

# Nocturnal Frontal Lobe Epilepsy

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# Case Presentation

- Patient is a 30 year old female presenting to clinic at 24 weeks gestation (G3P2) with complaints of seizure activity two weeks prior to visit.
- States her first seizure-like episode occurred at age 18 with jerking, rolling eyes, tongue-biting, and foaming at mouth. She has similar seizures once every 2 – 3 months. Recurring seizures always occur during sleep around 6 AM.
- Patient's provider overseas prescribed Topimax (topiramate) at 50 mg b.i.d. for past 7 years with minimal relief.

# Diagnosis

- Nocturnal Frontal Lobe Epilepsy:
  - Most common sleep-related focal epilepsy<sup>1</sup> (70%)
    - Simple / Complex Partial Seizure
    - Secondary generalized seizure
  - Onset in adolescence or early adulthood<sup>1</sup>
  - Genetic link (most common)<sup>2</sup>
    - Autosomal Dominant with variable penetrance
    - *CHRNA4* gene related to cholinergic system
  - Structural neurodevelopmental lesions<sup>3</sup>

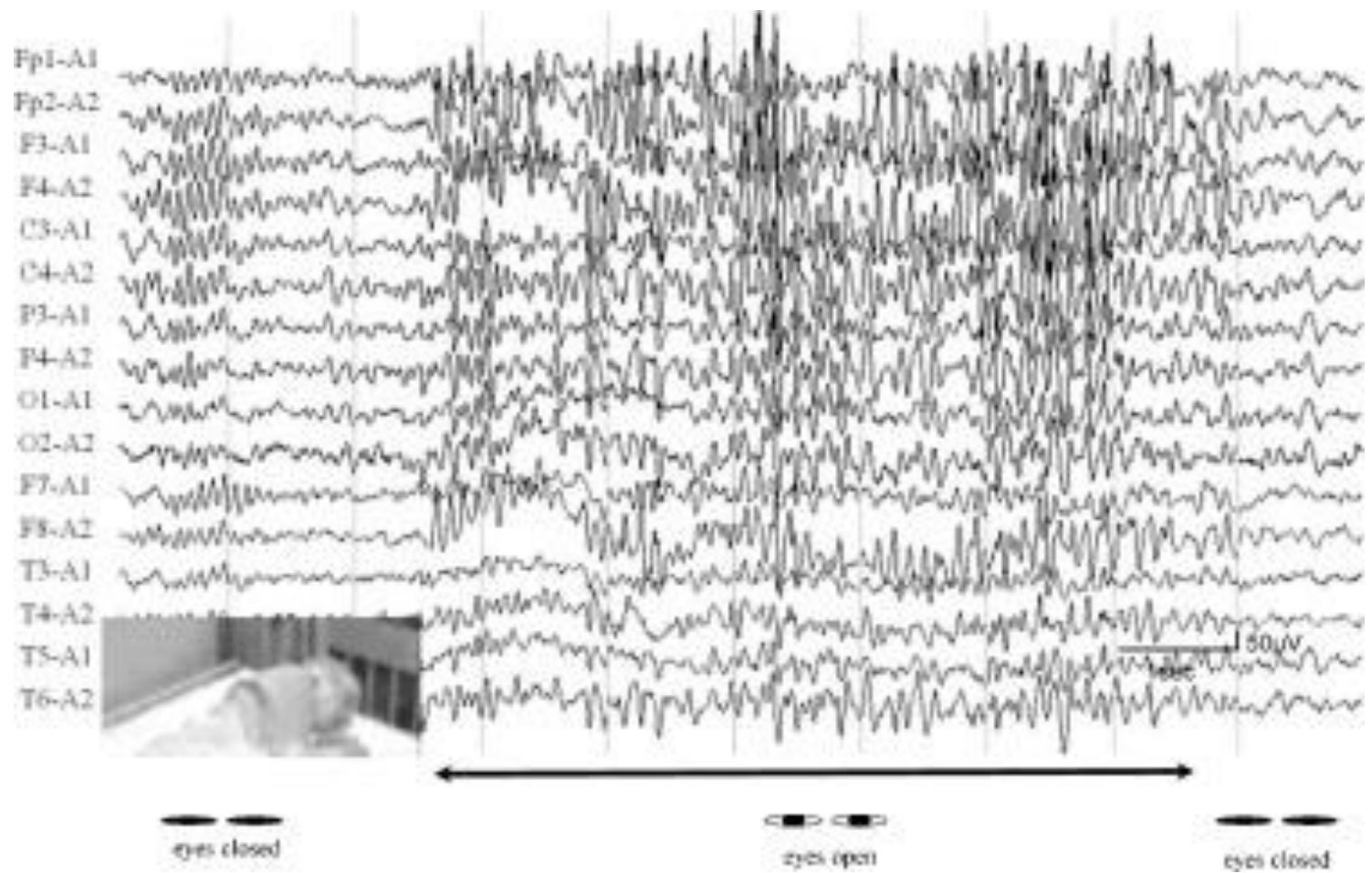


# Clinical Features

- Positive family history<sup>2</sup>
- 3 types:<sup>4</sup>
  - Short (2 – 4 seconds)
  - Sudden and brief (5 – 10 seconds)
  - Longer (up to two minutes)
    - Associated with parasomnias
- Multiple focal seizures, mostly in NREM sleep<sup>5</sup>
  - 28% in study had secondary generalized seizure

# Diagnostic Evaluation

- Presenting with multiple stereotyped motor events during sleep
- EEG<sup>6</sup>
  - Low interictal epileptiform discharge (IED) frequency + limited sampling of NREM sleep
  - Does not exclude diagnosis
- Asleep EEG repeat studies, all-night recording, polysomnography
- MRI recommended<sup>6</sup>



# Differential Diagnosis

- NREM parasomnias
- REM sleep behavior disorder (RBD)
- Periodic limb movements
- Psychogenic nonepileptic spells
- Nocturnal panic attacks



# Management

- Rare events
  - Clonazepam 0.5 – 1 mg at bedtime
- Anti-seizure drug therapy - lifelong

<b>Broad spectrum:</b>
Drugs used to treat a broad range of seizure types (both focal and generalized onset)
<ul style="list-style-type: none"><li>• Brivaracetam</li><li>• Clobazam</li><li>• Felbamate</li><li>• Lamotrigine<sup>§</sup></li><li>• Levetiracetam</li><li>• Perampanel</li><li>• Rufinamide</li><li>• Topiramate</li><li>• Valproate</li><li>• Zonisamide</li></ul>
<b>Narrow spectrum (focal):</b>
Drugs used primarily for focal-onset seizures (including focal evolving to bilateral convulsive seizures*)
<ul style="list-style-type: none"><li>• <b>Carbamazepine<sup>¶</sup></b></li><li>• Eslicarbazepine<sup>Δ</sup></li><li>• Gabapentin<sup>Δ</sup></li><li>• Lacosamide</li><li>• Oxcarbazepine<sup>Δ</sup></li><li>• Phenobarbital<sup>¶</sup></li><li>• Phenytoin<sup>¶</sup></li><li>• Pregabalin<sup>Δ</sup></li><li>• Tiagabine<sup>Δ</sup></li><li>• Vigabatrin<sup>Δ</sup></li></ul>
<b>Narrow spectrum (absence):</b>
Absence seizures only (a type of generalized seizure)
<ul style="list-style-type: none"><li>• Ethosuximide</li></ul>

# Management

- Counsel
  - Adequate sleep<sup>7</sup>
  - Seizure precautions<sup>8</sup>
  - Driving
- Pregnancy recommendations<sup>9</sup>
  - 6 months pre-pregnancy planning
  - Established pregnancy – no changes in drug regimen
    - Avoid **valproate**, carbamazepine, phenobarbital
  - Prenatal screen for malformations
  - Folic Acid and Vitamin K supplementation
  - Monitor blood levels: week 6, 10, once per trimester, postpartum
  - Not contraindicated in breast-feeding

# Patient Plan

- EEG
- Brain MRI
- Topiramate level
- Continue medication as previously prescribed
- Re-evaluate and adjust medications post-partum
  - Increase anti-seizure prevention and control

# Citations

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7. Hirshkowitz M, Whiton K, Albert SM, et al. National Sleep Foundation's updated sleep duration recommendations: final report. *Sleep Health* 2015; 1:233.
8. Thurman DJ, Logroscino G, Beghi E, et al. The burden of premature mortality of epilepsy in high-income countries: A systematic review from the Mortality Task Force of the International League Against Epilepsy. *Epilepsia* 2017; 58:17.
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# Questions