

## Support and Supervision of Individuals with Prader-Willi Syndrome December 2015

**Prader-Willi Syndrome (PWS)** is a complex genetic condition that can create life threatening medical and behavioral complications for individuals with developmental disabilities. Treatments are available and are typically directed at the secondary conditions associated with this condition, but there is no cure.

Common behavioral features of PWS include food seeking and overeating (hyperphagia) due to insatiable hunger, repetitive skin picking, compulsive behaviors such as hoarding (particularly food) and arranging items, tantrums, stubbornness, mood swings, impulsivity, anxiety, and aggression. Individuals with PWS may also have many medical concerns, including sleep disturbances/sleep apnea, scoliosis, and complications associated with obesity, diabetes, and risk for heart failure.

OPWDD regulations require that each individual must have an individualized plan of services suited to his or her unique physical, medical, behavioral, and social needs. It is important for service providers to be knowledgeable about an individual with PWS and his or her specific behaviors and medical issues in order to develop an appropriate individualized support plan.

All direct support professionals and clinicians involved in the support of individuals with PWS should be trained on PWS and the specific strategies to be implemented in the person's care and support. **Staff (including new, transferred, part-time, per diem) should not be assigned responsibility for the care or support of any individual(s) with PWS until it is assured that they have been trained and understand the individualized supports to be implemented. Also, staff should not be assigned to residences, day settings, or other programs where one or more persons with PWS are served until it is assured that they have been trained and understand the environmental strategies in place to support individuals with PWS.**

A plan for support and supervision of an individual with PWS should include the following elements:

- Specific behavioral concerns associated with their genetic condition and the strategies to address them.
  - *Strategies to create an environment for appropriate access to food.*  
Individuals with PWS benefit from a food-controlled environment in ways that go beyond prevention of obesity. When the food environment is controlled by others and mealtimes and snacks are carefully structured and managed, individuals with PWS are better able to focus on interests in their lives other than food. The plan should describe these environmental strategies. Where applicable, a plan including controls on access to food, or other rights modifications, must be reviewed by a Behavior Plan/Human Rights Committee in accordance with 14 NYCRR section 633.16.

- *Specific strategies for provision and participation in alternate preferred activities* that focus the individual's attention away from food seeking and other symptoms of PWS.
- Specific medical concerns and the associated signs and symptoms of the person's medical concerns, including those associated with PWS, and strategies to monitor and address and document
  - **Identify symptoms which require immediate attention and specific actions staff must take to provide medical care.** Symptoms requiring immediate attention include abdominal distention (bloating), abdominal pain, vomiting, and generalized discomfort. (Binge eating, which may occur with lapses in safeguards, increases the risks of abdominal complications and choking.)
  - **Identify the health issues which require routine monitoring** and intervention, particularly diabetes, hypertension, sleep apnea and scoliosis. The plan should identify specific actions, including frequency that staff must perform and document to support the health of individuals with PWS. The plan should also describe symptoms that staff should monitor for and the actions to take when symptoms are observed.
  - **Develop a plan to monitor individuals and their environment to prevent overheating and hypothermia.** The body of a person with PWS cannot regulate temperature effectively. The person with PWS may not recognize that they are cold or hot. Instruction to staff on how to monitor the person's status in hot or cold weather both indoors and outdoors is needed. External hot and cold weather conditions may cause the person's body temperature to become too cold or too hot, but individuals also require monitoring in indoor environments. Rooms often become very hot or cold depending on the effectiveness and setting of the air conditioning or heating unit.
  - **Identify monitoring and actions required for psychiatric issues.** Individuals with PWS who are also diagnosed with Obsessive-Compulsive Disorder or another Mood Disorder may benefit from treatment with anti-depressant medications.

Those providing care have a responsibility to follow every detail of the care plan for the individuals they support. All Provider Agencies must recognize their responsibility to properly train and support employees to implement service/support plans safely and effectively. The need to follow individualized plans should be reinforced frequently and supervisors should demonstrate best practices. Any breakdown in following an individual's plan can create risk to the person's health and well-being.

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The Prader-Willi Alliance of NY, Inc. through their advocacy on behalf of individuals with PWS assisted in the identification of the very specific supports needed by individuals with PWS included in this "best practice" document.

Information on Prader-Willi Syndrome for this document and additional information is available from the Prader-Willi Syndrome Association (USA) and the following link:  
<http://www.pwsausa.org/publications/ER%20Treatment%20%20MA-02.pdf>