Pittsburgh Partnership
Specialists in Prader-Willi Syndrome
www.pittsburghpartnership.com

Linda M. Gourash, MD
Developmental and Behavioral Pediatrics

Janice L. Forster, MD
Child and Adolescent Psychiatry

Prader-Willi Syndrome: The Behavioral 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. Extensive professional experience with the syndrome is rare but consultation may be 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 persons with 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 have been thus far ineffective in PWS.

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"). It is easy to confuse these behaviors with treatable psychiatric symptoms or

disorders. The most common error is to confuse the repetitive (perseverative) questions, actions and collecting behaviors of PWS with OCD. Please see “PWS Primer for Psychiatrists”.

No medication should be prescribed because it “is good for PWS behavior”. Expert consultation is recommended. More information is available through the PWSA-USA 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. After the developmental period, persons with PWS should never be encouraged to “outgrow” the structure and support which they have shown a need for to function. Experience shows that these supports once needed are generally not likely to be withdrawn successfully or diminished no matter how high the individual’s IQ is or how cooperative the patient is or appears to be. Removal of these supports can 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.

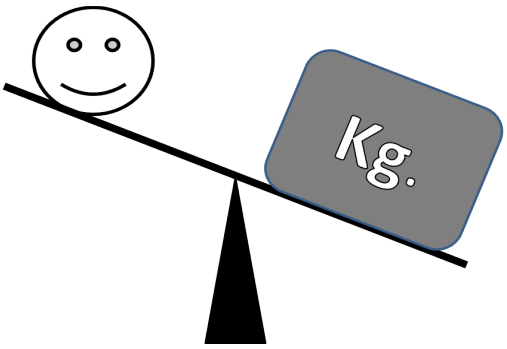
Food-Seeking Behavior

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 caregivers 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 incidents of success or when it is clear to the individual that opportunities are eliminated.

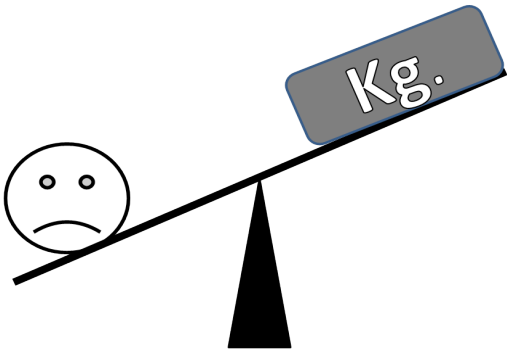
Behavior and Stress

Persons with PWS are highly stress sensitive. Food is a stressor. Most behavioral problems can be traced to a breaches in FOOD SECURITY or other unrecognized stressors 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.

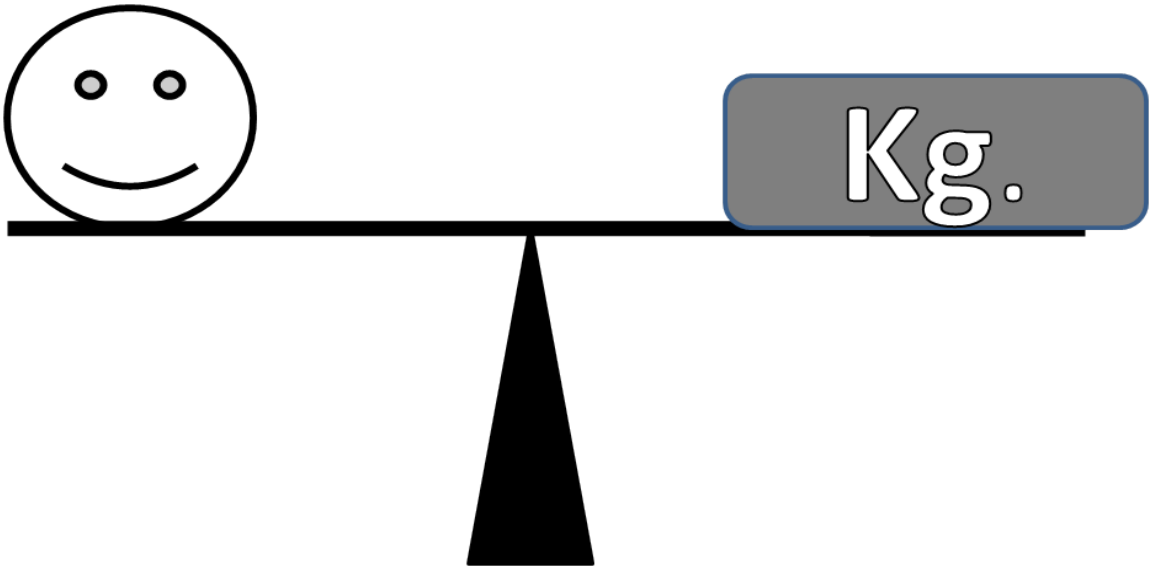
Food Security



Families and professionals often mistakenly believe that control of weight will result in unhappiness and behavioral problems. This is because efforts to limit food, if attempted without establishing FOOD SECURITY, cause increased stress in the form of doubts, hope and disappointments.



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



 www.pittsburghpartnership.com

Food Security

The measures needed to achieve FOOD SECURITY vary from individual depending on many factors including cognitive ability, age and prior experiences. See the Food Security Checklist to help you to evaluate your child's FOOD SECURITY.

Remember: "Food is STRESS"

Food Security includes:

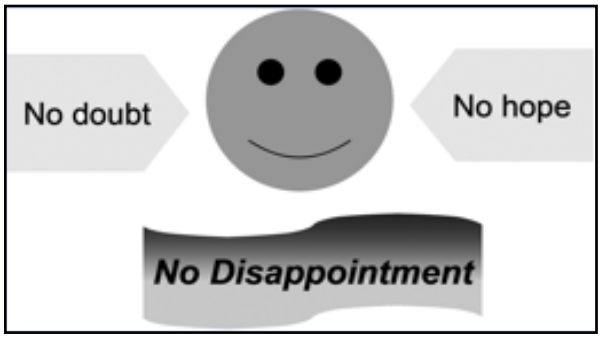
1. NO DOUBTS about what will be provided and when
2. NO HOPES of obtaining food outside the plan
3. NO DISAPPOINTMENTS concerning food

FOOD CONTROL

(locks, no access to food or money, supervision)
is the management of the *physical* environment needed to achieve no hopes and to maintain weight. FOOD SECURITY the psychologic state of the individual.
It is FOOD SECURITY that improves behavior

NO DOUBTS

- Any possible uncertainty about food must be eliminated as matches possible.:
- Advance planning meals
- A schedule of all the days events with the place of meals clearly identified
- Visual schedule
- Verbal reminders of these plans
- Food can be used carefully as an incentive* but never use food as a reward or a punishment.



Managing Expectations
prevents
DISAPPOINTMENTS,
a major source of behavior problems

NO HOPES (No opportunity/no chance)

- Food control and well-trained caregivers are essential.
- Caregivers must avoid saying anything that could create doubts for hopes about food.
- Matter-of-fact adherence to the plan in the face of manipulation, pestering and whining is an essential skill. The presence of These behaviors indicates that FOOD SECURITY is not yet achieved.

*used to motivate but can not be lost or earned: E.g. "Let's hurry up so we can get our lunch!" or "After math it will be time for your chewing gum."

 www.pittsburghpartnership.com