Sleep Disordered Breathing in Peudek Wille Syndrome

Outline

Pathophysiology of Sleep Disorders

Abnormal Sleep

- Adults and children present to the sleep clinic with 3 main symptoms:
  - Trouble falling asleep or staying asleep (insomnia)
  - Excessive daytime sleepiness
  - Abnormal events or episodes during sleep (sleepwalking, sleep talking, leg kicks, etc.)

The Most Common Sleep Disorders

- Insomnia
- Obstructive Sleep Apnea (OSA)
- Narcolepsy
- Restless Legs Syndrome (RLS)
- Narcolepsy

Pediatric Sleep Disorders by Time of Night

- Behavioral Inconsistencies
- Physical Developmental Delays
- Sleep-Related Cognitive Impairment
- Speech and Language Delays
- Increased Stimulation
- Insomnia
**Epidemiology in Typically Developing Children**

- 10% infant mortality
- OSA
  - When defined by pediatricians, the prevalence is 4 to 10%
  - As defined by adults, the overall prevalence is 1 to 4%
  - High prevalence between the ages of 3 and 8 years
  - Rare in infants

**OSA in Prader-Willi Syndrome**

- Systematic review of 14 published studies. 80% of children with Prader-Willi syndrome (PWS) had obstructive sleep apnea syndrome (OSAS)
- Peak incidence of OSA occurs at 3 to 6 years
- Similar to the general population
- Central sleep apnea (CSA) is more common in infants

**OSA in Prader-Willi Syndrome**

- Respiratory failure during pulmonary or systemic illness is the most common cause of death in patients with PWS
- Both CSA and OSA events increase during times of illness in patients with this syndrome
- Sleep apnea is thought to be the leading cause of sudden deaths

**Presentation and Symptoms**

- **Nocturnal Symptoms**
  - Loud, persistent snoring
  - Apnea events
  - Paradoxical movements of chest wall and abdomen during breathing
  - Restless sleep
  - Sweating during sleep
  - Mouth breathing
  - Abnormal sleeping positions

**Presentation and Symptoms**

- **Physical Diverting Symptoms**
  - Mouth breathing at the mouth
  - Chronic nasal congestion
  - Frequent sneezing
  - Morning headaches
  - Fatigue
  - Hypertension
  - Poor appetite

**Presentation and Symptoms**

- **Associated Features**
  - Enuresis (esp. secondary)
  - Increase in peripheral oral paraphilia
  - Central sleep problems
  - RESPI progeny or sleep apnea disorder may be secondary to OSA or causally independent
**Sleep Disordered Breathing in Prader-Willi Syndrome**

**Outline**
- Sleep Disordered Breathing
- Obstructive Sleep Apnea
- Nodoule
- Restless Leg Syndrome

**Pathophysiology of Sleep Disorders**

**Abnormal Sleep**
- Adults and children present at the sleep clinic with 3 main symptoms:
  - Trouble falling asleep or staying asleep/insomnia
  - Excessive daytime sleepiness
- Abnormal movements at night: leg movements equals restlessness (wagging your toes)

**The Most Common Sleep Disorders**
1. Insomnia
2. Obstructive Sleep Apnea (OSA)
3. Narcolepsy
4. Restless Leg Syndrome (RLS)
5. Other/undefined

**Pediatric Sleep Disorders by Time of Night**

**Clock Time**

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**Epidemiology in Typically Developing Children**

- 10% habitually snore
- OSA
  - When defined in uncomplicated sleep studies, the prevalence is 1 to 11%.
  - An asymptomatic overnight sleep study in the overall prevalence is 1 to 5%.
  - High prevalence between the ages of 1 and 8 years.
  - Recent studies.

**OSA in Prader Willi Syndrome**

- Systematic review of 14 published studies: 80% of children with Prader Willi syndrome (PWS) had obstructive sleep apnea syndrome (OSAS).
- Peak incidence of OSA occurs at 3 to 6 years.
- Similar to the general population.
- Central sleep apnea (CSA) is more common in infants.

**OSA in Prader Willi Syndrome**

- Respiratory failure during pulmonary or systemic illness is the most common cause of death in patients with PWS.
- Both CSA and OSA events increase during times of illness in patients with this syndrome.
- Sleep apnea is thought to be the leading cause of these sudden deaths.

**Presentation and Symptoms**

- **Nocturnal Symptoms**
  - Loud, continuous, rough snoring.
  - Apnea pauses.
  - Paradontal disease, ear pain, and children with obstructive sleep.
  - Restless sleep.
  - Snoring during sleep.
  - Mouth breathing.
  - Abnormal sleeping positions.

- **Physical Daytime Symptoms**
  - Mouth breathing.
  - Chronic nasal congestion and tinitus.
  - Hypertension.
  - Morning headache.
  - Frequent infections.
  - Daytime sleepiness.
  - Poor appetite.

**Presentation and Symptoms**

- **Associated Features**
  - Enuresis (eg, primary).
  - Increase in parental attention in young children.
  - Gastroesophageal reflux disease.
  - RLS, PLMD, and obstructive sleep disorders may be secondary to OSA or central sleep apnea.

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Preparing for the Sleep Study

- Set up a practice visit to the sleep center
- "Practice" electrode application at home

Preparing for the Sleep Study: Write a Social Story

A Social Story is an individualized social story which includes real-life visual and written instructions and words that can help a child understand and prepare for the sleep study. The story includes steps such as getting ready, going to bed, and waking up. The child can follow along with the story and practice the steps.

Create a Visual Schedule

Create picture books to help prepare children for the sleep study experience.
- Quiet play activities, stickers, activity books
- Create a visual schedule to help children understand the process.
**PAP Habituation**

- Always a challenge
- 20% drop out rate
- Patient report is subjective and
- Variable adherence (60-70%)
- No difference for fixed vs bilevel PAP

**Hypoglossal Nerve Stimulation (HNS)**

- InSpire Upper Airway Stimulation System (UAS)
  - FDA approved 2014
- Hypoglossal nerve stimulator (HNS)
  - Inspire
  - Synergy

**Upper Airway Stimulation**

- Components
  - Irreversible pulse generator
  - Sensing lead
  - Stimulation lead

**PAP Habituation: It's a Process**

- Establish that only patient breathes the mask
- Place on night or day
- Add humidifier
- Introduce flow
- Increase time with mask in place
- Avoid dehydration
- Reduce nausea

**Human Studies**

- The behavior (HNS) improves in an undisturbed (NO)
- DNA and nerve tissue damage decreases
  with the continued treatment
- Use of the HNS reduces frequency and severity of
  apnea symptoms
- Ambulatory study: 150 STS patients in 300