

Supporting Challenging Behaviors in Persons with Prader-Willi Syndrome

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ROUTINE & PREDICTABILITY

People with PWS need to receive information in a very orderly manner. There is a strong need for routine, sameness and consistency in lifestyle.

SUPPORT STRATEGIES:

- Accent routine with verbal reminders, written lists and schedules. Provide careful and simple explanations.
- Anticipate change and foreshadow with careful discussion until the change becomes part of the “new order” of thinking.

PERSEVERATION OF THOUGHT

Persons with PWS have a tendency to “get caught” on one issue or thought to a level where it overshadows the main theme of the learning or social activity. This tendency can contribute to loss of emotional control.

SUPPORT STRATEGIES:

- Carefully coax the individual to a resolution or to another topic with what ever strategy seems appropriate (empathy, leading questions, diversions)
- Avoid power struggles and ultimatums
- “Keep your cool” – even and especially voice tone and body language
- Consider psychiatric medications

TENUOUS EMOTIONAL CONTROL

Any seemingly minor combination of life stressors can contribute to loss of control as evidenced by yelling, swearing, aggression, destruction and/or self-injury. Once control is lost it is typically a period of time before control is regained. This is often followed by feelings sadness, remorse and guilt.

SUPPORT STRATEGIES:

- Be vigilant to life stressors and avoid or prepare person for the change.
- Read early signs of control loss and provide support
- Once control is lost, provide a safe area and wait it out. Afterwards, provide a “talk out.” Avoid negative consequences.

FOOD AND DIET

Persons with PWS experience a strong, physiological drive to obtain and eat food (hyperphagia) with impaired feeling of satiation after eating. This phenomenon is paired with a slowed metabolic rate that causes extraordinary weight gain. This combination of factors contributes to behavioral challenges including anger, stealing and lying.

SUPPORT STRATEGIES:

- Provide supervision at all times
- Keep food out of sight and secured in a locked pantry, refrigerator at all times.
- Serve pre-planned meals.
- Encourage daily aerobic exercise.
- Understand that behaviors such as stealing food, stealing money to purchase food, emotional outbursts, lying, etc. are a result of the physiological consequences of the syndrome.

SKIN PICKING

Picking is a common behavior. Persons with PWS engage in picking behavior as a means to manage the high level of anxiety or stress they may be feeling and/or when feeling bored. The combination of this activity with a high pain threshold can result in significant tissue damage.

SUPPORT STRATEGIES:

- Provide appropriate ways to “fidget” with the hands (e.g. tearing paper, shuffling cards)
- Provide supervision
- If picking increases view it as a symptom and work to reduce other life stressors.
- Praise and reward the absence of picking.
- Keep fingernails short, keep picking sites slippery with lotion and covered with bandages.
- Utilize rolling technique

PEER SOCIAL INTERACTIONS

Social interactions are often difficult. While persons with PWS need and value friendships, it may be an emotional strain to be exposed to the unpredictability of others for extended periods. The need for order can translate to issues of fairness. The outcome is often frustration and anger.

SUPPORT STRATEGIES:

- Employ professionals to teach social skills training
- One-to-one or small groups may be preferable.
- Time limit on social activities.
- Provide routine and structure to the activities.

SEXUALITY

Due to decreased sex hormone production, persons with PWS have underdeveloped sexual characteristics, yet the interest in sex is sometimes quite strong as well as confusing. There have been at least three documented cases of women with PWS becoming pregnant. It is still unknown whether men with PWS are capable of inducing pregnancy.

SUPPORT STRATEGIES:

- Education may be appropriate.
- Supervision may be warranted in order to avoid inappropriate sexual expression.

STAMINA

Typically, persons with PWS typically tire more quickly even when treated with growth hormone medication.

SUPPORT STRATEGIES:

- Morning is often the optimal time for productivity.
- Understand that the capacity for emotional control is diminished when fatigue is experienced.

CARETAKER STRESS ASSOCIATED WITH PROVIDING SUPPORT TO PERSONS WITH PWS

The amount of energy that is required to monitor and support persons with PWS is extraordinary. Family members are especially at risk for feelings of stress, anxiety, depression and a host of challenging feelings.

SUPPORT STRATEGIES:

- Seek out respite opportunities.
- Share parenting responsibilities with spouse, extended family members and trusted friends.
- Consider out of home support when appropriate.
- Monitor and provide support to all siblings of person with PWS.
- Seek out support groups and/or professional psychotherapy support as necessary.