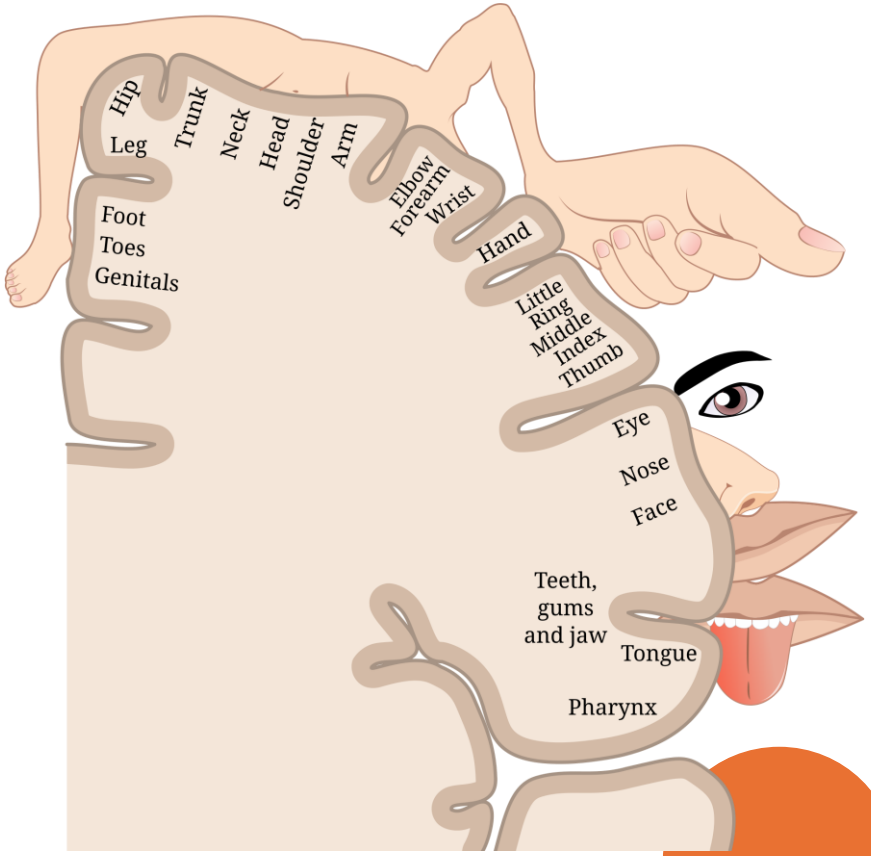
Additional Hx

- Typically first thing in the morning after waking up or just before going to bed
- Occurred in clusters – 1-4 in a month and then none for a few months
- Apparently decreasing in frequency over time

WL

Investigations

- 1st EEG: Normal background, right centrotemporal spikes, likely benign epilepsy with centrotemporal spikes (BECTS)
- MRI:
  - No epileptic focus



WL

SeLECTS

- Diagnosed with Self Limited Epilepsy with Centro-Temporal Spikes
  - Named for typical EEG appearance
- Most common focal epilepsy in children
- Developmentally normal school-age children
- Peak onset age 7-9 years
- Typical Jacksonian march
- Seizures mostly occur at bedtime/on waking or overnight
- Gradual decrease in frequency followed by spontaneous remission in most patients by age 12-13

WL

Next Steps

- Treatment optional
  - More recommended these days
- Family chose not to treat

WL

Next Steps

- Seizures progressed to secondary generalization
- Started as a typical seizure → gurgling/moaning in bed, tensed/hunched shoulders, unresponsive → post-ictal period x10 minutes
  - *(recommended AED)*
- 2 episodes of GTC seizures one weekend, after a busy week
  - one unwitnessed
  - other started as a typical focal seizure → GTC phase <1 minutes → post-ictal phase 10 minutes

WL

Treatment

- 1st line: clobazam (recommended in eConsult)
- Seizure-free for 6 months
- Developed frequent breakthroughs
  - Despite excellent adherence to medication
  - Good sleep hygiene
  - Eating and drinking well
  - Generally healthy

WL

Treatment

- 2nd line: oxcarbazepine
  - Cross-tapered from clobazam
  - Decreased clobazam by 20%/week
- Referred to neurology: likely atypical SeLECTS, no change in management recommended

WL

Outcome

- Stable and seizure-free on oxcarbazepine

WL

Takeaways

- Aura is a hint that seizures are focal
- SeLECTS is the most common childhood focal epilepsy
- Characterized by typical EEG pattern,nighttime events, Jacksonian march semiology, and gradual resolution of seizures over time
- No treatment necessary, but can treat per parent preference or if secondarily generalization
- Focal epilepsy → consider oxcarbazepine

Wrap-Up

Summary

- Distinguishing between focal and generalized seizures helps with diagnosis, investigation, and management
  - Focal epilepsy always needs an EEG and MRI
- Most common generalized epilepsy syndrome: CAE
- Most common focal epilepsy syndrome: SeLECTS

Quick Hits

- EEGs are 70% sensitive (at best)
- If there is an aura, think focal
- First line medications:
  - Generalized: levetiracetam (clobazam, VPA)
  - Focal: oxcarbazepine (carbamazepine)
  - Absence: ethosuximide

Thank You!

Any questions?