

Pittsburgh Partnership

Specialists in Prader-Willi Syndrome

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Prader-Willi Syndrome: The Behavioural Challenge A Brief Summary for Professionals

The Professional's Role: Coach

PWS is a highly complex disorder; families need assistance in finding expert information and advice and often need coaching to use that help. Part of the complexity of the disorder is the wide range of manifestations in each patient and the wide variability of severity of each manifestation from patient to patient. In depth and broad experience with the syndrome is rare but consultation is available locally and through www.pwsusa.org, and other national Prader-Willi syndrome Associations.

Professionals working with PWS must remain humble and expect to function on a steep learning curve; the most common error is to underestimate the complexity of the disorder. The role of the professional is to *remain involved over time* and to become an “expert” in his/her own patient by helping the family to use the resources available and to apply the information to their particular situation.

PWS and “Hunger”

Clinical observation as well as neurobiological research point to a distinction between “hunger” and lack of satiety. The most common clinical error with PWS is the assumption that the ability to consume large amounts of calories and the incessant search for food is “hunger”. Neurobiological studies indicate that the feedback mechanism producing *satiety* is defective in PWS and at the same time functional MRI studies appear to show an enhanced pleasure from consuming edibles. Words such as “ravenous” and “starving” are inappropriate in describing PWS and lead sympathetic persons including family members to do the exact wrong thing. Persons with PWS require and tolerate a very low calorie diet without discomfort when they are given Food Security (described below).

Appetite suppressant medications are therefore inappropriate and ineffective in PWS.

Behavior Medication

Psychotropic medications should be prescribed cautiously and for specific psychiatric indications following a thorough evaluation which includes a patient interview. The psychiatrist must be savvy about the basic personality traits and features of the syndrome (see “The PWS Personality” and “PWS Primer for Psychiatrists”) and not confuse these

behaviors with treatable psychiatric symptoms or disorders. No medication should be prescribed because it “is good for PWS behavior”. Expert consultation is recommended. More information is available through the PWSAUSA website: Prader-Willi Syndrome: A Primer for Psychiatrists. When weight is well controlled by food control and a daily exercise program, psychotropic medications associated with weight gain can be used with close monitoring of the patient.

Behavior Management

Behavioral management of PWS is based on an infrastructure which includes food security and a behavioral support system. Unlike many other persons with behavioral difficulties, the person with PWS will ALWAYS need the food security (see below) and behavioral supports (e.g. low expressed emotion, verbal praise, rules, incentives, token economy, daily schedule). The proper goal for the person with PWS is maximal functioning with supports, NOT independent functioning. Maximal functioning comes from consistent reliable use of these modalities. Unlike other adults with mild cognitive deficits, persons with PWS should never be encouraged to “outgrow” the structure and support they need to function. Experience shows that these supports once needed, cannot be withdrawn or diminished even in adulthood no matter how high the individual’s IQ is or how cooperative the patient is or appears to be. Removal of these supports will lead to life threatening weight gain. This is the second most common error in caring for persons with PWS. Advocacy by the clinician caring for the person with PWS must be based on this understanding which is countercultural in the world of developmental disabilities.

Food Seeking Behavior

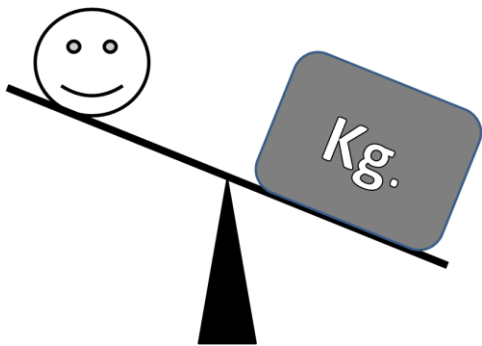
PWS is not a “food addiction” since it is an inborn abnormality and not acquired. Nevertheless, the third most common error in management of these patients is to underestimate their capacity for manipulation or the lengths that some persons will go to (very creatively) acquire food. Lying, manipulating, elaborate schemes and falsifications are all part of the PWS repertoire and should be expected and managed matter of factly and without registering surprise. It is incumbent on the caretakers to eliminate the possibility of obtaining extra food or of *successful* manipulation. These behaviors will not be “trained out” of the individual with PWS but attempts are minimized when food seeking is not rewarded by success or when opportunities are eliminated.

Behavior and Stress

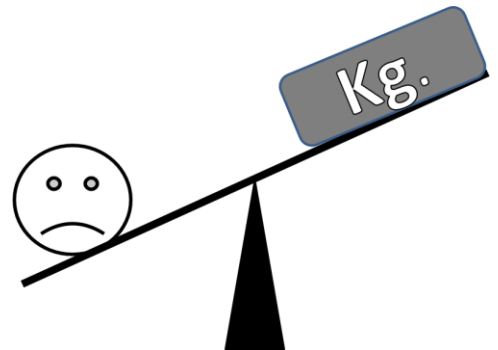
Persons with PWS are highly stress sensitive. Food is a stressor. Most behavioral problems can be traced to a breach in food security or other unrecognized stressor such as change in expectations by caretakers, loss of caretaker, mood disorder or medical illness. Angry, punitive, sarcastic or even annoyed caretakers can augment stress and increase behavioral problems.

Prader-Willi Syndrome

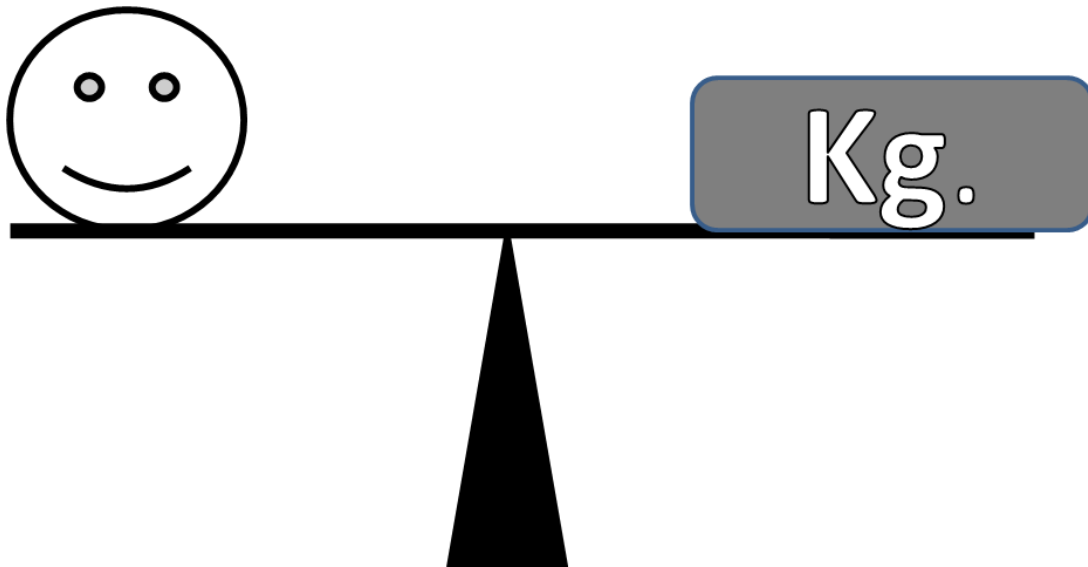
Food Security



Families and professionals often mistakenly believe that the patient cannot be happy unless he has as much food as he demands. Because efforts to limit food, if attempted without establishing food security cause increased stress and behavior problems.



When *Food Security* is fully implemented, weight and behavior are both managed successfully and simultaneously.



Food Security

Remember: "Food is STRESS"

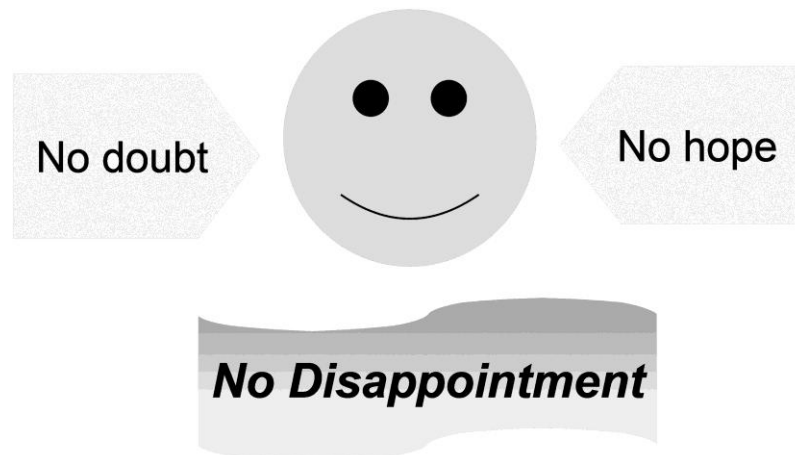
Food Security includes:

NO DOUBT

The person with PWS is able to relax and think less about food when he knows the plan for his food each day. This is achieved by a predictable routine for the day in which meals are scheduled reliably among his other activities. Focus on the *sequence* of events and not the *time* of each meal. Advanced planning assures the individual of what activities will precede the meal and which will follow. Advanced menu planning provides him with expectations which will be reliably fulfilled.

NO HOPE (NO CHANCE)

As children get older, opportunities for food acquisition increase and they require more measures to assure that they are not hopefully scouting for food all day. Chances to obtain food are stressful and therefore, as much as possible, should be eliminated. The measures taken will depend on the individual's history and capability of food acquisition



Successful Behavior Management of PWS means that uncertainty about food must be eliminated as much as possible. Advance planning of meals, a schedule of all the day's events with the place of meals clearly identified, reminders of these plans and a behavior program which requires completion of one task before the next activity (including meals) is begun, all contribute to successful behavior management.