



Prader-Willi syndrome (PWS) is a complex, lifelong, life-threatening disorder that causes myriad symptoms including low muscle tone, sensory integration dysfunction, and cognitive and learning challenges. But the hallmark symptoms of PWS are emotional lability and hyperphagia – an excessive drive to eat.

Persons with PWS always feel a physiological, overwhelming, and overriding drive to eat. At the same time, their bodies metabolize food at about half the normal rate which means they gain weight very quickly. Because the stomach muscles are weak and the ability to vomit is almost always absent, even a single gorge episode can lead to death due to gastric rupture or tissue death. It is critical to follow the *Principles of Food Security*\* and limit the individual's access to food at all times.

Individuals with PWS generally love life and people but frequently have a difficult time managing their behavior when they feel frustrated or anxious. It is important to know about the general Do's and Don'ts when living, playing, and working with someone who has PWS so that you can help them manage their feelings and behavior better.

### DO'S

- DO** Keep all food items inaccessible at all times other than mealtimes. People with PWS cannot fight the physiological drive to get food. Lock away all access to food. Know that the hand of someone with PWS is quicker than your eye!
- DO** Post the schedule of the day's events and a set of the rules. Read and discuss them at the onset of the day so they are clearly understood.
- DO** Give transition prompts (5, 3, 1 minute cues) before moving on to the next activity. It is important for someone with PWS to know of change a bit in advance.
- DO** Keep in mind that persons with PWS respond exceptionally well to praise, encouragement, positive reinforcement, smiles and hugs. Keep upbeat, positive, and encouraging despite what you may feel on the inside!
- DO** Remember to always speak to the individual with PWS calmly. Persons with PWS often feel a global sense of anxiety which is typically increased by raised, critical, or angry-sounding voices. Ignore inappropriate behavior that may be an attempt to get your attention. Respond to a raised voice or a temper outburst with calming words such as, "You want xyz. I understand. You want xyz and you feel frustrated. I am sorry you feel frustrated."
- DO** Be specific in what you want the individual *to do* rather than what *not* to do, e.g., "Please stop taking the items out of the box. Please put all of the items back into the box and place the lid on it. Thank you!"
- DO** Create written agreements if there is a disagreement over something. Agreements should be created collaboratively.
- DO** Refer to the written schedule or contract to avoid a power struggle or supposed misunderstanding.
- DO** Keep in mind that logic and reason will not prevail when the person with PWS gets upset and/or "stuck" on an idea or position. Showing empathy by repeating back to the person what he's said that he feels or wants can be helpful to get him "unstuck."
- DO** Avoid ambiguity. "In a while" or "Later" should be restated as, "After lunch at about 2:00"
- DO** Listen to the person who needs to talk. The time it takes to listen and express genuine empathy is worth it. If the person asks the same question repeatedly, ask the person what *he* thinks the answer is, confirm it, and then calmly let the person know that question is all done being asked and answered. Allow the person with PWS to have the last word.
- DO** Remember that the brain of someone with PWS is typically oppositional. Avoid asking a question when you should make a statement. E.g., If you need the individual to leave promptly, don't ask, "Are you ready to go now?" Instead, after providing 2-3 prior prompts, state, "Ok, it's time to go now. Thank you so much for getting up to go to the next adventure!"
- DO** Give a great deal of time for the individual to process what you are saying. Many people process information a bit slowly.
- DO** Keep in mind that persons with PWS are typically egocentric and self-focused.
- DO** Know that stealing, lying, and making up stories are common behaviors for some persons with PWS. Keep all of your personal belongings safely locked up.
- DO** Treat each person with respect and dignity.

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\*Term coined by Janice Forster, M.D. and Linda Gourash, M.D. of Pittsburgh Partnership, Specialists in Prader-Willi Syndrome [www.pittsburghpartnership.com](http://www.pittsburghpartnership.com)

## Strategies for Improving Behavior In Persons with Prader-Willi Syndrome

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### DO NOT'S

- DO NOT** leave food accessible. It is important to lock kitchens and other food sources to remove temptation. Food issues are one of the most common reasons for misbehavior and tantrums .
- DO NOT** take food out of anyone's hand or attempt to move it from them (unless it is spoiled and would cause illness if consumed) as this will initiate a behavioral outburst. Instead, encourage the person to put food item back or monitor person so that they don't choke.
- DO NOT** argue with the individual. If the person with PWS is building up to a disagreement, try to walk away or guide him to a private area and discuss it there. Frequently discussions can be ended if you allow the person to have the last word. Try to make the statement, "I will make one more comment and then you can make one more comment and then the subject is all finished being talked about."
- DO NOT** be sarcastic. People with PWS often find it difficult to understand sarcasm and other abstract concepts. Good-natured teasing is accepted, enjoyed, and looked forward to.
- DO NOT** promise anything that you cannot or will not do. People with PWS are uncomfortable with inconstancy and uncertainty. If changes are necessary, prepare the person so he does not feel deserted or threatened.
- DO NOT** ignore the person with PWS who has an obvious need to talk.
- DO NOT** try to talk anyone out of a disappointment or upset as they'll only become *more* upset in an attempt to get you to understand.

*Excerpted from Op Cit, Thompson, Dorothy C., et al, pages 125-127 and Prader-Willi Syndrome Association, Prader-Willi Syndrome Management pamphlet*



### MEDICAL ALERT INFORMATION

**Hyperphagia (Excessive Appetite):** Insatiable appetite may lead to life threatening weight gain, which can be very rapid and occur even on a low-calorie diet. Individuals with PWS must be supervised at all times in all settings where food is accessible.

**Respiratory Concerns:** Individuals with PWS may be at increased risk for respiratory difficulties. Obesity adds to these concerns.

**Body Temperature Abnormalities:** Idiopathic hyper- and hypothermia have been reported. Hyperthermia may occur during minor illness and in procedures requiring anesthesia. Fever may be absent despite serious infection.

**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. Rather than localized pain, there may be a general feeling of unwellness. If a child with PWS has these symptoms, especially following a known or suspected binge-eating episode, immediately notify parents and prepare for immediate hospitalization.

**Lack of Vomiting:** Vomiting rarely occurs in persons with PWS. The presence of vomiting may signal a life-threatening condition. If poisonous materials are ingested, emetics may be ineffective and repeated doses may cause toxicity.

**Adverse Reactions to Some Medications:** People with PWS may have unusual reactions to standard dosages of medications. Use extreme caution in giving medications that may cause sedation.

**High Pain Threshold:** Lack of typical pain signals is common and may mask the presence of infection or injury. Someone with PWS may not complain of pain until infection is severe or may have difficulty localizing the pain.

**Skin Lesions and Bruises:** Because of a habit that is common in PWS, open sores caused by skin picking may be apparent. Individual with PWS also tend to bruise easily.

#### FOR MORE INFORMATION ABOUT PRADER-WILLI SYNDROME CONTACT

**Prader-Willi California Foundation** 800.400.9994 | [info@pwcf.org](mailto:info@pwcf.org) | [www.PWCF.org](http://www.PWCF.org)

**Prader-Willi Syndrome Association (USA)** 800.926-4797 | [info@pwsausa.org](mailto:info@pwsausa.org) | [www/pwsausa.org](http://www/pwsausa.org)