

Prader-Willi Syndrome (PWS)

What is Prader-Willi syndrome (PWS)?

Prader-Willi syndrome (PWS) is a genetic condition that affects the brain and causes newborns and young infants to be weak and slow to gain weight but shifts to causing excessive hunger and weight gain in toddlers. It affects boys and girls equally.

How common is it?

Prader-Willi syndrome occurs at 1 in 15,000 to 30,000 births.

What are some common characteristics of children who have PWS or of PWS as children present with it?

Children with PWS tend to be short and to have obesity, with characteristic facial features. Findings include short stature, incomplete sexual development, intellectual deficits, and behavioral issues. They have obsessive behavior toward foods, which can cause problems, including aggressive behavior and eating inappropriate foods. They can have sleep problems with and without having obesity. Developmental delays are common, including gross-motor and speech delays. Autism spectrum disorder is diagnosed in about 25% of the children. Dry mouth and eye problems can also be seen, as well as hip and spine abnormalities. Obsessive behavior such as skin picking may occur, especially as the children age. Pain tolerance is very high. Children with PWS enjoy predictable routines.

Who might be on the treatment team?

- Pediatrician/primary care provider in the medical home
- Subspecialists, including geneticists, developmental-behavioral pediatricians, and endocrinologists
- Dietitians, who are crucial team members
- Physical, occupational, and speech-language therapists
- Classroom aides, who may be needed to help manage behaviors and meals

What adaptations may be needed?

Medications

Vitamins may be prescribed. Growth-hormone therapy is frequently prescribed but can be given at home.

Dietary Considerations

Management of food intake is important. A strict food plan may need to be in place. Placing locks on food storage sites and garbage cans may be necessary. Meals should be served on time. Other children should not carry food items that can be taken away.

Physical Environment and Other Considerations

- Limits on places where food is stored or disposed may be needed. Positive reinforcement of appropriate food intake may be needed. Behavior management programs that reward good behavior and strict routines can be very effective. Keep instructions short and simple. If the child keeps repeating the same question, write down the answer. Vigorous physical activity for 30 to 60 minutes daily is commonly recommended. More-frequent toothbrushing and oral care may be needed because of an increased risk for dental caries (cavities).
- Children with PWS should not be allowed to eat during bus transportation, and other children should not have food present.

What should be considered an emergency?

- **Breathing problems during sleep:** Monitor sleep closely, and report any breathing problems during sleep to the family.
- **Seizures:** Of children with PWS, 5% to 10% may develop seizures, especially with fever.
- **Vomiting:** Vomiting is not an emergency, but episodes of vomiting, especially after binge eating, should be reported to the family.

What are some related Quick Reference Sheets?

Autism Spectrum Disorder (ASD) (page 83)

What are some resources?

- American Academy of Pediatrics: www.aap.org—McCandless SE; American Academy of Pediatrics Committee on Genetics. Health supervision for children with Prader-Willi syndrome. *Pediatrics*. 2011;127(1):195–204
- Behavioral Data Collection Sheet (page 215) in Chapter 12
- International Prader-Willi Syndrome Organisation: www.ipwso.org
- Prader-Willi Syndrome Association (USA): www.pwsausa.org, 1-800-926-4797