

# Prader-Willi Syndrome

## Background

Prader-Willi syndrome (PWS) is a neurodevelopmental genetic disorder that causes physical, psychological, and behavioral challenges. Impaired hypothalamic development and function affect the body's ability to regulate essential functions, including hunger and metabolism. As a result, individuals experience chronic insatiable hunger and a slowed metabolism.

During infancy, children are often diagnosed with failure to thrive related to feeding difficulties, poor weight gain, and low tone. As they grow, muscle strength and tone typically improve. However, a constant sense of hunger often begins around age two. Motor and speech milestones are often delayed, and distinctive facial features may become more recognizable.

From early childhood to adulthood, food will be a trigger for individuals with PWS. Children may display patterns of behaviors including uncontrollable eating and unusual food-seeking. Low metabolic rate also significantly increases the risk for morbid obesity. Other features of PWS including poor growth and physical development, cognitive impairment, and behavioral problems will continue into adulthood.

Further physical symptoms may include sleep disorders, vision problems, impaired thermoregulation, and high pain threshold. Obesity related complications include type 2 diabetes, sleep apnea, heart disease and stroke. Mild to moderate learning disability, tantrums, obsessive-compulsive behaviors, or repetitive behaviors like skin picking are possible.

There is no cure for PWS. Effective treatment will be based on individual management of symptoms. Treatment plans could include a feeding protocol for infants who have difficulty eating and strict food intake supervision as the child grows. A low-calorie diet and regular exercise are important. Other treatments could include growth hormone therapy, physical therapy, and behavioral therapy.



## Top Takeaways for School

Prader-Willi syndrome (PWS) is characterized by uncontrollable hunger drive combined with decreased metabolism and unique behavior challenges.

Pain tolerance may be diminished or absent. Injuries should be reported to the school nurse. Subtle changes in condition or behavior should also be reported to rule out physical cause.

Staff working with the student should be aware of plans for food management (e.g., scheduled and predictable routines, supervision or support, safe storage). Food and snacks should not be used as incentives or rewards in the classroom.

Students with PWS may struggle with executive function but they have strength in visual learning. The use of visual aids for repetition, memory, planning, and organizing may be helpful.

## Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnoses: Risk for imbalanced nutrition, altered pain threshold, fatigue and impaired thought process
- Nutrition interventions and equipment (consider food management plan, supervision, etc.)
- Allergies or food restrictions
- Student-specific triggers, avoidance, or intervention strategies
- Temperature regulation considerations in school setting and transportation
- Use of specialized equipment, adaptive equipment and orthotics
- Consider emergency action plans (EAPs) and emergency evacuation plans (EEPs) related to special health care needs, including staff education/training

## Discussion Starters for the Educational Team

1. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
2. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
3. Does the student need additional adult support to access the academic curriculum in the least restrictive environment?
4. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?
5. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule
6. Will staff receive education/training to implement the student-specific emergency plan?

## Resources

Kennedy Krieger Institute: Neurology and Neurogenetics Clinics  
[kennedykrieger.org/patient-care/centers-and-programs/neurology-and-neurogenetics-clinics](https://kennedykrieger.org/patient-care/centers-and-programs/neurology-and-neurogenetics-clinics)

Foundation for Prader-Willi Research  
[fwr.org/what-is-prader-willi-syndrome](https://fwr.org/what-is-prader-willi-syndrome)

Prader Willi Syndrome Association (USA)  
[pwsausa.org/](https://pwsausa.org/)



For more information, please scan the QR code or visit: [KennedyKrieger.org/SHNIC](https://KennedyKrieger.org/SHNIC)

