

Sleep Abnormalities Associated with Behavioral Problems in Prader-Willi Syndrome

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Disclosure

- Consultancy: Soleno, Radius, Levo
- 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 **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

Sleep Disturbance as a Sign of Psychiatric Illness

- 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

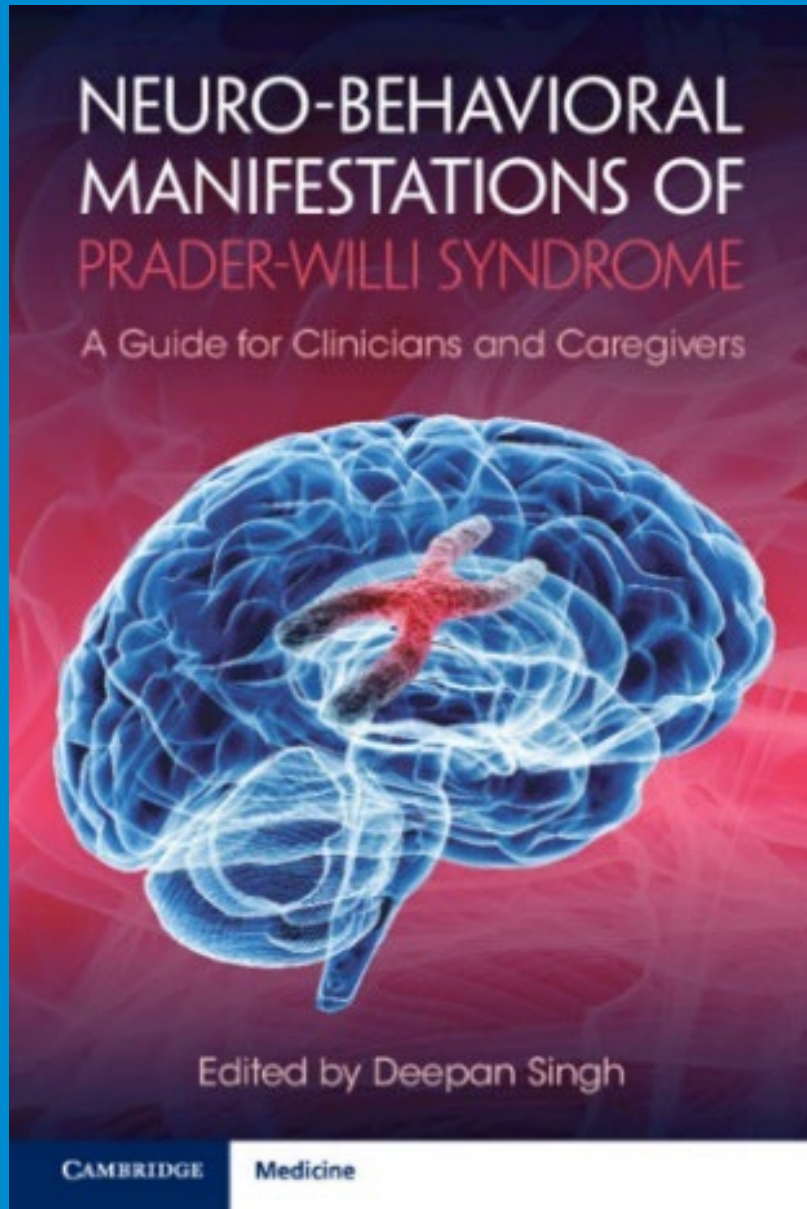
Sleep Hygiene

- 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- An Unexplored Treatment

- 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

Maimonides Research:

Bright Light Therapy

Radius

Guanfacine XR

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