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)**

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.
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.

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