Non-Respiratory Indication for Sleep Studies in Children

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Objectives

- Review Indications for PSG according to current (2005) AASM guidelines
- Review key diagnostic features on PSG for non-respiratory sleep disorders
- Discuss indications of the multiple sleep latency test (MSLT) and maintenance of wakefulness test (MWT)

Disclosures

- I have no financial relationships with commercial entities.
- I receive research support from Autism Speaks, Inc.

Polysomnogram (PSG)

- EEG
- EOG
- EMG
- Oral-Nasal airflow
- Nasal airway pressure
- Respiratory effort
- Oxygen saturation
- Carbon Dioxide monitoring
- ECG
- Video/audio
- Gastroesophageal pH

Indications for PSG

- Narcolepsy/Hypersomnia
- Nocturnal Seizures
- Sleep related Behavior that is violent or atypical of parasomnias
- Periodic Limb Movements

Hypersomnolence

- Hypersomnia: recurrent episodes of excessive daytime sleepiness (EDS) or prolonged nighttime sleep that affects the everyday life
  - Present for >3 months
  - No universally accepted definition of EDS in children


http://www.childrenshospital.org/gallery/indicators/AD/Slidepage.html
Presentation in Childhood

- Excessive sleepiness in child considered >5 y who continues to nap during day (unplanned/inappropriate situations) despite normal (>10 hours) nocturnal sleep
- Maybe difficult to wake in the morning
- Sleeps >2 hours longer on weekends compared to weekdays
- May have behavioral problems such as impaired attention, irritability, impulsiveness, emotional dysregulation

Utility of Polysomnogram

- Exclude sleep disorders
  - Periodic limb movements of sleep
  - Sleep disordered breathing
- Confirm sleep period
  - 6-10 hours without long sleep time
  - >10 hours with long sleep time.

Multiple Sleep Latency Test

- Quantifies the likelihood of falling asleep in quiet environment during the main wake period of the day.
- Allows 5 nap opportunities at 2-hour intervals in the daytime
- Started 2 hours after PSG documenting normal night sleep
- EOG, EEG, EMG, ECG
- Prior to MSLT
  - May be asked to have drug screen (even between naps)
  - Regular sleep/wake cycle (logs) for 2 weeks
  - Discontinue stimulants, antidepressants, psychotropic medications 2 weeks before PSG
  - Take into account half-life of drugs

DDx of Hypersomnia

- Idiopathic hypersomnia with long sleep time
  - "excessive" amounts of sleep at night (>10 hours) and at least 1 daytime nap (>1 hour)
- Idiopathic hypersomnia without long sleep time
  - "normal" sleep period of 6-10 hours

Common PSG findings in Hypersomnia

- Sleep latency <10 minutes
- Normal sleep efficiency >85%
- Decreased frequency of arousals
- Some may demonstrate increase in NREM sleep
- REM latency typically normal
- Normal total sleep time

MSL in Hypersomnia

<table>
<thead>
<tr>
<th>Mean Sleep Latency (MSL)</th>
<th>Stage 1</th>
<th>Stage 2</th>
<th>Stage 3/4</th>
<th>REM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Less than 10 min suggestive of hypersomnia</td>
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<tr>
<td>Normal 15-20 minutes in healthy children</td>
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<tr>
<td>Less than 5 minutes concerning for narcolepsy</td>
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<tr>
<td>No baseline MSLT values have been established for children &lt;8</td>
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Littner MR et al. SLEEP 2005
Narcolepsy

- Prevalence is 0.05% in the United States
- Nearly half of patients had onset prior to 15 years of age and 10% prior to age 5 years
- Early studies have suggested median delay between onset and diagnosis of 10 years
- Classic tetrad includes hypersomnolence, cataplexy, sleep paralysis and hypnagogic hallucinations
- Disturbed nocturnal sleep

Narcolepsy in Children

- Highly variable in children
- Cataplexy -60-90% with onset following EDS months to years later
- Hypnagogic Hallucinations-Ranges 10-89% and may vary by pubertal status
- Sleep paralysis-Ranges 0-78% and may vary by pubertal status
- Disturbed sleep-85-100%

Suggested Diagnostic Criteria

<table>
<thead>
<tr>
<th>Diagnostic criteria</th>
<th>Narcolepsy, cataplexy</th>
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<tbody>
<tr>
<td>1. Narcolepsy with cataplexy</td>
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<td>2. Disturbances of arousal</td>
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<td>3. REM onset linked to sleep onset</td>
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<td>4. Hypnagogic hallucinations</td>
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PSG and MSLT features

- PSG and MSLT: May show early REM period, typically highly fragmented sleep
- MSLT: Usually less than 8 min and most less than 5 minutes
- Sleep Onset REM Periods: 2 naps with REM latency <20 minutes

Before a PSG and MSLT...

- Establish Regular sleep/wake cycles
- Sleep Logs
- Avoid caffeine for 24 hours
- Stimulants, stimulant-like medications, and REM suppressing medications should ideally be stopped 2 weeks before MSLT
- No naps during the daytime

Parasomnia and Seizure Disorders

- Parasomnias are disorders of arousals occurring in NREM sleep and can be injurious to patient and result disruptions to sleep
- Common, uncomplicated and non-injurious parasomnias should be dx by clinical evaluation alone (standard)
- If sleep behavioral is atypical (age, onset, timing duration, frequency) or described as stereotypical, repetitive, or focal PSG is indicated (guideline)

Kushida et al. SLEEP 2005
Derry et al. SLEEP 2006
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Nocturnal seizures

- In largest reported case series of difficult to diagnose nocturnal paroxysmal events, 50% ultimately dx with sleep related epilepsy
- Nocturnal frontal lobe seizures can be particularly difficult to differentiate because of bizarre, hyperkinetic movements without clear loss of consciousness exclusive to sleep

Diagnostic Studies

- Routine EEG with sleep captured are often sufficient to establish diagnosis of sleep related seizure disorder
- PSG considered should be considered when:
  - Other sleep disturbances suspected that may result in increased seizure frequency
  - No response to conventional treatment
  - Trauma/personal injury
- Difficulty in differentiating parasomnias and seizures may necessitate video monitoring with extended bilateral montage EEG

Periodic Limb Movement of Sleep

- PLMS is considered a nocturnal non-epileptic myoclonic like movement with stereotyped, intermittent and repetitive movements of limbs during sleep
- Pediatric prevalence ranges from 5-26% of patients with description of restless sleep and limb twitching at night

Diagnostic Criteria

PLMD is diagnosed when the following are present:
1. PLMS documented by polysomnography
2. PLMS exceeding norms for age (≥ 5/h for children)
3. Clinical sleep disturbance or daytime fatigue
4. The absence of another primary sleep disorder or reason for the PLMS, including RLS and OSA.

PSG Characteristics

- PLMS can cause serious disruption to sleep:
  - Increased arousal index
  - Increased number of awakenings
  - Increase in N1 REM stage 1 sleep

Maintenance of Wakefulness Test

- Objective Measure of one’s ability to stay awake in a quiet environment for defined period of time
- Helpful to know volitional ability to stay awake vs tendency to fall asleep (i.e. if medication for hypersomnia or narcolepsy working, residual EDS in cases of SDB)
- Requires optimization of sleep night before
MWT

- Series of four 40 minute trials
  - Trial terminated if no sleep in 40 min or sleep (3 continuous epochs of stage 1 and 1 epoch of any other sleep) achieved.
  - Mean sleep latency less than 8 min abnormal in adults
  - Highly variable only 60% of normal subjects remained awake for all 4 trials!
- Has been used for assessment of narcolepsy for not standard of care:
  - Results of the 20 minute MWT showed a MSL of 6.0 with a SD of 4.8 for patients with narcolepsy.

Conclusion

- Non-respiratory indication of PSG limited per Practice Parameters to PLMS, specific subtype of parasomnia, nocturnal seizures and hypersomnia/narcolepsy
- Detailed history is key in sleep evaluation in order to assess if PSG needed
- MSLT and MWT can be useful diagnostic tools but norms for children still unclear
- Lab based sleep studies have specific indications and prerequisites to optimize utility