

### What is it?

Prader-Willi syndrome is a rare genetic disorder present at birth that results in physical, mental and behavioral problems. PWS, a defect on the 15th chromosome, is the most common genetic cause of life threatening obesity. PWS affect's the hypothalamus of the brain, which is part of the endocrine system. The hypothalamus helps to control and regulate bodily functions like hunger and satiety, pain, temperature, fertility and the sleep wake cycle. There are 2 stages associated with this syndrome. In stage 1, an infant displays low tone with a weak cry and poor suck. These infants are often diagnosed with failure to thrive because of their feeding difficulties. As they age, infants often gain strength and muscle tone. Stage 2 begins around age 2-8 years. These children will display the constant desire to eat, never feeling full. These overeating patterns, combined with a low metabolic rate, put a child at risk for morbid obesity.

### What are the common effects?

- Poor muscle tone
- Distinct facial features
- Choking
- Failure to thrive
- Lack of eye coordination
- Food cravings and weight gain
- Underdeveloped sex organs
- Learning disabilities
- Speech problems
- Behavioral problems
- Sleep disorders
- Obesity related complications
  - Type 2 diabetes
  - Heart disease
  - Stroke
  - Sleep apnea
- Higher threshold for pain
- Scratching and skin picking
- Deficient growth hormone
- Low tolerance for external temperature

### Safety at school

#### *Food seeking behavior*

- Create a "food-free" learning environment
- Be mindful of locations of a packed lunch (locker, when on school bus, etc.)
- Be aware of smells (think popcorn!) when planning classroom location
- Don't use food as reward
- Always supervise food areas, children may try to trade food items
- Also supervise with meals if concerns for pace and/or binging
- Discourage and/or report food sharing as calories may need to be adjusted for future intake
- Don't attempt to physically remove, try to negotiate or trade



#### *Poor stamina*

- Mornings typically are best for learning
- Send on errand/assign activity to get up and moving
- High energy activity after lunch
- Rest breaks as needed



## **Behaviors that impact learning**

### *Rigid thinking*

- Need consistency
- Visual schedule for the day, include pictures
- Keep instructions simple (short term memory)
- Use reflection
- Less is best
- Foreshadow change, allow discussion in safe place
- No promises!
- Compromise, within reason (also works as diversion)
- For field trips, show brochures

### *Perseverate or obsessive thinking*

- Use reflection, state back what you just said
- “Strategic timing” before lunch
- Set limits  
Ex. “You can ask 2 more questions.” “You can ask one more question before lunch.” Hold up fingers. Use tally sheet to track.
- Checklist of items so nothing is lost

### *Emotional control*

- Be aware of over-stimulating situations like hallways, transitions, arrivals and dismissals
- Plan for what the child can do if feeling angry or frustrated
- ”Safe zone”
- De-escalation activity
- Be aware of reasoning, set limits
- Set daily goals together
- Communicate behaviors you wish to see
- Create key words to communicate quickly

### *Peer interactions*

- Small groups
- Include them in planning of activities
- Practice social skills like sharing, taking turns
- Role play
- Write down Do’s/Don’ts for social situations

## **SHNIC school nurses information:**

### **Specific health issues for individual health care plans**

- Diagnosis including age and other characteristics the child may display
- Nutrition orders including calories for meals, snack
- Supervision for meal times
- Emergency protocol in event of severe stomach illness (possible binge episode)
- Emergency protocol for vomiting (children with PWS rarely vomit)
- Fever protocol– severe infection may present with low grade fever
- Baseline skin assessment and ordered skin checks to monitor picking
- Pain assessment including signs and symptoms, scale
- Child specific protocol for assessing injury
- Ordered orthotics for issues like scoliosis
- Climate control temperature regulations
- Education of staff and peers related to food sharing, etc.

## Resources & Manuals

**Foundation for Prader Willi Research**  
<http://www.fpwr.org/about-prader-willi-syndrome/>

**Youtube video: Prader Willi syndrome– Teacher tips**  
<https://www.youtube.com/watch?v=Y5LhSePDvqk>

**Prader Willi Syndrome Association**  
<http://www.pwsausa.org/>