

What is Prader-Willi Syndrome?

Prader-Willi Syndrome (PWS) is a genetic neurodevelopmental disorder characterized by:

- neonatal hypotonia (low muscle-tone),
- childhood-onset obesity,
- hyperphagia (excessive hunger),
- hypogonadism (defective reproductive system),
- developmental delay/mental retardation,
- behavioral difficulties (in particular food related),
- short stature due to growth hormone deficiency,
- characteristic dysmorphic facial features.

Compulsive behaviors in PWS patients include preoccupation with food or food seeking.

The prevalence of PWS is estimated at between 1 in 10000 to 1 in 20000 children.

How are Sleep Disorder and PWS Linked to Each Other?

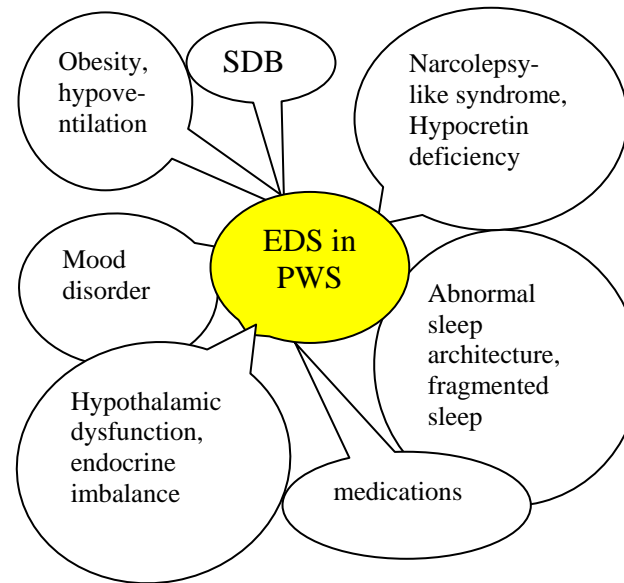
The most common Sleep Disorders associated with ADHD include:

- **Sleep Disordered Breathing** (SDB, e.g. sleep apnea)

- Narcolepsy-like symptoms
- Abnormal sleep architecture
- Fragmented sleep



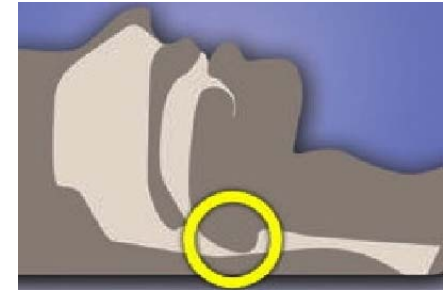
- **Excessive Daytime Sleepiness (EDS)**—EDS is a well-known feature of PWS. The following factors contribute to EDS in patients with PWS:



PWS patients exhibit significant behavioural disturbances, temper tantrums, lack of certain social skills, and depressive symptoms

Sleep Disorder Breathing in Patients with Prader-Willi Syndrome

Sleep Breathing Disorder (SBD) refers to a sleep disorder in which a person stops breathing repeatedly during sleep.



SBD is prevalent in patients with Prader-Willi Syndrome.

Risk factors for SBD include the following:

- obesity
- hypotonia
- adenotonsillar hypertrophy
- sticky secretion
- hypothalamic dysfunction
- restrictive lung disease
- scoliosis
- respiratory muscle weakness
- facial dysmorphism

Sleep Apnea in PWS and anesthesia – use of pre-and postoperative sedation could increase the risk of sleep-related apneic episodes. There is the need for well-planned anesthesia care for PWS patients. Polysomnography (sleep study), early diagnosis and treatment of upper airway obstruction are recommended.

How to Treat SDB in Patients with PWS?

Adenotonsillectomy: is a first choice of treatment of sleep apnea in children. Certain patients are at higher risk for postoperative complications. This includes:

- Age below 3 years, failure to thrive, obesity, cardiac complications, craniofacial anomalies, neuromuscular disease, OSA and sleep O₂ saturations <80%
- Preoperative sleep studies and postoperative overnight observation are recommended

Continuous Positive Airway Pressure (CPAP): allows for normalization of breathing pattern, improvement of nocturnal sleep, elimination of daytime somnolence, improved mental acuity, normalization of daytime blood gases
* However there is limitation of CPAP use in PWS patient due to behavioral and learning difficulties

Growth hormone commonly used in PWS children might increase the risk of Sleep Apnea and could lead to fatalities among these children. It is recommended that patients undergoing growth hormone therapy have a sleep study done at baseline and one done 6 weeks after the therapy has initiated.

For more copies of this pamphlet please visit:
www.youthdale.ca

REFERRAL FORM



Youthdale Child and Adolescent Sleep Centre
227 Victoria Street, Lower Level 2
Toronto, Ontario, M5B 1T8
Phone: (416) 703-0505 Fax: (416) 703-0507

Patient Information:

Name: _____
DOB: _____
Contact Phone #: _____
Age: _____ Male Female
Height: _____ Weight: _____

Referring Dentist / Doctor:

Name: _____
Address: _____
Phone #: _____
Fax #: _____

Reason for Referral: (Please Circle All Relevant)

Anatomical:

- Large tonsils
- Large adenoids

Nighttime Complaints:

- Insomnia
- Snoring, Breathing problems
- Sleep apnea
- Other: _____

Daytime Complaints:

- Difficulty waking up
- Excessive sleepiness
- Tiredness
- Irritability
- Hyperactivity
- Behavioral problems in school
- Other: _____

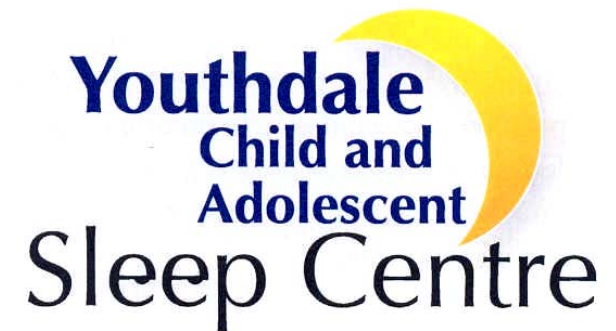
History and Medical Information:

Referring Dentist / Doctor Signature:

_____ Date: _____



PRADER-WILLI SYNDROME AND SLEEP DISORDERS



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