Sleep Abnormalities Associated with Behavioral Problems in Prader-Willi Syndrome

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Disclosure

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- FPWR grants- Guanfacine XR; Bright Light Therapy
- Book: Neurobehavioral Manifestations of PWS
Objectives

• Introduction
• Common sleep abnormalities in PWS
• Common behavioral abnormalities in PWS
• Determining the cause of sleep disturbance in PWS
• The role of sleep hygiene
• The role of psychopharmacotherapy
• Q/A
Introduction

• Sleep disorders are ubiquitous in PWS (today’s presentation)
• Genetics and hypothalamic dysfunction are biological causes (today’s presentation)
• Pulmonologists and Sleep Specialists should be core team members
• Important to differentiate chronic sleep abnormalities from acute changes
Problems with Sleep Initiation

- Onset - is it new or a chronic issue
- Napping during the day
- Change in daytime physical activity e.g. during COVID-19
- Associated behaviors
  - Excessive worrying (anxiousness)
  - Separation anxiety - any change at home?
  - Compulsive bedtime behaviors
Excessive Daytime Sleepiness

- Differentiate from sleep phase disturbances
  - Advanced- e.g. elderly
  - Delayed- e.g. adolescents
- Narcolepsy- very common
- Sleep Apnea
  - Central
  - Obstructive

Masquerades as depression, mood disorder, ADHD, even psychosis
Excessive Daytime Sleepiness (Epworth)

NODDING OFF:
• Sitting and reading
• Watching TV
• Sitting inactive in a public place (e.g., a theater or a meeting)
• As a passenger in a car for an hour without a break
• Lying down to rest in the afternoon when circumstances permit
• Sitting and talking to someone
• Sitting quietly after a meal
• In a car, while stopped for a few minutes in traffic
Inattention and sleep

- ADHD symptoms are common in PWS
- Hypothalamus and in-turn \textit{reticular activating system} abnormalities directly lead to inattention
- Hyperactivity is noted infrequently as compared to inattention
- Untreated sleep apnea
- Excessive daytime sleepiness
- Aggression- sometimes associated with “fighting the sleepiness”
- Role of stimulants
- Non-stimulants
- Therapeutic role of daytime naps
Mood Disorders

Sleep in Depression
  • Increased Latency
  • Early morning awakening
  • SIGECAPS

Sleep in Mania/Psychosis
  • Reduced need for sleep...sudden resolution of hyperphagia
  • Cycloid Psychosis

Cataplexy vs. Catalepsy- associated hallucinations differ (hypnogogic/pompic)

Decreased need for sleep is unusual and should be taken seriously if sudden
Medication-induced sleep problems

• Timing of dosage matters with many medications

• Sleepiness:
  • Benzodiazepines
  • Anticholinergics
  • Some antidepressants
  • Antipsychotics- except with Akathisia
  • Alpha-2 Agonists

• Insomnia:
  • Stimulants
  • Modafinil/Armodafinil
  • Theophylline
  • Bupropion
  • Most antidepressants

MAY BE USED AS TREATMENT OF EDS
Important for both patients and caregivers (YOUR SLEEP IS IMPORTANT TOO!!)

Chronic sleep deprivation can affect physical and mental health

Have a specific bedtime and wake-up time that does not vary much.

Create a ritual that signals to the body that it’s time to sleep

Refraining from caffeinated beverages (coffee, black tea, caffeinated sodas) after the late afternoon

Avoid high-intensity exercise within three hours of bedtime

Turn off electronics at least 30 minutes before bedtime
• Bright light therapy: 9000 lux or more for 30 minutes
• Well established treatment for SAD (seasonal affective d/o)
• Also studied in patients with TBI and Parkinson’s
• Well tolerated, may help with weight and cognition
• 30 minutes twice a day
• Home based within natural settings
• Maimonides Research Enrolling NOW- Call (718) 283-8170
THANK YOU

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Book now available: Thank you Ms. Rivard and PWSA!