To Refer or Not To Refer: Sleep Evaluation is the Question

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Objectives

• Have a basic understanding of different sleep disorders
• Know risk factors associated with different sleep disorders
• Be aware of common signs and symptoms associated with sleep disorders
• Understand when a sleep evaluation referral is needed
Common Sleep Problems in Developmentally Delayed Individuals

- Obstructive Sleep Apnea
- Circadian Rhythm Disorders
- Insomnia
- Hypersomnia
- Parasomnias
- Sleep Related Movement Disorders
Contributors to Sleep Disorders

- Intrinsic brain abnormalities
  - Brain chemicals (neurotransmitters) of sleep and wake
  - Circadian rhythm Modulators
    - Superchiasmatic Nucleus
    - Melatonin Regulation
  - Homeostasis Modulators
    - Sleep Pressure
    - Autonomic Nervous System
  - Hormone regulation
    - Cortisol
    - Testosterone
    - Estrogen
  - Seizures
  - Breathing centers in brain stem
    - Messages about when and how to breathe
Contributors to Sleep Disorders

- Medical illnesses
  - Obesity
    - Airway Obstruction
    - Diminished Breathing Effort
  - Cardiac Disease
    - Arrhythmias
    - Central Sleep Apnea
  - Pulmonary disease
    - Hypoxemia
    - Hypercarbia
  - Diabetes Mellitus
    - Energy Metabolism
  - Kidney Disease
    - Fluid Balance
  - Pain
    - Hyperarousal
    - Restlessness
  - Seizures
    - Hyperarousal
Contributors to Sleep Disorders

- Other primary sleep disorders
  - Obstructive Sleep Apnea
  - Central Sleep Apnea
  - Circadian Rhythm Disorders
  - Parasomnias
  - Movement disorders
Contributors to Sleep Disorders

- Medications
  - Sedating Medications
    - Anti-seizure medications
    - Hypertension medications
    - Muscle relaxants
    - Sleep aids
  - Stimulant Medications
    - Stimulants
    - Modafinil
    - Caffeine
    - Antidepressants
  - Psychotropic Medications
    - Antipsychotics
    - Antidepressants
    - Anti-anxiety medications
Contributors to Sleep Disorders

- Psychiatric and Behavioral factors
  - Anxiety
  - Depression
  - Obsessive Compulsive Behaviors
  - Cognitive Problems
  - Caregiver Interactions
Common Sleep Problems in Developmentally Delayed Individuals

- Obstructive Sleep Apnea
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Obstructive Sleep Apnea

- Repeated pauses in breathing interrupting sleep
  - At least 10 second duration
  - Oxygen desaturation
  - Brain interruption
- Associated with many medical and behavioral problems
- Risk Factors:
  - Body Habitus
  - Facial Features
  - Muscle Tone
Sleep Apnea Cycle

- Physiologic stressors:
  - Cyclic hypoxemia
    - 11,911 adults – 41% systemic hypertension
  - Strenuous respiration
  - Sympathetic activation
  - Reduced total sleep time

Tkacova, Eur Respir J. 2014
Untreated Sleep Apnea
1868 subjects followed 20 years

<table>
<thead>
<tr>
<th>Incidence of multiple organ diseases higher in OSA Group:</th>
<th>32% with OSA</th>
<th>68% without OSA</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Hypertension:</td>
<td>79%</td>
<td>25%</td>
</tr>
<tr>
<td>• Coronary heart disease:</td>
<td>56%</td>
<td>24%</td>
</tr>
<tr>
<td>• Pulmonary heart disease:</td>
<td>6%</td>
<td>4%</td>
</tr>
<tr>
<td>• Stroke:</td>
<td>27%</td>
<td>7.5%</td>
</tr>
<tr>
<td>• Diabetes:</td>
<td>12%</td>
<td>5.4%</td>
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</tbody>
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Ci SP, 2011 (Article in Chinese)
Signs/Symptoms May Vary With Age

- **Children**
  - Pauses in breathing
  - Secondary enuresis
  - Hyperactivity
  - Morning headache

- **Middle Age**
  - Pauses in breathing
  - Snoring
  - Excessive daytime sleepiness
  - BMI > 35, Neck > 16/17

- **Older > 60 years**
  - Not feeling well rested
  - Nocturia ≥ 3
Obstructive Sleep Apnea Symptom: SNORING

- Children
  - 10%
- Age 30
  - 20% men
  - 5% women
- Age 60
  - 60% men
  - 40% women

20-35% of habitual snorers have OSA
Physical Findings on Exam Which May Predispose to OSA

BMI > 35

Neck Circumference
- > 16 in women
- > 17 in men
Physical Findings on Exam Which May Predispose to OSA

- Crowded Oropharynx
- High arched narrow palate
- Low laying palate
- Large uvula
- Narrow posterior oropharynx
- Tonsillar hypertrophy

http://yoursmileyourstyle.com/files/2014/05/What-your-dentist-looks-for-in-diagnosing-sleep-apnea.jpg
Physical Findings on Exam Which May Predispose to OSA

- Chronic nasal congestion
- Nasal speech
- Obligate mouth breather
  - adenoidal hypertrophy
- Mandibular retrognathia
- Floppy Eye Syndrome
  - AKA Ectropion
  - 38/45 patients (85%) had OSA
  - 65% had severe OSA

STOP-BANG: Quick Screening Tool for OSA

Answer each of the following “yes” or “no”:

- Do you **SNORE** loudly (louder than talking or loud enough to be heard through closed doors)?
- Do you often feel **TIRED**, fatigued, or sleepy during daytime?
- Has anyone **OBSERVED** you stop breathing during your sleep?
- Do you have or are you being treated for high blood **PRESSURE**?
- **BMI** more than 35?
- **AGE** over 50 years old?
- **NECK** circumference > 15.75 inches?
- Male **GENDER**?

≥ 3 “yes” answers = High-risk for OSA
< 3 “yes” answers = Low-risk for OSA
OSA Summary

Most Common Symptoms:
- Snoring
- Pauses in Breathing
- Excessive Daytime Sleepiness

Treat to Reduce Morbidities:
- Cardiovascular
- Neurological
- Behavioral
Common Sleep Problems in Developmentally Delayed Individuals

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Biologic Clocks Influence All
Circadian Rhythm Disorders

- Misalignment of internal sleep/wake rhythm and the desired (or required) time for sleep
- Desire for sleep and wakefulness at inappropriate times
- Risk Factors
  - Intrinsic brain abnormalities
  - Blindness
  - Genetic predisposition
Common Sleep Problems in Developmentally Delayed Individuals

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Insomnia

- Hard to sleep
  - Difficulty going to sleep
  - Difficulty staying asleep
  - Waking up too early
  - Poor quality sleep
- Risk Factors:
  - Hyperarousal
  - Environment
  - Habit
  - Medication
Reasons for Insomnia

- Lack of sports, leisure
- Inability to relax
- Genetics
- Work shifts
- Circadian rhythm
- Illnesses
- Nutrition
- Environment, noise
- Relationship, family
- Coffee, alcohol, drugs
- Stress, worries, anxiety, depression

SensitiveSleepers
Acute Insomnia

- Acute Insomnia
  - < 3 month duration
  - In isolation
    - Often a known stressor/cause
  - Comorbid to medical or mental health condition
    - Improves with treatment for underlying cause
Chronic Insomnia

- Chronic Insomnia
  - > 3 months
  - 3+ nights per week
- Subtypes:
  - Psychophysiological Insomnia
    - “Trained” by habit
    - Worse in usual sleep environment, better in different environment
    - Excessive worry about not sleeping
  - Idiopathic Insomnia
    - Longstanding and persistent
    - Often starts in childhood
    - No sustained remission
Chronic Insomnia

- Paradoxical Insomnia
  - Sleep state misperception
  - Report of very little to no sleep
  - Appear to have normal sleep on objective measures of sleep (PSG)
  - Evidence of altered sleep/wake arousal system
- Inadequate Sleep Hygiene
  - Variable sleep schedule
  - Daytime napping
  - Use of sleep-disruptive products
    - Caffeine, tobacco alcohol
    - Electronic devices
Chronic Insomnia

- Behavioral insomnia of childhood
  - Improper sleep training or limit setting
  - Sleep-Onset Association Type
    - Dependence on specific stimulation or object to fall asleep
  - Limit-Setting Type
    - Bedtime stalling
    - Bedtime refusal
    - Poor limit setting by caregiver
- Mixed Type
  - Combination of both
Secondary Insomnias

- Insomnia due to a mental health disorder
- Insomnia due to a medical condition
- Insomnia due to a drug or substance
Insomnia Summary

- Behavioral interventions are most effective
- Acute insomnia often resolves when primary reason is resolved
- Chronic Insomnia can be
  - Lifelong
  - Formed by habit
  - Due to other medical or behavioral factors
  - Worsened by other sleep disorders

Refer if:
- Acute Insomnia persists for unclear reasons
- Over the counter medications to help with sleep are being used nightly
- Medications being used for insomnia are not effective
- Concern about underlying reason
  - OSA
  - Hypoxemia
  - RLS
  - PLMD
  - Seizures
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Hypersomnia

• Too Much Sleep
  • Excessive duration of sleep
  • Excessive depth of sleep
  • Excessive frequency of sleep episodes

• Risk Factors
  • Intrinsic
  • Genetic
  • Medication
Hypersomnia

- Narcolepsy
  - Due to loss of orexin secreting neurons
  - Genetic predisposition
  - Autoimmune component
  - Symptoms:
    - Sleep attacks
    - Sleep Onset latency of < 8 minutes
    - Sleep onset REM period
    - Sleep Onset hallucinations
    - Sleep/wake instability
      - Too sleepy in the day
      - Hard to sleep soundly during the night
- Narcolepsy Type I
  - Cataplexy
    - Sudden loss of muscle tone
    - Precipitated by strong emotions
    - Retain Consciousness
- Narcolepsy Type II
  - No cataplexy
Hypersomnia

- Idiopathic hypersomnia
- Klein-Levin Syndrome
  - Hypersomnia
    - 2-5 week duration
    - Recurrent at least < every 18 months
  - At least one during episode:
    - Cognitive dysfunction
    - Anorexia or hyperphagia
    - Disinhibition
    - Altered perception
Hypersomnia

- Hypersomnia due to a medical disorder
  - Parkinson’s disease
  - Post traumatic
  - Genetic disorders
    - Prader Willi
    - Myotonic dystrophy
    - Moebius syndrome
    - Fragile X syndrome
  - Brain tumors
  - CNS infections/lesions
  - Endocrine disorders
    - Hypothyroidism
  - Metabolic encephalopathy
  - Residual sleepiness in those with adequately treated OSA
Hypersomnia

- Hypersomnia due to a medication
  - Sedating medication
  - Substance abuse
  - Stimulant withdrawal
- Hypersomnia associated with a psychiatric disorder
  - Mood disorder
  - Somatoform disorders
  - Schizoaffective disorder
  - Adjustment disorder
  - Personality disorders
- Insufficient sleep syndrome
  - Common in teens
Hypersomnia Summary

• A known factor in many neurodevelopmental syndromes due to:
  • Intrinsic factors
  • Medical morbidities
  • Medications
  • Other sleep disorders

• Refer if:
  • Concern for underlying cause
    • OSA
    • Seizures
    • Overmedicated
Common Sleep Problems in Developmentally Delayed Individuals

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Parasomnias

- Unwanted nocturnal behaviors
  - Simple or Complex
  - Routine behaviors
  - Inappropriate behaviors
- Consciously unaware
- Familial Pattern
  - Neither parent affected
    - 22% of children sleepwalk
  - One parent affected
    - 45%
  - Both parents affected
    - 60%
- Predisposing, priming and precipitating factors involved
NREM Parasomnias

- Often in the first third of the night
- More Common in children
  - Typically from NREM 3 sleep
- Increased with:
  - Sleep deprivation
  - Sickness
  - Stress
  - Side effects to medications
Parasomnias
During NREM Sleep

• Night Terrors
  • Episodes of abrupt terror
  • Intense fear
  • Autonomic arousal
  • Inconsolable
  • Eyes open
  • Brief to 30+ minutes

• Confusional Arousals
  • Mental confusion or confused behavior
  • Absence of terror or ambulation
Parasomnias During NREM Sleep

- Sleep walking
  - Somnambulism
- Sleep talking
  - Somniloquy
- Groaning during Sleep
  - Catathrenia
- Bedwetting
  - Enuresis
- Teeth Grinding
  - Bruxism
- Sleep related eating disorders
  - Eating while asleep
  - Variant of sleepwalking
Parasomnias During REM Sleep

- REM Behavioral Disorder
  - Dream enactment
  - Usually Brief
  - Recall intact upon awaking
  - Themes of being pursued/fear
  - Associated with neurodegenerative diseases
    - Predate onset of Alpha-synucleinopathies by years
      - Parkinson's Disease
      - Multiple systems atrophy
      - Dementia with Lewy Bodies
    - Lesions affecting brain stem
      - Multiple Sclerosis
      - Narcolepsy
      - Stroke
  - Medications
    - Antidepressants

- Associated with neurodegenerative diseases
Parasomnias During REM Sleep

- Nightmares
  - Recall intact upon awaking
  - Second half of the night

- Sleep Paralysis
  - Awake but unable to move

- Sleep related hallucinations
  - Hypnogogic
    - When falling asleep
  - Hypnopompic
    - When waking up
Parasomnia Summary:

- Parasomnias can be caused by:
  - Sleep deprivation
  - Sickness
  - Stress
  - Side effects to medications
  - Sleep disorders

- Refer if:
  - Occur nightly
  - Causing safety concerns
  - Persist over long periods
Common Sleep Problems in Developmentally Delayed Individuals

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Sleep Related Movement Disorders

- Restless Leg Syndrome
  
  **U**- Urge to move limbs
  **R**- Rest makes it worse
  **G**- Getting up/moving helps
  **E**- Evening or night
  **S**- Symptoms not due to other cause
Sleep Related Movement Disorders

• Restless leg syndrome
  • Bothersome while awake
  • Associated with iron deficiency
    • Ferritin < 50
  • Familial
  • Overlap with Periodic Limb Movements Disorder
    • 80% of those with RLS have PLMD
Sleep Related Movement Disorders

• Periodic Limb Movement Disorder
  • During Sleep
  • Movements interrupt/worsen sleep
  • Overlap with RLS
    • 20% of those with PLMD have RLS
Sleep Related Movement Disorders

- Sleep Related Rhythmic Movement Disorder
  - Repetitive, stereotyped and rhythmic motor behaviors
    - Body rocking
    - Head banging
    - Leg banging
  - Occurs when drowsy or asleep

- A “problem” when:
  - Interferes with sleep
  - Daytime impairment
  - Risk of Self Injury
Sleep Related Movement Disorder Summary

- RLS can mimic insomnia
  - Check ferritin level is > 50
- Periodic Limb Movements may not be a disorder:
  - if not interrupting sleep
  - not causing daytime dysfunction
- Rhythmic Movements can be a coping tool
- Dream enactment may be a harbinger of a neurodegenerative illness

- Refer if:
  - Daytime functional impairment
  - Safety concerns
Developmentally Delayed Populations at High Risk for Sleep Disorders

- Down Syndrome
  - Obstructive Sleep Apnea - incidence of 50-100%
  - Insomnia
- Prader Willi
  - Obstructive Sleep Apnea
  - Central Hypersomnia
  - Nocturnal Eating
- Myotonic Dystrophy
  - Obstructive Sleep Apnea
  - Central Sleep Apnea
  - Central Hypersomnia
- Smith Magenis Syndrome
  - Circadian Rhythm Disorders
  - Insomnia
  - Obstructive Sleep Apnea
Developmentally Delayed Populations at High Risk for Sleep Disorders

- Syndromes with Dysmorphic Faces
  - Obstructive Sleep Apnea
- Autism Spectrum
  - Insomnia
  - Sleep related rhythmic movement disorders
- Individuals with Blindness
  - Circadian Rhythm Disorders
- Syndromes with Epilepsy
  - Obstructive sleep apnea
  - Hypersomnia
  - Parasomnias
Case: Mr. Willi

Mr. Prader-Willi is a 39 year old morbidly obese male who reports that he wakes every morning with a headache and feels tired during the day.

He sleeps an average of 12 hours a night.

He often wakes up hungry in the middle of the night and sneaks to the kitchen to get a snack, while other times he wakes up with crumbs in his bed though does not remember getting up to eat.

His roommate complains that he is “noisy” at night, which makes Mr. Willi feel persecuted.
Case: Mr. Willi:
Key Points

- Some developmental disorders, like Prader-Willi, have known hypersomnia
- Obesity is a risk factor for OSA
- Waking unrefreshed after sufficient sleep and taking day time naps is a major red flag for OSA
- OSA is a risk factor for parasomnias due to interruptions in sleep
- OSA contributes to problems with glucose regulation and impulse control
- Snoring may bother others more than the patient
- Obesity increases the likelihood of obesity hypoventilation
  - Morning headache due to accumulation of CO2 due to insufficient expiration/hypoventilation
  - Resolves when CO2 is “blown off” with normal respirations when awake
Case: Ms. Nellie

Mrs. Nellie is a 57 year old female with autism spectrum disorder who has always had insomnia and been a nervous person, but over the last several years has been having even more difficulty falling asleep and has been waking up in the middle of the night “in a panic”.

She sometimes wakes and can’t move her body for several minutes, which is very frightening to her. Now she fears going to sleep.

She returns for follow up after starting lorazepam 1mg at bedtime noting that she is falling asleep faster, but still wakes with anxiety.

She has been having memory problems.

She wakes to urinate at 3am and can’t return to sleep.

Sometimes rocking her body helps her go to sleep.
Case: Ms. Nellie
Key Points

- Rates of sleep apnea in women increase after menopause
- Repeated episodes of hypoxemia and increased sympathetic response can contribute to anxiety
- Sedating medications decrease muscle tone in airway and can worsen OSA
- Repeated hypoxemia and sleep fragmentation contributes to short term memory problems and word finding difficulties
- OSA is typically worse during REM sleep
- OSA increases/causes nocturia
- Rhythmic movements of sleep can be a self soothing tool for some
  - Warrants treatment/is a disorder if causing harm or poor sleep
Case: Ms. Oxy

Ms. Oxy is a 28 year old female treated with opioid pain medications for a Chiari malformation that causes intense headaches.

She has had a few “scary” episodes where her roommate wakes and thinks she is dead because she can’t see Ms. Oxy breathing.

Ms. Oxy notes sometimes waking with a racing heart. She requests something to help with her anxiety at night.
Case: Ms. Roxy

Key Points

• Brain injuries or malformations can affect the sleep wake circuitry and breathing centers in the brainstem
• Opioid medications decrease respiratory drive during sleep
  • Brain forgets to send a signal to lungs to breathe
• Decreased capacity to arouse when hypoxemic
• Concurrent use of benzodiazepines and narcotic medications are a “double whammy”
  • Increased OSA risk from loss of muscle tone
  • Central sleep apnea from blocking mu receptors
Mr. Stumper is a 43 year old male with trisomy 21, hypertension and diabetes, presenting for follow up.

He recently started a third antihypertensive agent and increased his long acting insulin.

His blood pressure and diabetes are still poorly controlled.

He has had difficulties with medication compliance in the past, and you suspect that he is not taking his medications as directed.

When asked about this, he becomes very upset and leaves the office, tearful when noting that he is always the bad guy.
Case: Mr. Stumper

Key Points

- OSA increases risk of hypertension four fold
  - OSA prevents nocturnal dipping
    - Blood pressure usually drops 10-20% during sleep
- OSA causes insulin resistance due to stress response
  - Also true in non-diabetics

Harding SM, J Clin Sleep Med, 2014
Case: Annie Antsy

Annie Antsy is a non-verbal 13 year old female who has recently been having agitation at bed time. She recently started menstruating. She has always been a “picky” eater. She was started on a mirtazapine to help her sleep, which seemed to make things even worse.
Case: Annie Antsy

Key Points

- Restless legs can manifest as behavioral issues, especially in those not able to communicate.
- Iron deficiency is a cause of restless legs.
- Medications, especially antidepressants can cause restless legs.
  - 30% of people on mirtazapine get RLS symptoms.
What questions do you have?

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References: