

Weight & Behavior Management

Managing Prader-Willi Syndrome

Two of the most outstanding characteristics of Prader-Willi Syndrome (PWS) are insatiable appetite and behavior problems.

Prader-Willi Syndrome is a birth defect also characterized by hypotonia (weak muscles), developmental delays, short stature if not treated with growth hormone, small hands and feet, incomplete sexual development, and cognitive deficits which may or may not be at the level of mental retardation.

The craving for food is complicated by the fact that people with PWS have poor calorie utilization, gaining weight on approximately one-half the calories required by a normal person. The constant presence of food in our society creates further difficulty in the struggle to control appetite.

There are no easy answers to any of the problems surrounding the management of this unique condition. The following “do and don’t” suggestions have come from parents, professionals, and observations of persons with PWS. In addition, use good humor, kindness, affection, determination and respect, seasoned with flexibility and good judgement, and there will be a good amount of success.

Behavior management should be intervention/prevention oriented. For best results, pre-schedule, preplan, avoid arguments and anticipate.

Weight management should be individualized and include daily or weekly weigh-ins, complete control of food intake, environmental barriers to food access and diets acknowledging that fewer calories are required.

Prader-Willi: A Multistage Syndrome

The initial stage of Prader-Willi syndrome is characterized by low birth weight and subsequent failure to thrive.

The infant is very weak, usually cannot nurse or suck, and must be fed with special nipples or tubes. NOT gaining weight is a problem. Depending on the degree of hypotonia, this stage can continue for the first couple of years.

A program of good nutrition, proper food intake, and exercise started as soon as possible, rather than after weight starts to accumulate, will be extremely beneficial.

Behavior problems may be few or nonexistent in the young child, although individuals vary. A little stubbornness usually appears first, often progressing to temper tantrums.

The next stage reflects thriving almost too well. The food compulsion may surface with improvement of hypotonia, usually between the ages of two and four, but possibly earlier or later. Behavior may include greater stubbornness, temper tantrums increasing in number, length and intensity, and food sneaking, stealing, and hoarding.

When weight gain starts, complete control of caloric intake, aided by locks on cabinets and refrigerators, is essential. Without such control, 95 percent develop weight problems. A well-balanced diet limiting calories is a lifelong necessity.

DO

- Keep food inaccessible at all times. Persons with PWS cannot fight their compulsion to get at it. Put it away and lock the cupboards and refrigerator
- Keep their lives structured. They need structure. Preplan changes
- Praise and recognize good performances. A lot of mileage can be obtained with a few words, smiles and hugs
- Listen to a person who needs to talk. The time it takes may alleviate or prevent unpleasantness later
- Include the person with PWS in planning and programming. They feel a need for some control and will cooperate 100 percent if they feel it was their idea
- Keep sight of the fact that the hand of a person with PWS is quicker than your eye
- Enjoy your treats in private. It is very hard for the person with PWS to watch others enjoying the goodies he is denied
- Use smaller plates and cups, spread the food out, and add extra nonfattening items such as carrots, dill pickles or diet Jello to make the amount on the plate look larger
- Inform neighbors, relatives, teachers, babysitters, classmates, everyone with whom the child with PWS comes in contact – and then inform them again!
- Be consistent. They thrive on routines and knowing exactly what the guidelines are
- Remember that logic and reason will not prevail when the person with PWS gets upset and/or “struck” on an idea or position

DON'T

- Use food as a reward or punishment except on a very limited basis
- Assume, if the person has lost weight, that the problem is now “cured”
- Nag. Once a behavior has been dealt with, do not bring it up again. Discuss temper tantrums and then forget it
- Argue. Make the statement, allow the person one more comment, warn that the discussion is over and stick to it. You will never win an argument
- Tease, be sarcastic, or even use subtle humor. People with PWS do not respond well to such tactics
- Ignore bad behavior. Try interventions to prevent it
- Lose your temper. Easier said than done but do whatever it takes to keep your cool; nothing will be gained if you lose control
- Promise anything that you cannot or will not do. Persons with PWS rarely accept change
- Ignore poor table manners. They are capable of using utensils; they can slow down; they can stay until the meal is finished without additional food
- Try to talk things out with a child with PWS. It does not work!

- Lose sight of the humorous aspects of all of this. Hang on to your sense of humor while figuring out “how to lock up the apple tree”
- Hesitate to ask for professional help. IPWSO can give good references
- Forget that this is a life-threatening situation

Rebranded November 2022