PRADER-WILLI SYNDROME

Background

Prader-Willi syndrome (PWS) is a neurodevelopmental genetic disorder causing physical, psychological, and behavioral conditions. Impaired hypothalamic development and function affects the ability to control and regulate bodily functions and chronic feelings of insatiable hunger and slowed metabolism are present.

During infancy, infants are often diagnosed with failure to thrive related to feeding difficulties, poor weight gain, and low tone. As they age, infants often gain strength and muscle tone before a constant sense of hunger onsets usually around age two. Motor and speech milestones are often delayed, and distinctive facial features may begin to be recognized.

From early childhood to adulthood, food will be a trigger for individuals with PWS. Children may display patterns of behaviors including:

- Uncontrollable eating
- Unusual food seeking behavior
- Low metabolic rate that significantly increases risk for morbid obesity

Other features of PSW such as cognitive impairment, poor growth and physical development, and behavioral problems will remain throughout life. Mild to moderate learning disability, tantrums, obsessive-compulsive behaviors, or repetitive behaviors like skin picking are possible. 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.

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 Considerations

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 diagnosis of risk for imbalanced nutrition, altered pain threshold, fatigue and impaired thought process
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Nutrition interventions and equipment needs (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 care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained to implement the student-specific emergency plan?
2. 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?
3. 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)?
4. Does the student need additional adult support to access the academic curriculum in the least restrictive environment?
5. 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)?
6. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?

Resources

Kennedy Krieger Institute: Neurology and Neurogenetics Clinics
kennedykrieger.org

Foundation for Prader-Willi Research
fpwr.org/what-is-prader-willi-syndrome

Prader Willi Syndrome Association (USA)
pwsausa.org/

Scan QR code or visit **KennedyKrieger.org/Redirect** for more information.

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