



Prader-Willi syndrome Causes an Insatiable Appetite

Prader-Willi syndrome (PWS) is a non-inherited genetic disorder that causes a host of symptoms including hyperphagia, or insatiable appetite and often very difficult behavior problems. Persons with PWS do not feel full no matter how much they eat; the brain continuously and relentlessly drives someone with PWS to seek out and eat more food. At the same time, the body metabolizes food at about half the normal rate, so someone with PWS will gain weight at a tremendous rate on few calories.

There is no known treatment or appetite suppressing medication yet to eliminate or even reduce PWS' hyperphagia symptom. There is no "behavior modification" in regard to food. It is not realistic to expect that the student with PWS can "learn to control himself" and not eat food that is available. While some children may be able to manage their behavior for a day or even a week or two, this does not constitute genuine modified behavior; there will come a day when the child, despite knowing that he should not, will attempt to eat excess food. Individuals with PWS know they're not supposed to eat "unauthorized" foods; they simply cannot overcome their biochemical drive to eat.

For the student with PWS the emphasis is always on prevention. **The key to managing the hyperphagia caused by PWS and reduce the incidence of food-related behavior issues is to restrict access to food within all environments and provide continuous supervision of the child at all times.**

But Just "One" Can't Hurt... Can It?

For the child with PWS, just "one" or just one food binge *can* hurt... It can even be deadly. Here's why:

Rapid Weight Gain

Due to a reduced metabolic rate and other endocrine issues persons with PWS can quickly gain large amounts of weight on very few calories. A diet considered healthy for a child without PWS can cause rapid weight gain for the child with PWS. The daily caloric intake of the child with PWS must be strictly monitored; even just one cookie or one extra slice of pizza every now and again can mean the difference between a healthy weight and a tip of the scale into morbid obesity. If a child is known to have obtained extra food during the school day, parents must be notified so that they may compensate and reduce the child's caloric intake for the remainder of the day or week.

Choking

Choking is a particular hazard for persons with PWS who often eat much too quickly and do not adequately chew their food before swallowing. Sneaked or stolen food will be eaten very, very quickly to avoid detection or loss of the forbidden item. Low muscle tone and poor oral-motor coordination exacerbate the choking risk.

Severe Gastric Illness

Abdominal distention or bloating, pain, and/or vomiting may be signs of life-threatening gastric inflammation or necrosis and is more common in PWS than in the general population. Thinner individuals are at even greater risk. Rather than localized pain the student may complain of a general feeling of unwellness. If a child with PWS has these symptoms, especially following a known or suspected binge episode, immediate parental notification and hospitalization is warranted as this may be a life-threatening situation.

Lack of Vomiting

Vomiting rarely occurs in individuals with PWS due to an impaired Central Nervous System. Emetics may be ineffective and repeated doses may cause toxicity. The presence of vomiting may signal a life-threatening illness and warrants immediate notification of the parents and possible hospitalization.

The Principles of Food Security

Food Security is a term coined by PWS specialists Linda Gourash, M.D. and Janice Forster, M.D. and refers to "the ready availability of nutritionally adequate and safe foods with an assured ability to acquire acceptable foods in socially acceptable ways."

The Principles of Food Security are:

No Doubt when meals will occur and what foods will be served *plus*

No Hope/Chance to obtain additional, unauthorized foods *equals*

No Disappointment related to unmet expectations. *No disappointment means fewer behavior problems.*

The Hand is Quicker than the Eye

Regardless of cognitive function, a child with PWS can be quite adept at obtaining food. Educators should be aware that children with PWS have been known to:

- Take food out of someone else's lunchbox or off another's lunch tray, or from the teachers' lounge
- Trade valuable items for food such as jewelry
- Steal money to purchase food from the cafeteria, vending machine, and other students
- Eat their own packed lunch and attempt to obtain more food from the cafeteria
- Take half-eaten or discarded food from the trash
- Take food from backpacks left in corridors while on the way to the restroom
- Eat all of their lunch on the bus while on the way to school

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Children with PWS may not be truthful about whether or what they've eaten regardless of the "evidence." It is generally recommended not to ask "Did you eat that?" but rather presume food was eaten and notify the parents.

PWS' Impact in the Classroom

Giving in to tantrums, particularly tantrums for food, creates spiraling out-of-control behavior. It is unrealistic to expect a student with PWS to maintain focus on the task at hand when he knows that a food or beverage may be obtainable; the drive to eat is too strong and will affect behavior and school performance. For most students with PWS, having any food anywhere in the classroom will impede their ability to concentrate and learn. Maintaining Food Security will help improve learning and the overall behavior of the student with PWS.

Tips to Maintain Food Security

- Educate and inform *all* people working with the student including bus drivers, custodians, secretaries and volunteers.
- Secure access to food across all settings including the cafeteria, lunch boxes, hallways, vending machines and money
- Supervise food exposure at all times, including during bus rides and especially in the lunchroom and wherever there are food related activities. In some cases the student may need to eat snack or lunch in his classroom with a peer or friend. Some students may require supervision in hallways
- Post the schedule for snack and lunch times
- If the student states he has not had breakfast call the parents or caregiver *before* giving food. The child may have already eaten a healthy breakfast and is hoping to obtain more food.
- Lock up all sources of money including purses. Avoid allowing the student to carry money.
- Address any stealing or trading of food in private. Never be punitive—stealing and lying are symptoms of syndrome!
- Never try to take food out of the child's hand unless it is spoiled or dangerous as this may lead to meltdown behavior. Instead, encourage the individual to put the food down and later inform the parents of the eaten food. Secure that food source to ensure against a reoccurrence.
- Follow parental guidelines for treats or eating of extra food. Communication with home is very important.
- Don't delay snack or lunch when at all possible. If delays are necessary discuss ahead of time with the student.
- Limit availability and visibility of food. Remove candy dishes. Do not offer a food treat to another child in the presence of the child with PWS who cannot also enjoy that food treat.
- Do not use food as a reward, incentive, or punishment. Instead, the use of other systems of reward or incentive is encouraged, such as stickers, small toys, tokens for the "purchase" of small toys, etc.
- Be aware of food aromas. There is nothing like the smell of popcorn or cookies to make a student with PWS agitated and increase the potential for anxiety and a behavior problem.
- When going on a field trip or other outing, discuss all food-related issues *ahead* of time. Will you bring a snack along or will it be purchased? If purchased, what will it be? Will the outing interfere with the time of a meal?
- Obtain weekly weight by school nurse if indicated.

For More Information about PWS

For more information about PWS, speak with someone about how to organize the classroom to assist the student, or to request a school training, contact PWCF at 310-372-5053 info@pwc.org or visit www.PWCF.org