NOCTURNAL SEIZURES

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NOVEMBER 3, 2018 11:30AM-12:15PM
• To become an EEG expert – this takes months, years of practice

Electroencephalography is like a beautiful park with a sign posted at the entrance: “For Persons with Total Commitment only” – Ernst Niedermeyer, MD (1920-2012)
LEARNING OBJECTIVES

• Learn basic nomenclature related to seizure activity
• Recognize abnormal EEG activity
• Be able to respond to patient having a seizure in the sleep lab safely
PRIESTESS OF DELPHI (1891)
John Collier, Art Gallery of South Australia
THE SACRED DISEASE

- Epilepsy known the “sacred disease” in antiquity
- prevailed for many centuries even after Hippocrates expressed his opposition to this widespread belief in his work On the Sacred Disease (5th C. BCE)
- Aristotle considered sleep “as an epilepsy”
WHAT IS A SEIZURE? WHAT IS EPILEPSY?

- **Seizures** are changes in awareness or behavior brought about by an abnormal discharge of electrical activity in the brain.
- **Epilepsy** is the tendency toward recurrent, unprovoked seizures.
EPILEPSY

- background EEG is usually normal in patients with epilepsy
- abnormal interictal EEG manifestations may include non-epileptiform abnormalities and interictal epileptiform discharges (IEDs)
- Patients with epilepsy may show generalized or focal slowing of the background, but most useful diagnostic finding supportive of a diagnosis of epilepsy is the activation of IEDs, which may be either focal or generalized
- IEDs must be carefully distinguished from benign variants, artifact or normal brain waves to avoid over-interpretation
EPILEPSY AND SLEEP

• 7.5 to 45 percent of people who have epilepsy have seizures mostly during sleep
• Sleep disorders more prevalent in epileptic population
• Nocturnal seizures usually occur towards the end of the sleep period (5am to 6am)
• Chances of capturing seizure in lab are low, even in patients with known epilepsy


Al-Biltagi MA. Childhood epilepsy and sleep
WHY MAY SEIZURES HAPPEN IN SLEEP?

• Epileptic seizures are often strongly influenced by the sleep-wake cycle
• change of state has effect on the brain’s epileptic activity; nocturnal seizures are triggered by changes in the electrical activity in your brain when moving between the different stages of sleep, and between sleep and wake

NREM

• occurrence of generalized discharges and clinical seizures mainly in NREM sleep, most IEDs in SWS
• can enhance IEDs in both focal and generalized seizures
• seizure activating role of NREM sleep has been attributed to increased neuronal synchronization within thalamo-cortical projection neurons with robust activation of epileptic ictal and interictal activity
• Most sleep-related seizures start during N2
• few seizures occur during N3 slow wave sleep (SWS)
• REM sleep limits spread of epileptic discharges outside the area that started seizure activity
• Few/no seizures in stage R
HOW TO RECOGNIZE A SEIZURE

- Short attention blackouts that look like daydreaming, Dazed behavior
- Sudden falls for no reason
- Lack of response for brief periods
- Unusual sleepiness and irritability when wakened from sleep
- Head nodding
- Rapid blinking
- Frequent complaints from the child that things look, sound, taste, smell or feel “funny.”
- Clusters of “jackknife” movements by babies who are sitting or laying down
- Clusters of grabbing movements with both arms in babies lying on their backs
- Sudden stomach pain followed by confusion and sleepiness
- Repeated movements that look out of place or unnatural

- A blank stare, followed by chewing, picking at clothes, mumbling, random movements
- Sudden fear, anger, or panic for no reason
- Muscle jerks of arms, legs, or body, especially in the early morning
- Odd changes in the way things look, sound, smell or feel
- Memory gaps
- Dazed behavior
- Being unable to talk or communicate for a short time
HOW TO RECOGNIZE SEIZURE ON EEG – CAN BE CHALLENGING FOR SLEEP TECHS

• number and placement of EEG electrodes
• experience of the sleep tech
• size and location of the ictal generator
• paper speed
HOW TO DESCRIBE WHAT YOU SEE

• Frequency
• Localization
• Amplitude
• Variability
HOW TO DESCRIBE WHAT YOU SEE

- Frequency
- Localization
- Amplitude
- Variability
DEFINITIONS

- **Interictal epileptiform discharges (IEDs)**
  - EEG isolated spikes, or sharp waves or spike-wave complexes
  - no behavioural change

- **Ictal**
  - EEG contains spikes and sharp waves lasting seconds/minutes
  - usually clinical seizure
WHAT IS A SPIKE?

- Epileptogenic waves are made up of spike waves and sharp waves.
- Spike wave = 20-70 ms while.
- Sharp waves duration 70-200 ms.
- Usually negative (up in PSG).
<table>
<thead>
<tr>
<th>EEG speed</th>
<th>PSG speed</th>
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<tr>
<td>10 sec page (30 mm/sec)</td>
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**Sharp waves**

**Spikes**

**Spike and wave**

**Polyspikes**

**Polyspike and wave**

1 second
WHAT IS A SPIKE AND WAVE?

- 2 different patients with childhood absence epilepsy
- (a) A typical spike–wave oscillation (3Hz)
- (b) additional spikes may be seen
# Classification - International League Against Epilepsy

## ILAE 2017 Classification of Seizure Types Expanded Version

<table>
<thead>
<tr>
<th>Focal Onset</th>
<th>Generalized Onset</th>
<th>Unknown Onset</th>
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<td>Non-Motor</td>
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<td>atonic</td>
<td>behavior arrest</td>
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- **focal to bilateral tonic-clonic**

1. Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms
2. Degree of awareness usually is not specified
3. Due to inadequate information or inability to place in other categories
ADVANTAGE OF PSG EEG

• interictal epileptiform discharges (IEDs) most sensitive and specific markers for seizure diagnosis
• routine EEG studies, which typically record for only 20–30 min
• routine EEG chance of recording clinical seizure is rare, IEDs are important
• Longer, ambulatory EEG studies do not provide sufficient information regarding whether a potential sleep disorder is present due to lack of respiratory and electromyography (EMG) channels
ADVANTAGE OF PSG EEG

- Seizures can be triggered by OSA which is highly prevalent among adults with epilepsy, with a prevalence rate around 40%, and is moderate-to-severe in 16%
- OSA fragments sleep and produces a state of chronic sleep deprivation (which increase seizures), one would expect a benefit from routine screening for OSA in adults with epilepsy

BEST MONTAGE

- AASM PSG: Referential montage F4-M1 C4-M1 O2-M1 (backup F3, C3, O1, M2)
- Advantages of limited EEG montage include reduced technical time for electrode placement, physician time for data analysis (and cost of data storage?)
- Difficult to differentiate during PSG between epileptic seizures, non-epileptic behavioral events, arousals, and artifacts
- Many discharges and other EEG abnormalities focal, likely have been missed, misinterpreted, or required more evaluation if a limited EEG montage was used

Practice Parameters for the Indications for Polysomnography and Related Procedures: “4.4.3.1 Polysomnography, with additional EEG derivations in an **extended bilateral montage**, and video recording, is recommended to assist with the diagnosis of paroxysmal arousals or other sleep disruptions that are thought to be seizure related when the initial clinical evaluation and results of a standard EEG are inconclusive.”


WHAT TYPES OF EPILEPSIES FEATURE SLEEP-RELATED SEIZURES?

- Juvenile Myoclonic Epilepsy (JME)
- Generalized Tonic-Clonic
- Benign Childhood epilepsy with centrottemporal spikes (BECTS) aka Benign Rolandic aka Benign Focal Epilepsy of Childhood
- Electrical Status Epilepticus of Sleep (ESES or CSWS)
- Lennox-Gastaut Syndrome
- Landau-Kleffner Syndrome (LKS)
- Frontal Onset Seizures (such as Nocturnal Frontal Lobe Epilepsy)
- Nocturnal Temporal Lobe Epilepsy
BENIGN CHILDHOOD EPILEPSY WITH CENTROTEMPORAL SPIKES (BECTS)

- Aka Benign Rolandic epilepsy
- Focal, idiopathic
- Most children will outgrow (starts ~ 3-13 with a peak around 8–9 years and ends around age 14-18)
- May feature altered speech, salivation, syncope
BECTS

• Two coexisting types of benign paroxysm BECTS and childhood epilepsy with occipital paroxysm (benign occipital childhood epilepsy) in a 10-yo girl with a hx of generalized tonic-clonic seizures

• Note the two independent BECTS at C3/T3 and C4/T4
BECTS

- (A) 12 y.o. girl w. hx of nocturnal generalized tonic-clonic convulsions with postictal aphasia and right arm weakness (Todd’s paralysis); EEG shows spikes maximum at C3
- (B) 10-year-old boy w/ hx of nocturnal generalized tonic-clonic convulsions; EEG shows spikes maximum at T4
GENERALIZED TONIC-CLONIC

- Tonic - limbs stiffen, body rigid
- Clonic - after period of stiffening, limbs and face begin to jerk rhythmically
- may have loss of bowel or bladder control
- afterwards: confusion, fatigue, headache

Tonic – muscle tone

Video: Nguyen Tri Phuong Neurologic Department
Onset of the ictal event in this generalized seizure consists of low-voltage, rhythmic beta-range fast activity, with progressively increasing amplitude and decreasing frequency. Followed by generalized spike-wave bursts, which become progressively slower in frequency and less rhythmic toward the end of the seizure. EEG suppressed during the immediate postictal period and is then followed by the appearance of postictal delta activity. Clinically, the initial fast activity corresponds with the tonic phase and the subsequent spike-wave bursts coincide with the clonic phase of the seizure. During the ictal events, EEG activities are largely obscured by muscle and movement artifacts.
GENERALIZED TONIC-CLONIC

- (A) generalized ictal event in a 25-yo man with hx of seizures since childhood

- Note the sudden flattening of electroencephalography (EEG) activity at the onset, followed by beta activity peeking through the massive electromyogram (EMG) artifact during the tonic phase of seizure

• (B) Toward the end of the seizure, ictal discharges changed to periodic spike-wave discharges, which were contaminated by muscle artifact (clonic phase)

• Afterward, there was postictal suppression of EEG activity
NOCTURNAL FRONTAL LOBE EPILEPSY

- sudden awakening with violent movement, dystonic or tonic posturing, and hyperactive behavior
- resembles night terror or RBD
- EEG may be normal or may show frontal slowing or spikes
- cluster of short fits, rapid start on/off
- sudden battering movements during sleep, head jerks to one side, upper limbs rising into brief, frozen state
- Behavioral automatisms eg rocking, bicycle pedaling movements and repetitive hand movements
NOCTURNAL FRONTAL LOBE EPILEPSY

- (A) epileptiform discharges Wake
- (B) epileptiform discharges in Sleep over midline and frontal region

Successful epilepsy surgery in frontal lobe epilepsy with startle seizures: a SEEG study. Epileptic Disorders. Volume 17, issue 4, December 2015
CASE STUDY 15 YO MALE W GENERALIZED EPILEPSY

- Intractable seizures, NDD, autism, nephrocalcinosis
- Presented in infancy with West Syndrome
- Enalapril, Topamax (topiramate), Zarontin (ethosuximide), Clobazam, vit B6
- Referred for snoring, gasping, pauses in breathing overnight, daytime sleepiness/fatigue, unable to stay awake during the day recently, more frequent naps
- initiated on APAP in community
LFF 0.3Hz HFF 35Hz Notch 60Hz Sens 7uV/mm
30s/page 10 mm/s

Body Position: Left

10/10/2018 21:16:48
Lights Off
10/10/2018 21:16:51
Lights have to stay on until sleep onset due to seizures

2/1101
*Left
100.0/1000
LFF 0.3Hz HFF 35Hz Notch 60Hz Sens 7uV/mm
30s/page 10 mm/s
LFF 0.3Hz HFF 35Hz Notch 60Hz Sens 7uV/mm
30s/page 10 mm/s
Good ventilation 19/12cmH2O R=12
Poor ventilation BiLevel 19/12cmH2O R=12
LFF 0.3Hz HFF 35Hz Notch 60Hz Sens 7uV/mm
10s/page 30 mm/s
Spike-wave complexes 2-4Hz

LFF 0.3Hz HFF 35Hz Notch 60Hz Sens 7uV/mm – 10s/page 30 mm/s
Time: 0526h

LFF 0.3Hz HFF 35Hz Notch 60Hz Sens 7uV/mm
30s/page 10 mm/s
So absence in sleep??

Generalized motor seizure with tonic component

- Characteristics are 3 Hz spike wave complex
- Appears and goes of abruptly on normal background activity
- Maximum at frontal and midline region
- Starts at 4 Hz then slows down to 3.5 Hz then up to 2.5 Hz
- Hyperventilation precipitate such attacks
- Paroxysm of more than 5 sec leads to clinical seizure
BENIGN VARIANTS

- “of uncertain clinical significance”
- E.g. wicket waves, rhythmic mid-temporal theta of drowsiness (RMTD or RTTD), benign small sharp spikes (BSSS, sometimes denoted BSST for benign small sleep transients), 14 and 6 positive spikes, 6-Hz “phantom” spike and wave, subclinical rhythmic EEG discharge of adults (SREDA), and the midline theta rhythm of Ciganek
- Are they reproducible? Play with the software, Filters, Montages, Use the referential montage, Get a repeat EEG, Get the context
- Many people have IEDs, some will develop epilepsy, some have diagnosed or undiagnosed neurological conditions but no clinical seizures
HOW DO YOU SCORE IT?

• Occasionally whole record will be abnormal EEG, lacking recognizable sleep features
8 y.o. female with Mitochondrial Myopathy, seizure disorder

LFF 0.3Hz, HFF 35Hz, notch 60Hz, sens 15uV/mm, 10mm/sec (30s/page)
HOW DO YOU SCORE IT?

• Richard Rosenberg, PhD ‘I wouldn’t even try’

• Use the pediatric scoring set (if available), and score as NREM vs REM vs W, or even just NREM vs W

https://www.aastweb.org/blog/author/richard-rosenberg-phd
WHAT HAPPENS WHEN A PATIENT HAS A SEIZURE IN THE SLEEP LAB?

REMAIN CALM!
ALL IS WELL!!!
HAVE A PLAN (AND KNOW THE PLAN)

Before the study:

- P&P
- Familiarize yourself with the names of anti-epileptic medications
- Talk to the patient, ask about seizures - when diagnosed? controlled? when was last seizure? how does it manifest?
KNOW THE COMMON TRIGGERS

- Sleep deprivation
- Alcohol intake
- Stress
- OSA
- photosensitive patients, seizures can be precipitated by television

https://www.epilepsy.com/learn/professionals/refractor-y-seizures/actions-take/identify-and-reduce-triggers
EPILEPSY ACTION PLAN - IS IT AN EMERGENCY?

A seizure is generally considered an emergency when:

- Convulsive (tonic-clonic) seizure lasts longer than 5 minutes
- Or repeated seizures without regaining consciousness
- = Status Epilepticus
- types of status epilepticus: convulsive and non-convulsive
- Convulsive status epilepticus requires emergency treatment by trained medical personnel in a hospital setting as it may be life-threatening

EPILEPSY ACTION PLAN

- Communication - other techs
- Be reassuring, comforting and stay calm
- Track and record time, and note as much detail as possible (time, description of movements)
- Basic 1st aid:
  - Environment - recovery position, remove objects, protect head (pillows, rolled up sheets)
  - Do not restrain
  - Do not put anything in mouth
  - Keep airway open and watch breathing
  - Stay with them until fully conscious
  - Call your MD

DIFFERENTIAL DIAGNOSIS

• 3 m.o. infant, in PICU, grandma reports frequent seizures
• Upon pre-study interview, reported to sleep tech unusual eye movements when asleep
• EEG showed normal sleep stages, no apparent spikes/spike waves, no clinical seizures
• LAMF, low chin EMG, and eye movements

“His eyes go all ‘funny’. I’ve had 3 children and many grandchildren and none of them ever did this”

“Oh look, there he goes, he’s doing it!”

St R with REMs!