Nocturnal Seizures and Parasomnias

Objectives

- Create a differential diagnosis for complex nocturnal behaviors
- Discuss the differentiation of nocturnal seizures of frontal origin and disorders of arousal including episode timing, duration, semiology, and provoking factors
- Identify the more common sleep related epilepsies
- Describe the basis of the shared pathophysiology of nocturnal frontal lobe seizures and NREM disorders of arousal parasomnias
- Discuss current understanding of the etiology of nocturnal paroxysmal dystonia
Nocturnal Seizures and Disorders of Arousal from NREM Sleep

• Occur during entry into sleep, within sleep, or during arousals from sleep

• Have a broad range of semiology including autonomic nervous system changes, skeletal muscle activation, and seemingly purposeful, goal-directed complex behaviors outside consciousness

• Can have ill-defined EEG manifestations

• Are activated by sleep deprivation and stress
Parasomnias
Para = alongside of (Gk)
Somnus = events that accompany sleep (Latin)

- Undesirable events or experiences that occur during entry into sleep, within sleep, or during arousals from sleep
- Predominant features include ANS changes and skeletal muscle activation often with dream mentation
- Distinguished from sleep-related movement disorders by seemingly purposeful and goal-directed complex behaviors outside consciousness
ICSD 2 Classification of Parasomnias

• Disorders of Arousal (NREM)
  – Confusional Arousals
  – Sleepwalking
  – Sleep Terrors

• REM Parasomnias
  – REM Sleep Behavior Disorder
  – Recurrent Sleep Paralysis
  – Nightmare Disorder

• Other Parasomnias
  – Sleep-Related Dissociative Disorder
  – Sleep Enuresis
  – Sleep-Related Groaning
  – Exploding Head Syndrome
  – Sleep-Related Hallucinations
  – Sleep-Related Eating Disorder
  – Due to Drugs, Substances and Medical Conditions
Sleep Related Epilepsies

• Focal
  – Benign focal epilepsy of childhood with centro-temporal spikes (BECTS)
  – Frontal lobe epilepsy (FLE)
    – Supplementary sensorimotor area (SSMA)
    – Autosomal dominant nocturnal FLE (ADNFLE)
  – Nocturnal temporal lobe epilepsy

• Generalized
  – Lennox-Gastaut syndrome (LGS)

• Unknown
  – Epilepsy with continuous spike wave in sleep (CSWS)
Nocturnal Frontal Lobe Epilepsy

• Heterogeneous disorder characterized by seizures occurring exclusively or predominately during sleep

• “Frontal lobe semiology” of variable complexity and duration
  – Minor motor events: 2-4 sec stereotyped movements
  – Paroxysmal arousals: abrupt, 5-10 sec arousals accompanied by stereotyped movements of trunk and head
  – Major attacks (NPD): 20-30 sec

• Personal or family history of parasomnia in 34% and 39% of cases, respectively

• Autosomal dominant inheritance varies from 8-43%; sporadic and familial forms phenotypically similar

• Medical resistance observed in 30% of cases
Autosomal Dominant Nocturnal Frontal Lobe Epilepsy

- Clusters of motor attacks of increasing complexity and duration often encompassing dystonic, dyskinetic features or complex or violent behaviors
- Onset in childhood
- Considerable intra-familial variation
- Daytime seizures in 1/3
- Difficulty in waking, morning tiredness or EDS in 58%
- Medication-sensitive
- Mutations in nACHR subunit genes *CHRNA4*, *CHRNBB2*, *CHRRA2* and possibly CRH

Is there a Common Substrate for Nocturnal Seizures and Arousal Disorders?

- Seizures and parasomnias are produced by activation of Common Pattern Generators (CPGs)

- Neuronal networks capable of generating stereotyped, rhythmic motor patterns that underlie vital functions

## Differentiating Epilepsy and Arousal Disorders

<table>
<thead>
<tr>
<th></th>
<th>Epilepsy</th>
<th>Arousal Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age at onset</strong></td>
<td>Any age</td>
<td>Usually childhood</td>
</tr>
<tr>
<td><strong>Frequency</strong></td>
<td>Nightly</td>
<td>Several/month</td>
</tr>
<tr>
<td><strong>Event onset</strong></td>
<td>Abrupt</td>
<td>Gradual</td>
</tr>
<tr>
<td><strong>Semiology</strong></td>
<td>Stereotyped; EP features</td>
<td>Variable; no EP features</td>
</tr>
<tr>
<td><strong>Distribution</strong></td>
<td>Repetitive, clusters</td>
<td>First 1/3 of night; isolated</td>
</tr>
<tr>
<td><strong>Event duration</strong></td>
<td>&lt; 1 minute</td>
<td>Several minutes</td>
</tr>
<tr>
<td><strong>Sleep stage</strong></td>
<td>N2 &gt; N3</td>
<td>N3</td>
</tr>
<tr>
<td><strong>Natural history</strong></td>
<td>70% respond to AEDs</td>
<td>Spontaneous remission</td>
</tr>
</tbody>
</table>

Evaluation of Suspected Nocturnal Seizures

• Comprehensive history and examination
  – Rule out other causes of episodic cerebral dysfunction including syncope, transient ischemic attack/stroke, migraine, psychiatric disorders, parasomnias

• High resolution MRI
  – Etiology of focal epilepsy includes malformations of cortical development, head trauma, stroke, tumor, CNS infection

• Routine EEG

• Video PSG/EEG or longterm VEEG in epilepsy unit
  – Nightly episodes
  – Unusual or injurious episodes
  – Features suggestive of epilepsy
  – Failure to respond to conventional therapy
10-20 International System of Electrode Placement