

Prader-Willi Syndrome



An overview of how to manage food and behavior when caring for someone with Prader-Willi Syndrome especially for extended family, babysitters, and community personnel.



Never give in to a temper tantrum or a sit down strike whether it's for food or anything. Once you give in you have taught the person with PWS that next time you will eventually give in if they just cry louder, longer, or behave with increased aggression.

Have patience. And then have more patience. People with PWS typically process or think about things a bit more slowly and may need extra time to comply with a request.

Have fun! When the needs of the individual with PWS are addressed there will be few if any behavioral problems. So, enjoy the time you spend in the company of your loved one or friend with Prader-Willi syndrome!

Prader-Willi California Foundation and the national **Prader-Willi Syndrome Association (USA)** have an extensive amount of materials available for families, friends, physicians, therapists, caseworkers, and school staff. Please contact PWCF or PWSA (USA) when we may provide more information, support and advocacy services to you, your child, your friend, patient, or your student.

To learn more about Prader-Willi syndrome and how you can help, call us or visit www.PWCF.org

Prader-Willi California Foundation is a non-profit 501(c)(3) charitable corporation established in 1979. An affiliate of Prader-Willi Syndrome Association (USA), the Foundation is dedicated to assisting individuals with Prader-Willi syndrome, their families, and the professionals who serve them.



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PRADER-WILLI SYNDROME ASSOCIATION USA
Still hungry for a cure.

Low Tolerance to Frustration Almost all people with PWS have difficulty managing feelings of frustration and disappointment. To the extent possible, eliminate the potential for frustrators and disappointment and use positive-toned language. For example, the person with PWS says, "I want to go to the movies" but you know you can't schedule it today. Instead of saying, "No, we can't do that today" reply, "That sounds great! I'd like to go to the movies too! Today we need to xyz. Let's see what movies are playing and plan to go tomorrow!"

Hyper-Reactive Almost all people with PWS can be described as "hyper-reactive," meaning they tend to become overly upset about a seemingly minor incident. When someone with PWS becomes upset it's best not try to talk to them out of their upset as this will cause them to become even *more* upset and work even *harder* to show you how upset they are. Instead, immediately and calmly demonstrate understanding and empathy as this will help them recover more quickly. For example, "You want to see a movie today. You don't want to wait. I'm so sorry." Then stop talking or only occasionally and calmly repeat their upset. People with PWS are often concerned with "fairness." Sometimes simply agreeing to show you understand their upset will be calming, "You're right, it's not fair and I am sorry."

Oppositional Most people with PWS are oppositional; the brain reflexively thinks, "No!" Strategies to counter this are to refrain from giving "yes or no" choices. For example instead of asking, "Are you ready to get dressed now?" give two or three preferred choices such as, "Do you want to get dressed in 3 minutes or 5 minutes?" As much control over things the individual with PWS perceives he or she has, the better will be their behavior.

Inflexible People with PWS often have a difficult time transitioning from one thought or activity to the next. Give prompts to end a conversation about a particular topic: "This has been fun to talk about xyz. You can say one more thing and then let's talk about something else." Give incremental prompts before moving to the next activity. For example prompt, "Five minutes before we need to leave... Ok, 3 minutes before we need to leave so start putting away your books... Ok, 1 minute. Everything should be put away. Get your shoes on." Some people with PWS move more slowly so make sure you carve out lots of time for transitions.

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This rare medical disorder, pronounced PRAH-der WILL-ee, affects multiple systems in the body and causes many different symptoms. Many of these symptoms are very serious and some of them are life-threatening.

People with Prader-Willi syndrome (PWS) experience a host of challenges including weak muscles, problems with balance and coordination, developmental delays, speech and language problems, temperature regulation problems, orthopedic problems, and sleep problems to name a few.

This brochure will provide you with an overview of important information you need to know when someone with Prader-Willi syndrome is in your care.

Food and Weight Challenges

Prader-Willi syndrome is best known for its symptom called “hyperphagia” which means literally “over eating.” For reasons not yet understood, the brain of someone with PWS drives the individual to want to eat as much food as possible all of the time. Persons with PWS feel a constant drive to eat that is *never* satisfied no matter *how* much food is eaten. At the same time, metabolism rate slows to about half what it should be. **If caloric intake is not significantly reduced and strictly controlled, persons with PWS will quickly gain an enormous amount of weight leading to morbid obesity and premature death.** People with PWS are vulnerable to sudden death due to choking, stomach rupture, or stomach necrosis.

Just because someone has PWS doesn’t mean they are destined to become obese. People with PWS can absolutely maintain a healthy weight, but it takes constant and continuous supervision from not only their parents or primary care providers, but from *everyone* in whose care they are entrusted.

Behavioral Challenges

Second only to hyperphagia on the list of PWS challenges is the misbehavior that many people with PWS exhibit.

The brain of someone with PWS causes individuals to feel a global sense of anxiety, think oppositionally, have a low tolerance to frustration, be argumentative, and react to disappointment and frustration with tears or anger. Regardless of age, persons with PWS can exhibit temper tantrums that can include yelling, foul language, hitting, kicking, sitting and other refusal behaviors.

It’s important to understand how persons with PWS typically think and react so that you can better avoid the behavioral problems that are common in persons with PWS.

Strategies to Manage Symptoms

There are currently no known medications that eliminate or even reduce the insatiable appetite that is the hallmark symptom of PWS. There are, however, therapeutic and environmental interventions that can help manage weight, improve behavior, save lives, and improve the quality of life of *all* who are impacted by Prader-Willi syndrome.

Food Security

People with PWS cannot manage their own food intake because their brain tells them to eat as much food as possible which means they must depend upon everyone in their environment to make sure they are safe around food. The *Principles of Food Security*, a concept coined by PWS specialists Linda Gourash, MD and Janice Forster, MD, describe the basics of how to manage the food environment:

No Doubt that appropriate foods will be served at the appropriate time **plus No Hope** that extra, unauthorized foods can be obtained **equals No Disappointment**. No disappointment means fewer behavioral problems.

People with PWS need to know *that* they will eat and *when* they will eat. Having a written schedule when meals and snacks will be served is very helpful because once it’s written, it’s often not argued about. Stick to the schedule as persons with PWS cannot be expected to be flexible.

It’s helpful for the person with PWS to eat every 2 1/2 to 3 hours. Eating this frequently, however, requires a daily diet *very* low in calories, fat, and carbohydrates. Some people with PWS cannot exceed 800 calories per day.

Never use food as a reward or punishment. For someone with PWS food must be as stable as oxygen, *never* contingent upon good behavior and *never* taken away for poor behavior.

It is imperative to follow the menu instructions *exactly* as dictated by the parent. Because the metabolic rate is very slow, every calorie consumed by someone with PWS can potentially cause weight gain. Giving someone with PWS extra food causes not only weight gain but gives them hope that they will receive extra food from you later, and that hope will likely lead to a behavior problem some time down the road. There are many ways other than food to provide a special treat or show you care such as stickers, small toys, time playing together, employing the individual as your special helper, and hugs.

Before offering the individual with PWS food, ask the parent outside the presence of the individual with PWS what food options you may offer.

Secure and lock up all food sources. Almost all families who live with someone with PWS lock their refrigerator and food pantry. Not only does locking away food keep the individual with PWS safe, it tells their brain that food is not available and reduces the hopeful anxiety they feel to obtain that food. In addition, secure wallets, purses, and loose change because children and adults with PWS know that money can purchase food. Stealing is a symptom of PWS.

You will show the person with PWS and their family that you care about them by securing all food sources and by following all parental instructions.

Behavioral Interventions

It’s important to familiarize yourself with how PWS typically impacts the brain so you can minimize or avoid behavior problems. In general, work to create an environment that is predictable, positive, and free of stress. Use positive words, praise and encouragement; use a calm tone of voice; use rewards such as your attention, stickers or small toys; and engage the individual as your ‘special helper.’

Anxiety Almost all persons with PWS feel an underlying degree of anxiety all of the time and do best when they know what to expect and what is expected of them. Create a written schedule of the day’s activities including approximate timing of all snacks and meals. “Paint a picture” of new activities or situations; talk about what things might look or sound like, what can be expected to happen, etc. Talk about your expectations of behavior, and make a plan for what will happen if there is a behavior problem. Stick to the schedule to the extent possible as persons with PWS cannot be expected to be flexible.