PRADER-WILLI SYNDROME

What is it?

Prader-Willi syndrome (PWS) is a complex genetic disorder affecting metabolism, appetite, growth, cognitive and behavioral functions. PWS is a defect on the 15th chromosome and the most common genetic cause of life-threatening obesity. The key feature of PWS is a constant sense of hunger that usually begins at age 2. PWS affects the hypothalamus; responsible for helping 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 during infancy, an infant displays low tone, weak cry, and poor suck. These infants are often diagnosed with failure to thrive because of their feeding difficulties and poor weight gain. 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 characteristics or complications?

Signs and symptoms that may be present at birth include poor muscle tone, distinct facial features, poor feeding, unusual tiredness or generally poor responsiveness, and underdeveloped genitals. Motor milestone and language developed are often delayed. As the child grows into early childhood and adult, the signs and symptoms of PWS that appear will remain throughout life. These features include:

- Food craving, uncontrollable eating
- Weight gain
- Unusual food seeking behavior
- Cognitive impairment
- Poor growth and physical development
- Delayed motor development
- Hypothyroidism
- Behavioral problems including tantrums, obsessive/compulsive behavior, skin picking
- Sleep disorders
- Higher pain threshold
- Low tolerance for external temperature
- Scoliosis
- Underdeveloped sex organs
- Dental issues, thick saliva
- Obesity related complications
  - Type 2 diabetes
  - Heart disease
  - Stroke
  - Sleep apnea

What is the treatment?

There is no cure for PWS and effective treatment will be based on individual management of symptoms. Treatment plans could include infant feeding protocol for difficulty eating as well as strict food intake supervision as the child grows. A low calorie diet and regular exercise are important. Height, weight, and BMI should be monitored. Other treatment could include growth hormone therapy, physical therapy, and behavioral therapy.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

The team should consider the needs of the student that could include safety, stamina, behavior, and learning disabilities. Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluations
- Communication with parents/guardians regarding food policy at school during special events
- Plan for food security
- Structured day
- A "safe zone" or cooling down area
- Supervision when the student is eating
- Assistive technology
- Extended time
- Visual schedule
- Behavioral Support Plan
- Extra time to ambulate in hallway
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis including age and other characteristics the child may display
- Nutrition orders including calories for meals, snack
- Supervision for meal times
- Pain assessment including signs and symptoms, scale
- Child specific protocol for assessing injury
- Ordered orthotics or braces, assess skin and follow protocols for devices
- Climate control temperature regulations
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each (including event of severe stomach illness as possible binge episode or for vomiting as children with PWS rarely vomit)

Resources & Manuals

National Institute of Health (NIH)- Prader-Willi
https://ghr.nlm.nih.gov/condition/prader-willi-syndrome#genes

Prader Willi Syndome Association (USA)
https://www.pwsausa.org/

Prader-Willi Syndrome Association, Information and advice for school professionals
https://www.pwsausa.org/wp-content/uploads/2015/12/Supporting-Students-with-PWS-AAB-Resource.pdf

Foundation for Prader-Willi Research
https://www.fpwr.org/about-prader-willi-syndrome