



# Behavioral Health Nutrition

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## BHN Fact Sheet: Prader-Willi Syndrome Children

Prader-Willi syndrome (PWS) is a genetic condition caused by the absence of chromosomal material. Early identification becomes the key to avoidance of poor growth in infancy and prevention of obesity in children and adults. Nutritional guidance should be obtained throughout childhood, adolescence, and into adulthood.

### General Information:

PWS occurs with a frequency of 1 in 10,000 to 1 in 25,000 births. It is estimated that 50% of individuals with PWS are developmentally delayed and have learning disabilities. Characteristic physical health and developmental features of PWS may include:

- Developmental delays
- Low muscle tone (described as feeling floppy)
- Short stature
- Small hands and feet
- Incomplete sexual development
- Unusual facial features: turned down mouth, thin upper lip, narrowing head at the temples
- Insatiable appetite usually leading to obesity
- Behavioral issues

### Nutrition Concerns/ Implications

#### Infancy

In infancy the low muscle tone often results in feeding problems and poor weight gain. Infants are often given a diagnosis of "Failure to Thrive" because their rate of weight gain is lower than other children at the same age.

Infants often have low muscle tone and a weak suck, which may cause low rate of weight gain and poor intake. Infants may also have generally poor responsiveness, seem to lack energy, and sleep excessively. Later in infancy, body composition changes can be seen. Muscle mass decreases and body fat increases. Body fat is usually deposited in the thighs, buttocks and abdominal areas. Delayed motor development (sitting up, crawling and walking) decreases calorie needs and leads to weight gain.

#### Toddler to Adulthood

Short stature in an individual with PWS has been attributed to growth hormone deficiency. Growth hormone therapy was approved by the Food and Drug Administration in 2000. Once identified, treatment should begin before 18 months of age. Good nutrition support is key to ensuring optimal growth and preventing excessive weight gain.

At about 15 months of age, weight changes are seen along with an increase in appetite and food intake.

Insatiable appetites begin to appear and the child may become upset if food is not provided. As the child ages food searching may become an issue. Often the kitchen is locked along with cupboards and the refrigerator.

Children with PWS may have low blood levels of insulin. Insulin is a hormone that helps regulate glucose (sugar) in the blood. A lack of insulin can cause a form of diabetes. Your medical doctor and registered dietitian nutritionist (RD/RDN) may recommend regular blood sugar tests.

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continued

## Nutrition Tips & Advice:

### Infancy

Breast milk or formula feeding is recommended until 1 year of age. If the infant is unable to suck adequate amounts of milk, concentrating either the breast milk or the formula may be required. The RDN can determine the amount of milk to give and calories needed. The RDN may also recommend feeding therapy with an occupational therapist who can assist with the infant's weak sucking. Baby food can be started between 4-6 months of age if head control and trunk stability are achieved.

### Toddler and Preschool Age

Begin making feeding and mealtime a structured event. Give small amounts of meats, fruits, vegetables, grains, milk in a regular 3-meal pattern with snacks between meals. Avoid grazing or eating throughout the day. Introduce cup drinking at 10 months of age for gradual weaning from the breast or bottle.

### School Age to Adult

Talk with the school food service program if a low calorie diet is needed. Your physician can provide a prescription.

Meals should be planned to consist of a variety of vegetables, lean meat, fish or poultry, fruits, grains and dairy products. Low fat or nonfat milk should be used. The size of any food served should be small with the possibility of a second helping of vegetables or fruits.

Include the individual in any form of physical activity to increase energy expenditure. Supervised swimming is an excellent sport for the child or adult with PWS when physical balance is a problem.

Prevention of obesity is critical for the child or adult with PWS. The constant craving for food and rapid weight gain require frequent monitoring of weight. Obesity can lead to sleep disorders, apnea, daytime sleepiness, type 2 diabetes, heart disease or stroke.

Work with a behavioral therapist as needed when access to and desire for food results in adverse or negative behavior.

## Typical Nutrition Intervention

- Measure height, weight, head circumference (infant and toddler)
- Plot measures on the appropriate growth chart or weight record
- Review usual food and beverage intake, frequency and timing of meals and snacks, and size of servings offered strategies
- Discuss parent/caregiver's description of feeding ability, problems, and concerns
- Observe actual feeding or eating
- Review clinical factors, including available laboratory data and sleep issues
- Recommend calorie and nutrient needs
- Work with the individual/family/caregiver/team to identify and apply strategies that best support the person with PWS

## Resources for Further Information

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

Miller JL, Lynn CH, Driscoll DC, et al. Nutritional Phases in Prader-Willi Syndrome. American Journal of Medical Genetics Part a. 2011;155A(5):1040-1049. doi:10.1002/ajmg.a.33951.

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More topics and resources available at [bhndpg.org](http://bhndpg.org).

This information does not take the place of medical advice. Please contact a Registered Dietitian/Nutritionist or medical doctor for more information.

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