PARASOMNIAS REVISITED

NEW MEXICO THORACIC SOCIETY

SAPNA BHATIA MD

02/25/17
OBJECTIVES

• Appreciate the clinical semiology to help differentiate between REM and NREM parasomnias
• Appreciate the ICSD III Classification Scheme for the major parasomnias
• Understand management modalities including behavioral and pharmacological for NREM and REM parasomnias
• Understand the difference between parasomnias and seizures
WHAT ARE PARASOMNIAS?

Undesirable motor, or verbal phenomena that arise from sleep or sleep-wake transition
PARASOMNIAS: OVERLAPPING STATES

REM PARASOMNIAS

WAKE

REM

NREM

NREM PARASOMNIAS
PARASOMNIAS: DIFFERENTIAL DIAGNOSIS

REM

RBD
Recurrent Isolated Sleep Paralysis
Nightmare Disorder

NREM

Confusional Arousals
Sleepwalking
Sleep Terrors
Sleep Related Eating Disorder

SEIZURES

NFLE

PSYCHOGENIC SPELLS

Dissociative Disorder
<table>
<thead>
<tr>
<th>ICSD II</th>
<th>ICSD III</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>REM Parasomnias</strong></td>
<td></td>
</tr>
<tr>
<td>• RBD</td>
<td>• RBD</td>
</tr>
<tr>
<td>• Recurrent Isolated Sleep Paralysis</td>
<td>• Recurrent Isolated Sleep Paralysis</td>
</tr>
<tr>
<td>• Nightmare Disorder</td>
<td>• Nightmare Disorder</td>
</tr>
<tr>
<td><strong>Disorders of arousal (NREM sleep)</strong></td>
<td></td>
</tr>
<tr>
<td>• Confusional Arousals</td>
<td>• Confusional Arousals</td>
</tr>
<tr>
<td>• Sleepwalking</td>
<td>• Sleep Walking</td>
</tr>
<tr>
<td>• Sleep Terrors</td>
<td>• Sleep Terrors</td>
</tr>
<tr>
<td><strong>Other Parasomnias</strong></td>
<td></td>
</tr>
<tr>
<td>• SRED</td>
<td>• SRED SDB</td>
</tr>
<tr>
<td>• Sleep Related Dissociative Disorders</td>
<td>• Sleep Related Dissociative Disorders</td>
</tr>
<tr>
<td>• Sleep Enuresis</td>
<td>• Sleep Enuresis</td>
</tr>
<tr>
<td>• Catathrenia</td>
<td>• Catathrenia</td>
</tr>
<tr>
<td>• Exploding Head syndrome</td>
<td>• Exploding Head syndrome</td>
</tr>
<tr>
<td>• Sleep Related Hallucinations</td>
<td>• Sleep Related Hallucinations</td>
</tr>
<tr>
<td>• Parasomnia, Unspecified</td>
<td>• Parasomnia, Unspecified</td>
</tr>
<tr>
<td>• Parasomnia due to Drug or Substance</td>
<td>• Parasomnia due to Drug or Substance</td>
</tr>
<tr>
<td>• Parasomnia Due to Medical Condition</td>
<td>• Parasomnia Due to Medical Condition</td>
</tr>
<tr>
<td><strong>Isolated Symptoms Normal Variants</strong></td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Sleep Talking</td>
</tr>
</tbody>
</table>
# ICSD III CLASSIFICATION

## REM PARASOMNIAS
- RBD
- Recurrent Isolated Sleep Paralysis
- Nightmare Disorder

## DISORDERS OF AROUSAL (NREM PARASOMNIAS)
- Confusional Arousals
- Sleepwalking
- Sleep Terrors
- SRED

## OTHER PARASOMNIAS
- Sleep Related Dissociative Disorders
- Sleep Enuresis
- Exploding Head Syndrome
- Sleep Related Hallucinations
- Parasomnia, Unspecified
- Parasomnia due to Drug or Substance
- Parasomnia due to Medical Condition

## ISOLATED SYMPTOMS & NORMAL VARIANTS
- Sleep Talking
CATCH ME IF YOU CAN
• Philip is a 62 yo R handed man who was referred to the sleep clinic for evaluation of confusion and violent behaviors during sleep.

• 7 years prior, his wife first noticed episodes of talking and groaning in his sleep, sometimes accompanied by unusual body movements.

• Over the past 1 year, his night behaviors became more dramatic with increasing frequency of 2-3 x per week. These episodes typically occurred 2 hours after he fell asleep.

• His wife recalled the first violent episode, when she woke up to find him sitting at the edge of the bed facing the bedside table with the lamp on the floor and blood dripping from his L eyelid. He was shouting and sweating profusely. Once she was able to calm him down, he told her that he was dreaming a man was chasing him while he was jogging at a park. He decided to hide behind a tree and planned to jump his aggressor as he passed in an effort to catch him.
• He and his wife denied urine or stool incontinence, and convulsive movements during episodes.

• No history of cataplexy, hypnogogic hallucinations, sleep paralysis, snoring, choking at night, or symptoms of restless legs syndrome.

• One year before presentation, Philip began to experience episodes of confusion during the day, and his wife noticed trouble with his memory.

• Once, while driving home alone from the grocery store, he forgot how to get back home and could not recall his address when he waved down a police officer for assistance.

• Evaluation with his PCP at that time included a normal brain MRI, daytime EEG, TSH, RPR, vitamin B12 and CBC. Ultimately it was determined that Philip had depression and he was placed on venlafaxine.

• Philip maintained a regular sleep scheduled during weekdays and weekends. He went to bed by 10 pm, fell asleep in 5 mins, and usually woke by 7 am. He often woke feeling unrefreshed, though he did not take naps during the day. Epworth Sleepiness Scale was was 5 (normal), and Patient Health Questionnaire-9, and index of depression, was normal at 3.
DIFFERENTIAL DIAGNOSIS?

• Nocturnal Seizures?
• Nightmare Disorder?
• REM Behavior Disorder?

AN IN-LAB DIAGNOSTIC NOCTURNAL POLYSOMNOGRAM WAS THEN PERFORMED.
REM Sleep Behavior Disorder
Lewy Body Disease
REM BEHAVIOUR DISORDER (RBD)

• A REM parasomnia that usually emerges later in life, typically after the age of 50

• The presenting complaint in RBD is recurrent dream-enacting behaviors that have the potential to result in injury to the affected individual or the bed partner

ICSD III DIAGNOSTIC CRITERIA FOR RBD

• **PSG abnormality** – Elevated EMG tone during REM sleep in either submental or limb leads

• Either a **history** of dream enactment behavior or **observation** of abnormal REM sleep during the PSG

• Absence of EEG epileptiform activity during REM sleep

• The disturbance is not explained by another sleep/medical/neurological/mental disorder, and is not related to medication/substance use
THE SEMIOLOGY OF RBD

- REM SLEEP
- SUDDEN AROUSAL
- POSSIBLE INJURY
- COMPLEX BEHAVIOR
- RECALL
REM SLEEP BEHAVIOR DISORDER

• RBD associated with neurodegenerative disease, particularly \( \alpha \)-Synucleinopathies
  • Parkinson’s Disease
  • Dementia with Lewy Bodies
  • Multiple System Atrophy

Positive \( \alpha \)-Synuclein staining of a Lewy body in a patient with Parkinson’s disease
Stage 1 & 2
Dorsal motor, olfactory, RBD

Stage 3
Motor Symptoms

Stage 4-6
Cortical Involvement (dementia)

Spread of α-Synuclein in the Brain

Via olfactory bulb

Premotor symptoms

Via vagus nerve

Motor symptoms

Brainstem Lewy body

Cortical Lewy body

Disturbances in sleep should be seen before motor symptoms

### ACUTE RBD

<table>
<thead>
<tr>
<th>DRUGS</th>
<th>MEDICAL AND NEUROLOGIC DISORDERS</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Alcohol intoxication, psychoactive drugs, withdrawal states: intense REM rebound states&lt;br&gt;• Medication intoxication (e.g. anticholinergics)&lt;br&gt;• Therapeutic pharmacotherapy: SSRIs, TCAs, MAOIs, venlafaxine</td>
<td>• Relapsing Multiple Sclerosis&lt;br&gt;• Vascular&lt;br&gt;• Toxic/Metabolic&lt;br&gt;• Brainstem Tumors&lt;br&gt;• Infectious, post-infectious&lt;br&gt;• Paraneoplastic&lt;br&gt;• Traumatic</td>
</tr>
</tbody>
</table>
PATHOPHYSIOLOGY OF RBD

Pathophysiology of REM sleep behavior disorder

Lesion

- Pons Medulla
- Lack of Excitation
- Medullary Inhibitory Area

Excite

Inhibit

Motor Neurons

Lack of Inhibition

Motor Neurons

Medullary Inhibitory Area

Lesion

Pons Medulla

Muscles

Net effect

Muscle Atonia

Net effect

Lack of Muscle Atonia

- Via magnocellularis neurons
- Via the lateral tegmentoreticular tract
WHY ORDER SLEEP STUDIES IN RBD?

• (A) RBD is the only parasomnia requiring PSG confirmation

• (B) Readily available assay for confirmation of RBD and exclusion of RBD mimics

• (C) RBD heralds the future diagnosis of α-synucleinopathies: Dx is crucial in the neurologic follow-up of these patients

• (D) Dream-enacting behaviors are not unique to RBD- may be seen in OSA, NREM parasomnias, and nocturnal seizures
<table>
<thead>
<tr>
<th></th>
<th>RBD</th>
<th>Nocturnal Seizures</th>
<th>Nightmare Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Time</strong></td>
<td>Late</td>
<td>Any</td>
<td>Late</td>
</tr>
<tr>
<td><strong>Sleep Stage</strong></td>
<td>REM</td>
<td>NREM&gt;&gt;REM</td>
<td>REM</td>
</tr>
<tr>
<td><strong>EEG discharges</strong></td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td><strong>Behavior</strong></td>
<td>++++</td>
<td>++++</td>
<td>+</td>
</tr>
<tr>
<td></td>
<td>Not stereotyped</td>
<td>Stereotyped,</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>monomorphic</td>
<td></td>
</tr>
<tr>
<td><strong>Duration</strong></td>
<td>1-10 min</td>
<td>5-15 min</td>
<td>3-20 min</td>
</tr>
<tr>
<td><strong>Post event confusion</strong></td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td><strong>Age</strong></td>
<td>Older adult</td>
<td>Young adult</td>
<td>Child/Young adult</td>
</tr>
</tbody>
</table>
TREATMENT SUGGESTIONS FOR RBD

• Confirmation

• Formal Neurologic Exam

• Safety is the first step before any pharmacologic intervention

• LEVEL A: Safety intervention
  • Clonazepam – 92% effective, 0.25-0.5 mg
    • Use with caution in patients with dementia, gait disorders, or concomitant OSA. S/E include sedation, impotence, motor incoordination, confusion and memory dysfunction
  • LEVEL B: Melatonin – 87% effective, 9-12 mg
    • Effective in patients with α-synucleinopathies, memory problems, and sleep disordered breathing. S/E include headaches, sleepiness, delusions/hallucinations

• Avoid aggravating Rx: SSRIs, venlafaxine

RBD – SAFETY: LEVEL A EVIDENCE

• Modifying sleep environment:
  • Bedroom Safe
  • Remove hard/safe objects
  • Sleep in padded mattress
  • Place a mattress on floor
  • Cover windows with heavy curtain
  • Use pillow barricades

• Until managed, sleep alone
• Sleep in sleeping bag until treated

YOUNG AND AFRAID
• A mother brings her 6 yo daughter, Jamie, to the sleep clinic for evaluation of loud screaming episodes at night.

• Starting at the age of 3, Jamie would wake up and scream at night. At first, these episodes occurred a few times per week, but then her bouts became more frequent.

• The timing of these episodes were fairly consistent. Almost every night around 9pm, Jamie screamed out and sat up in bed crying. She looked frightened, as if she had just seen a ghost.

• Her eyes were partially open, her breathing was labored, and she was covered with sweat. Sometimes she moved aimlessly around the bed and tugged on her blankets.

• She appeared to be confused and did not recognize her mother when she tried to comfort her. Her crying gradually subsided and within 30 mins, Jamie returned back to sleep. In the mornings, Jamie would not recall these episodes.
• Jamie’s sleep history revealed that she went to bed at 7 pm and woke up at 7 am for school on both weekdays and weekends.

• Once asleep, she did not exhibit any snoring, leg jerking, tongue biting or bed-wetting behavior.

• Jamie’s medical history was unremarkable. She was delivered without complications at 40 weeks via C-section, weight 9 lbs, 7 oz. Her physical development has been normal, and there had been no reports of behavioral or academic problems at school.

• Physical exam was normal. Specifically, her cranio-facial morphology was unremarkable, without evidence of retrognathia or nasal abnormality.

• Her tonsils were small without erythema or exudates, and her tongue was proportional to her mouth size.
DIFFERENTIAL DIAGNOSIS?

- Sleep Terrors?
- Confusional Arousal?
- Nightmare Disorder?

AN IN-LAB DIAGNOSTIC NOCTURNAL POLYSOMNOGRAM WAS THEN PERFORMED
Sleep Terrors
SLEEP TERRORS

• AKA parvor nocturnus

• The most prominent characteristic of sleep terrors is a sudden arousal from sleep that is associated with an intense yell or cry occurring in the first part of the night, typically during SWS (NREM)

• Activation of the sympathetic nervous system manifesting as tachycardia, tachypnea, diaphoresis, and mydriasis are common

• Other characteristics include confusion through the entire episode, inconsolability, and poor recollection of the event the morning after

• Sleep terrors typically peak in early elementary school ages and resolve by middle school age

ICSD III DIAGNOSTIC CRITERIA FOR SLEEP TERRORS

• This criteria meets general criteria for NREM disorders of arousal
  1) Repeated episodes of partial or incomplete awakening from NREM sleep
  2) Patient responds inappropriately, or does not respond to efforts of observers to intervene or redirect him/her during the episode
  3) Little or absent cognition or dream imagery following the event
  4) Fragmented or no recollection for the episode
  5) Absence of other primary sleep disorder, mental disorder, medical condition, or substance use to help explain the disturbance

• The arousals are characterized by episodes of abrupt terror, typically beginning with an alarming vocalization such as a frightening scream

• There is intense fear and signs of autonomic arousal, including mydriasis, tachycardia, tachypnea, and diaphoresis during and episode
THE SEMIOLOGY OF SLEEP TERRORS

SWS
- Sudden Arousal
- Tremendous autonomic discharges
- Confusion/Disorientation
- No response to parents
- Mumbled speech, confusion
- Attempts to interrupt heighten confusion
- Amnesia for the event
- Return to sleep and baseline
## DIFFERENTIAL DIAGNOSIS

<table>
<thead>
<tr>
<th></th>
<th>Sleep Terrors</th>
<th>Confusional Arousals</th>
<th>Nightmare Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Time</strong></td>
<td>Early</td>
<td>Early</td>
<td>Late</td>
</tr>
<tr>
<td><strong>Sleep Stage</strong></td>
<td>NREM (SWS)</td>
<td>NREM (SWS)</td>
<td>REM</td>
</tr>
<tr>
<td><strong>EEG discharges</strong></td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td><strong>Scream</strong></td>
<td>+++</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td><strong>CNS activation</strong></td>
<td>+++</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td><strong>Post event confusion</strong></td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td><strong>Post event awakening</strong></td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td><strong>Duration</strong></td>
<td>1-10 min</td>
<td>0.5-10 mins</td>
<td>3-20 min</td>
</tr>
<tr>
<td><strong>Age</strong></td>
<td>Child</td>
<td>Child</td>
<td>Child/Young Adult</td>
</tr>
<tr>
<td><strong>Genetics</strong></td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>
SLEEP TERRORS: MANAGEMENT

• The diagnosis of sleep terrors is often distressing to the entire family, but it is mostly benign and does not warrant aggressive pharmaceutical intervention

• It does not help to try and to intervene by awakening the child during the episode

• The mainstays of therapy for sleep terrors include reassurance, adequate sleep time, regular sleep schedule, treatment of secondary triggers like OSA, safety precautions

SLEEP TERRORS: MANAGEMENT

• If all episodes are predictable, a trial of scheduled awakenings
• This involves disturbing the child’s sleep just enough to cause stirring and arousal without total awakening
• The parent is asked to do this several minutes prior to the predicted event
• Particularly resistant cases can be tried on benzodiazepines or tricyclic antidepressants

• Jill is a 31 year old woman who comes to the sleep medicine clinic with complaints of binge eating at night for nearly 10 years.

• These episodes initially occurred in times of extreme stress but they increased dramatically over the past year when she was promoted at work to host a morning radio talk show.

• She made excursions to the kitchen 3 to 4 times per week, often multiple times in the night, typically during the first half of the sleep period.

• She usually ate high-carbohydrate foods, such as chocolates, cakes, cereal, pastries, muffins, and cookies.

• One morning, she found a half eaten dish of lasagna in the microwave. In her refrigerator, she found a pie pan with only crumbs and a dirty spoon.

• On awakening, she would find crumbs and chocolate smears on her fingers and face, on the bed, or on the doorknob; frosting and coffee grounds in her hair; and peanut butter in the pockets of her robe.

• She had no recall of her nighttime foraging; she only saw the disturbing evidence left behind and an annoyingly increase in numbers on her weight scale.

• Curiously, during the daytime, she had no appetite.
• In terms of Jill’s sleep history, she typically went to bed at 11 pm. It took 30 mins to fall asleep and, due to her new job, woke up between 4:30 and 5 am feeling unrefreshed.

• In addition, her PCP had started a sleeping pill approximately a year ago as Jill complained of insomnia when her new job started.

• Jill’s history revealed that she had a history of sleepwalking and nightmares in childhood, both of which remitted in her teen years. Her father was a sleepwalker in childhood as well.

• She denied symptoms of RLS, narcolepsy, and psychiatric and eating disorders. Her ESS was notable for a score of 11/24 indicating mild sleepiness.

• Physical exam was unremarkable except for a BMI of 30.
DIFFERENTIAL DIAGNOSIS?

• Kleine-Levin Syndrome?
• Sleep Related Eating Disorder?
• Nocturnal Eating Disorder?

AN IN-LAB DIAGNOSTIC NOCTURNAL POLYSOMNOGRAM WAS THEN PERFORMED
Sleep Related Eating Disorder
SLEEP RELATED EATING DISORDER (SRED)

- SRED is characterized by compulsive involuntary nocturnal eating
- Consumption of inappropriate food items or food items that are rich in carbohydrates
- Incidence < 1%
- Female predominant, mean onset 3rd decade of life
- SRED patients have an increased risk of injuries, such as lacerations from food preparation, choking on thick foods, drinking excessively hot liquids, ingesting poisonous substances, and consuming foods to which they are allergic

SLEEP RELATED EATING DISORDER (SRED)

• Idiopathic, drug-induced or linked to other sleep disorders
• OSA could trigger partial awakenings during NREM periods, and phenomena like SRED may ensue
• A history of sleepwalking is most common
• Medication-induced SRED with zolpidem has been well described

ICSD III DIAGNOSTIC CRITERIA FOR SRED

• Repeated abnormal eating episodes following an arousal during the main sleep period

• The presence of at least one of the following in conjunction with recurrent episodes of involuntary eating:
  1) **Consumption of bizarre and unusual forms or mix of food or inedible or noxious substances**
  2) **Potentially harmful and injurious sleep behaviors performed while searching for food or while preparing/cooking food**
  3) **Deleterious and adverse health consequences from recurrent nocturnal eating**

• During the eating episode, patients experience incomplete or complete loss of conscious awareness, with subsequent impaired recall

• This disorder is not better accounted for or explained by another sleep disorder, medical disorder, mental disorder, substance use, or medication
THE SEMIOLOGY OF SRED

SWS

Return to sleep and baseline

Amnesia for the event

Mumbled speech, disorientation, confusion

Sudden Arousal

Ambulation/Displacement

OSA

SRED
<table>
<thead>
<tr>
<th></th>
<th>SRED</th>
<th>NES</th>
<th>KLS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nocturnal eating</td>
<td>Asleep</td>
<td>Asleep- &gt; Awake</td>
<td>Awake</td>
</tr>
<tr>
<td>Gender and age</td>
<td>F</td>
<td>F</td>
<td>M</td>
</tr>
<tr>
<td>Associated disorders</td>
<td>Sleepwalking, RLS, PLMS, OSA</td>
<td>Depression, Anxiety</td>
<td>Disinhibited Behaviors (Hypersexuality)</td>
</tr>
<tr>
<td>Daytime sleepiness</td>
<td>-</td>
<td>-</td>
<td>++++</td>
</tr>
<tr>
<td>PSG abnormalities</td>
<td>Arousal from SWS</td>
<td>Decreased sleep efficiency, Decreased TST</td>
<td>Increased TST</td>
</tr>
<tr>
<td>Amnesia</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Food consumed</td>
<td>Inedible substances</td>
<td>High carbohydrate</td>
<td>High carbohydrate</td>
</tr>
<tr>
<td>Spontaneous remission</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
</tbody>
</table>
**SRED: MANAGEMENT**

- Environmental Safety
- Topiramate: ? Mechanism of action
- Dopaminergic agonists: decrease motor activity during sleep, thus decreasing arousals


TAKE HOME POINTS

• Detailed history is key in the diagnosis of parasomnias.

• REM sleep behavior disorder is the only parasomnia requiring PSG confirmation.

• A diagnosis of RBD should prompt an evaluation for α-synucleinopathies.

• Sleep related eating disorder may be considered a continuum of sleepwalking. Evaluation of factors known to precipitate sleepwalking is important in the evaluation and treatment of SRED.

• Sleep terrors can seem terrifying, but often self-resolve without need for pharmacologic intervention.
QUESTIONS?

THANK YOU FOR YOUR ATTENTION