

# **Section J: Support Services for Persons with Prader- Willi Syndrome**

## SUPPORT SERVICES FOR PERSONS WITH PRADER-WILLI SYNDROME (PWS)

### PROCEDURE - Specific for Persons Served with Prader - Willi Syndrome

At specific program sites (group homes) and in the personal homes of persons served (individualized supported living) our goal is to provide treatment to meet the needs of people with Prader-Willi Syndrome (PWS), including services and assistance to lessen the impact of health problems which shorten lives, and common characteristics of the syndrome which impair community integration and self-sufficiency.

Prader-Willi syndrome is a genetic condition that affects 1 in 12,000- 15,000 live births (both sexes, all races). PWS is a complex disorder that impacts multiple physiological systems which makes diagnosis and treatment of related ailments challenging. We serve some people who do not have genetic PWS but whose disability has severely impacted their reaction to food in their environment and thus a similarly structured environment allows them to be safely supported.

#### **Health conditions common to Prader-Willi Syndrome (PWS)**

Certain conditions require extra diligence to detect, and in some cases require specialized medical care. Employees shall be provided with resources and training regarding the syndrome, and trained to observe health changes which may indicate a deteriorating or potentially harmful condition. The Community RN must be consulted if employees observe any changes in health status or have questions regarding symptoms observed during the course of their shift.

The following health conditions are common to people with PWS and require close observation:

- a) **Weight (Obesity)** - Individuals with PWS have excessive appetite due to failure of the brain to tell them when they are full. It is important to understand that this urge to eat is an actual physical disorder, overwhelming, difficult to control and requiring almost constant supervision. Individuals with a diagnosis of PWS require fewer calories due to their low muscle tone, low metabolism, and inactivity. A registered dietician will be used to create a proper diet with regular review. Weight control depends on restricting food from the individual. To date, no medication or surgeries have been found that would eliminate the need for strict dieting and supervision around food. Obtaining body weights must be done consistently and documented daily per program procedures. Weight fluctuations may indicate food foraging behavior (that may prove life threatening), fluid retention, or diuresis (excessive discharge of urine). A sudden fluctuation of plus-or-minus 5 pounds should be reported to the Community RN or Registered Dietitian for recommendations. Persistent fluctuations will be brought to the attention of the physician for further recommendations.
  
- b) **Bone fractures - or suspected bone injury** – Weak bones (osteoporosis) may occur earlier in life than usual and may cause fractures, so a proper plan of care must ensure adequate calcium, vitamin D, and weight-bearing exercise. Bone density tests are recommended. Bone breaks should be suspected following falls or other injuries. The person served will be monitored closely for changes in gait (the particular way a person moves on foot), ambulation and posturing. Injured areas will also be observed for deformities, swelling and for bruising. Since individuals with PWS frequently demonstrate increased tolerance to pain (see also d. below), this informal assessment / observation should be done even if the person denies pain or discomfort in the injured area. There have been many cases where there was no complaint of pain by the individual but visual symptoms have led us to a medical examination where a bone fracture is discovered. Contact the Community RN or physician if bone injury is suspected or if symptoms are observed. A

qualified medical professional may then be needed to assess the individual for injury.

- c) Sores, Skin lesions and Bruises - Open areas on the skin may occur due to scratches, insect bites, and/or breakdown in skin folds. Chronic skin picking behaviors, or self-injurious behaviors, keep these areas open. Open sores may be frequently present and prone to skin infections so should be treated with minor first aid or as recommended by the physician. The sores should be observed daily for signs of acute infection including pain, warmth, redness, streaking, bleeding and/or pus. If signs of infection are observed, report to the Community RN and primary care physician. Individuals with a diagnosis of PWS also tend to bruise easily. Bruises should be documented and watched for increase in size and/or change in color. If changes are observed in the bruised area, report to the Community RN. In addition to skin picking, the second most common type of self-injurious behavior is rectal digging, which may cause bleeding. With individuals who have exhibited this behavior, the support team may determine that limiting time in the bathroom is the best intervention since this is where it most often occurs. In rare instances bleeding becomes excessive, perirectal abscess occurs or the integrity of the anal sphincter is compromised. If there are signs of rectal digging, report to the Community RN and/or physician.
- d) High Pain Tolerance - Because individuals with the syndrome frequently demonstrate decreased pain sensitivity, they can experience acute medical emergencies without demonstrating typical symptoms. Lack of typical pain signals is common and may cover up the presence of infection or injury. Someone with PWS may not complain of pain until the infection is severe and may have a difficult time telling you where the pain is. In the absence of a verbal complaint of pain, other symptoms of specific injuries or illness should be evaluated. These observations include; changes in appetite (increased food foraging or reports of excess food intake or refusal of or hiding of food), long periods spent in the bathroom, excessive sleeping (change in normal sleeping pattern) and or noted change in interactions with others. Report any slight changes in condition or behavior to a medical professional and/or Community RN for investigation into the cause. Because pain complaints are rare, when an individual actually presents with a complaint of pain, symptoms should be carefully noted and immediately reported to the Community RN or physician for further instructions.
- e) Lack of Vomiting - For individuals with genetic PWS, vomiting rarely occurs. Due to the excessive eating and the possible eating of uncooked, spoiled and otherwise unhealthy food items, lack of vomiting is of particular concern. Medications used to induce vomiting may not work, and repeated doses may cause poisoning. The presence of vomiting in someone with a diagnosis of PWS may signify a life-threatening illness. All identified and reported episodes must be thoroughly investigated. If at all possible employees should view the vomit for signs of blood or other remarkable descriptors including amount, color, appearance etc. An episode of vomiting must always be followed with an examination by a physician to rule out a serious gastrointestinal problem such as gastric perforation or bowel obstruction. If a person served with genetic PWS vomits, call 911.
- f) Body Temperature – Unexplained high or low body temperatures are common for people with PWS. High body temperature may occur during minor illness and in surgical procedures requiring anesthesia. Fever may be absent even with severe infection. Body temperatures should always be taken when illness is suspected. Even minor changes in temperature may be significant. Contact the Community RN for temperatures above or below normal.
- g) Severe Stomach Illness - If left unsupervised an individual with PWS can consume life threatening amounts of food. Abdominal bloating, pain and vomiting may be signs of life-threatening stomach problems which are more common with individuals with a diagnosis of PWS than in the general population. Rather than localized pain, there may be a

general feeling of unwellness. Symptoms may include vomiting, complaints of constipation, diarrhea, stomachache, long periods of time spent in restroom, decrease or increase in appetite, or other change in usual pattern of behavior or activities. If an individual with PWS has these symptoms, contact a medical professional immediately.

- h) Breathing Problems & Sleep Apnea - Individuals with a diagnosis of PWS may be at increased risk for possible breathing problems related to low muscle tone, weak chest muscles and airway obstruction while sleeping. Sleep apnea is a sleep disorder associated with decreased oxygen saturation, irregular breathing pattern, cessation of breathing and nocturnal oxygen desaturation. Signs to watch for include sleepiness/grogginess during the day, and inability to concentrate or stay awake. Anyone with significant snoring, regardless of age, should have a medical evaluation to look for obstructive sleep apnea. Individuals identified with sleep apnea will have individualized monitoring and a treatment plan to address specific needs including the use of specialized equipment, positioning for sleep and a monitoring schedule.
- i) Diabetes Mellitus - Individuals with diabetes require careful monitoring of blood sugar levels. Individuals with diabetes will have blood glucose parameters included in their medical record. Employees should be familiar with individual parameters and actions to take if glucose levels exceed these parameters. Symptoms of a diabetic emergency include confusion, unsteady gait, faintness, rapid pulse. If an individual with history of diabetes exhibits these symptoms, their blood glucose should be checked immediately and the RN or physician should be contacted for recommendations.
- j) Other Common Concerns
- Vision concerns related to weak muscles and crossed eyes are common. Typically, annual eye exams are needed, or more often if recommended by the physician.
  - Scoliosis can occur unusually early and may be difficult to detect without x-rays. Kyphosis is a condition of over-curvature of the upper back. If symptoms are discovered the condition must be followed by a physician for potential continued medical concerns.
  - Dental problems may include soft tooth enamel, thick sticky saliva, poor teeth brushing and teeth grinding. Annual dental exams are needed, or more often if recommended by the dentist.
  - Other obesity related problems included high blood pressure, right-sided heart failure, bed sores, stasis ulcers, cellulitis, and skin problems in fat folds.
  - Nighttime enuresis—common at all ages; desmopressin acetate should be used in lower than normal doses
  - Puberty is usually delayed and incomplete. Menses are irregular and scant, and genital development is rarely complete.

In addition to the above health conditions there are psychiatric symptoms common to people with PWS that require close observation:

Psychiatric Symptoms - Secondary to the physiological aspects of the syndrome, individuals with the syndrome can experience behavior problems as a result of psychiatric issues. Typical behavior issues that tend to persist are: argumentativeness, verbal perseveration, moodiness, impulsivity, compulsivity, non-compliance, rigidity and temper “tantrums.” Other behaviors observed may include repetitive thought process, behaviors that are harmful to themselves or others, extreme aggression, hallucinations, delusional thoughts, and significant change in affect. Psychiatric and behavioral management should be provided by professionals knowledgeable about PWS. Psychoactive medications in addition to behavior management programs may be helpful. An above average

percentage of persons served with PWS may be on psychotropic medication or experience symptoms of a psychiatric illness. All observed signs and symptoms should be immediately reported to the psychiatrist and RN. In addition, employees should report any behavioral changes to the psychiatrist at the follow-up appointments.

## **ENVIRONMENTAL STRUCTURE FOR COMMUNITY LIVING SERVICES FOR PEOPLE WITH PWS**

### **PROCEDURE - Specific for Persons Served with Prader - Willi Syndrome**

For an individual with a diagnosis of PWS to succeed in living a healthy and productive quality of life, a basic level of support conducive to promoting safety and personal growth must be in place. In addition to procedures refined over 25 years of specialized services for people with PWS, Open Options has included the 2009 ***Environmental Structure for PWS Living-Best Practice Guidelines***, developed at the proceedings of the 2008 and 2009 International PWS Caregivers' Conference. (see other references at the end of this section). The preferred form of living arrangement to protect the life and improve longevity and health for people with PWS is to have a specific environment tailored to the needs manifested by the syndrome. This should include a family like environment in which each individual is able to have their own personal space while the unique support needs (specialized dietary needs, cognitive, and behavioral traits that are specific to PWS) are addressed. These traits require specialized programming and employee management skills that are not typical with services for people with other developmental disabilities. The program we provide is designed to provide this structure with some variances per individual. The typical environmental structures described in detail in this section are:

- Weight Management & Food Security
- Food Service
- Daily Routine and Structure for Mail, Money, & Personal Belongings
- Rationale for Restrictions in Individual Support Plans
- Trained Employees
- Continuum of Care Throughout the Day

#### **Weight Management & Food Security**

Weight loss improves the health status and emotional well-being of individuals with a diagnosis of PWS. The heart and lungs function better, and self-image, social acceptance, and perhaps even intellectual functioning are increased. Therefore, given the high health risks related to weight for PWS individuals, dietary compliance (including food security) and exercise shall be considered the top program priorities for services to individuals supported within this program. Prevention of obesity and/or malnutrition is one of the most lifesaving treatments in supporting individuals with a diagnosis of PWS. The following procedures support this goal:

- a) **Diet Establishment** - An individual with a diagnosis of PWS needs a lifelong diet that is individually based upon body composition, BMI and daily activity. Persons served shall be served a prescribed diet, which shall be determined by the Registered Dietician. All meals and snacks will be counted within the daily caloric limit. Each individual will have an assessment completed by a Registered Dietician (RD) administered annually and reviewed on a monthly basis. Ideal weight shall be determined by the method of determining BMI by weight in kg divided by height in inches. Due to lack of muscle mass for individuals with a diagnosis of PWS, the individual diets will be determined by a plan of weight loss being 6-8 kcal/cm in height and weight maintenance being 10-12 kcal/cm in height. Individual diets for individuals with PWS-like diagnosis will be based on most current dietary guidelines to include taking the individual's age, gender,

height, weight, and lifestyle, and portion sizes will be based on the most current USDA method for portion sizes to maintain healthy weight and body. The use of the RED-YELLOW-GREEN diet will be used to make administration of food easier for individuals with a diagnosis of PWS, their families, and employees. Meals should be managed and structured in a formal way to ensure everyone knows how meals are to be planned, monitored, and served.

- b) Food Security - In order for an individual with PWS to be successful in reaching and maintaining his or her goal weight, all unsupervised access to food must be eliminated from the environment. In home environments, all food storage areas must be kept locked at all times. Alternately or in addition to locked storage, alarms or limitations on the amount of food available at the site may be instituted. Person served access to the kitchen may be restricted to ensure the health and safety of the individual.

There will be 24-hour-day support in place, which includes direct care employee support and/or locks/alarms that provides food security and a quick response to assist with individual needs. This support should be available in both home and vocational environments. Assistance with menu planning and food preparation will be determined on an individual basis and written in their Individual Support Plans. Past behavior around food is the best predictor of future behavior and needs to be taken in account when developing an individualized plan.

- c) Psychological Benefits of Food Security - In addition to controlling intake of food, the physical security of food may assist persons served in reducing anxiety about food and the compulsive thoughts about food seeking. The Prader-Willi Syndrome Association (USA) advocates for three principles of food security - “no doubt, no hope, no disappointment” - to assist the individual with their psychological state in respect to food. When an environment is secure, individuals will experience “no doubt” by knowing their prescribed portions in advance, have knowledge of advanced meal planning, and have reliable scheduled foodservice times. Individuals will experience “no hope” by knowing they will have no opportunity to obtain additional food because of secured access to food, supervision in food accessible situations, and training of employees. Individuals will experience “no disappointment” or emotional let-down due to false anticipation or unfilled expectations about food. These principles in practice can help persons with a diagnosis of PWS to ignore food that is truly inaccessible and never potentially accessible and achieve greater peace of mind.

- d) Weight Measurement - Weight will be measured at least weekly based on individual history and current recommendations of the Registered Dietitian and/or Community RN. The following procedure will be followed for taking weights:

- All recommended weights will be taken in the morning with typical dress and no shoes.
- If additional weigh-ins are recommended they must be approved by the individual’s planning team. This may include before leaving for and/or return visits with family or friends (when Open Options PWS-trained employees are not present), and return from structured day activities where Open Options PWS-trained employees are not present.
- The scale shall be located in a room that lends itself to privacy during weigh-ins. Employees shall take reasonable measures to ensure that an individual’s weight is collected, verbally shared with the person served, and recorded on the weight record in confidence.
- Employees should remain neutral regarding any topic related to weight. Encouragement should be positive and related to overall health rather than focus on weight. If a person served has concerns about weight, caloric guidelines and changes in diet, employees should immediately defer the conversation to the dietician.

Employees should not become involved in conversations, negotiations, or debates about diet and weight control issues.

If a weight gain or loss of 5 pounds or greater within a 24-hour period is recorded it must be reported to the Registered Dietitian and Community RN for further instruction to protect the health and safety of the individual. If the individual has a weight gain/loss of 10 pounds within a seven-day period, this also must be reported to the Registered Dietitian and Community RN for further instruction to protect the health and safety of the individual.

- e) Food Outside the Home - For events in which the individual is exposed to an unsecured food setting every attempt will be made to remain within dietary guidelines. Whenever possible, when eating in a community setting, menus will be reviewed and food choices will be determined before the individual attends the event. In the case where this is not possible, plans will be made to assist the individual to make choices and reduce anxiety. In the event that persons served are attending a social gathering where food measurement is unavailable, the individual will participate with knowledge of at least one of the following to include the Division Director, Registered Dietitian, and/or Community RN. Employees will be present to encourage the individual to make good food choices and portion sizes per prescribed diets. Extra precautions will be taken to ensure the health and safety of the individual to include but not limited to extra monitoring for the following 24 hours for signs of distress or vomiting. Weights will be taken after the event for individual health and safety.
- f) Obtaining Food Outside the Diet - In the event an individual with a diagnosis of PWS obtains food that was unsecured, employees will ask the individual to return the item. If the individual does return the item there will be no further mention of the item or the event and the food will remain with them except in cases of a large quantity of food discovered in which case the Division Director must be consulted. Food obtained through foraging behavior (foraging is defined as obtaining food outside of the regular diet through covert behavior) will not be counted in the daily caloric limit because an exact determination cannot be made of the amount consumed or caloric content. There shall be no substitution or removal of food from the regular menu as a response or consequence for foraging behavior, unless specifically ordered by the Registered Dietician. In cases when we discover that food has been consumed outside the diet the person served will be monitored for signs of distress and vomiting for the following 72 hours and the Community RN will be advised of the situation.
- g) Exercise - Exercise should be a part of life routine including some levels of movement or fitness every day with the amount of time determined on an individual basis. Exercise is important to individuals with a diagnosis of PWS due to need for weight loss and maintenance. In addition to weight loss it is important for muscle strength and joint protection, cardio-respiratory efficiency, increased metabolic rate, bone density, hdl-cholesterol, treatment of co-morbidities (diabetes hypertension, obstructive sleep apnea, and interstitial edema) and enhancement of overall well-being. Recommendations for exercise will be included on the annual assessment completed by the Registered Dietitian and any physical limitations requested by the physician will be taken into account when developing the plan. The exercise should be completed in a method that increases the individual's heart rate. Methods of exercise will be determined by the planning team with individual preferences taken into account. Methods of exercise include but are not limited to: walking, riding stationary bikes, playing on a sports team, exercise videos, exercise video games, etc.

### **Food Service**

- a) Menus - Menus will be planned and posted based on diets developed as described above. Employees will make

reasonable efforts to address the preferences and desires of persons served in the menus. Written menus will be retained for review by the Registered Dietician. Substitutions must be recorded.

- b) Meal Service - Meal times should to be on a fixed schedule and all employees should worked diligently to adhere to that schedule. Food shall be served on individual plates from the kitchen. All food shall be weighed and measured in the kitchen according to the individual's meal plan. There will be no "family style" servings from the table. Food is only allowed in the kitchen and dining room. Employees will sit and dine with persons served during meal time, providing good role modeling and monitoring of choke hazards. Immediately after the meal the plates will be taken to the kitchen by persons served or employees where appropriate. The kitchen shall be cleaned by employees and leftover food shall be returned to the storage unit immediately. All edibles not saved shall be placed in the garbage disposal - only non-edible trash can be placed in the trash compactor/trash container. The Division Director shall establish other procedures as necessary to govern food service.
- c) Employees at Meals - Employees will be present at all times food is consumed by the persons served to provide feedback on eating behaviors and be available due to the increased risk of choking. Employees will be offered the same foods at the same portion sizes as the individuals in the home and at no time may employees bring any outside food or drink into the home. If an employee decides not to eat with the individuals at the site, they will need to eat before or after their shifts and away from the site. Whether eating or not, employees will sit at the dining table with the individuals in the home to ensure safety and role-model appropriate eating habits.

#### **Daily Routine and Structure for Mail, Money, & Personal Belongings**

The "best practices" described in the professional literature by physicians and psychologists are validated by the experience of families and support service providers: routine and consistency is essential for maintaining a sense of security and well-being for most people with PWS. This is accomplished through planning daily routine and special events (including the individual's ideas and preferences) and clear expectations and structure. The home where the individual with a diagnosis of PWS receives supports may also be structured with additional protocols, including the following:

- a) Daily Schedule - Some individuals in the program may benefit from having an input driven daily schedule and routine in the form that makes them most comfortable (such as appointment books, emailed schedules, picture schedules, etc.) The schedule will be provided and updated based on each individual's needed timeframe such as daily, weekly or monthly. The schedule may include meals and menu, medical appointments, activities in the community, chores and/or other activities important to the individual. If the structure and routine must be changed, as much notice as possible will be given to the person served to include the explanation of the change. If the notice of the structure and routine needs to be changed with less than 24-hour notice an explanation of the change must be given along with a replacement activity that is meaningful to the individual.
- b) Mail - When the individual receives mail at their home, it is given immediately to the individual but it is opened with the supervision of employees. The employee reviews the mail to ensure it does not have coupons or food items. If an item is located, it is taken without comment and placed in a secured area and, if appropriate, the item can be incorporated into their scheduled diet.

- c) Money – Money belonging to persons served or Open Options will be secured and maintained by employees. Each individual is given access to the balance of their personal needs account and they are able to use their finances to purchase items they desire but may not purchase food-related items outside of their prescribed diet.
- d) Personal Belongings – Persons served will have access to a lock and key to their bedroom door and will be encouraged to keep their bedroom secure when they are not within the area of their bedroom. Employees shall not search rooms based on theft accusations or suspicions without prior approval of the Division Director. Employees are expected to keep their personal belongings locked and secure at all times.
- e) Room Searches for Food - Certain circumstances raise justifiable suspicion that food has been brought into the facility and is being consumed by the individuals served, outside of their diets. Such circumstances include numerous and unexplained weight gain, food wrappers found in common areas, and confidential reports from other persons served. Since the health and safety of an individual is compromised by access to unplanned food, a room search may be completed within the following guidelines.
- The Division Director and Support Coordinator must be contacted and verbal authorization from both parties must be obtained (in the event that one of the two cannot be reached the Director of Community Living can authorize the search).
  - The room search will only be completed by the supervisor of that location with prior authorization.
  - The individual may choose to be present at the time of search unless otherwise indicated within the Individual Support Plan.
  - Any food found will be removed without comment. If the food is non-perishable it will be placed in a secured area and incorporated into the individual's diet, but if it is perishable it will be disposed.
  - An Event Report will be completed to explain the rationale and results of all room searches and submitted to the individual's Support Coordinator.

Room searches are highly intrusive and must be completed with the utmost professionalism and sensitivity. Employees serving people with Prader-Willi Syndrome have an obligation to maintain a secure environment which includes the use of the proactive methods to inhibit the presence of illicit food in the home. At the same time, the inherent degradation from a search through one's personal property must be minimized as much as possible through employees' respectful demeanor throughout the process.

Additional house rules can be developed by the individuals that live in the home. These rules will be developed and reviewed monthly in-house meetings. These rules will be written and signed by the individuals of the home and given to the Division Director for review. This will then be submitted to the Director of Community Living for review and final approval.

### **Rationale for Restrictions in Individual Support Plans**

Each person served has an Individual Support Plan per regulations, standards of care and procedure. For persons with PWS, the plan will include the program specific restrictions with reasons for the restrictions. The degree of the limitation will be based on the individual needs of each individual. While individualized, the prevalence of dangers related to food

seeking behaviors associated with PWS tell us there are no medical or behavioral interventions to relieve the need for these restrictions in some degree. The following list of restrictions may be implemented on an individual basis and with the approval of the support team:

- **Limited freedom in their home and the inability to participate in grocery shopping, menu planning, meal preparation, and food service:** Individuals with a diagnosis with PWS do not have the physiological ability to feel full. For this reason, they have the capability to literally eat themselves to death. They experience anxiety around the topic of food which can manifest itself into unproductive behaviors that could lead to harm for themselves or others. Because of both the risk of serious illness or death if an individual accessed food and ate excessively, and the corresponding anxiety around food and the food preparation area, the persons served are not allowed access to food storage and preparation areas. Meals are planned, prepared and served by employees that have had specialized training in their dietary requirements. A Registered Dietitian monitors all of the diets on a monthly basis and only foods within these dietary guidelines are allowed. Research on Prader-Willi syndrome has shown that individuals with the diagnosis actually feel less anxiety when the environment is structured in such a way that there is no “hope” to obtain additional foods or unapproved foods. The individuals we support can verbalize that they feel more comfortable and safe with us managing the food in such a manner. The individuals in the program do tell us foods they may dislike and foods they enjoy and we use that information to base our meals.
- **Dietary Requirements:** Each individual has a diet and exercise plan that is developed by a Registered Dietician who has had special training in the diagnosis of PWS. The diet is based on the dietician determining their individual body mass index and setting a goal weight based on that number. The dietician then determines a calorie based diet either on a maintenance weight or a weight loss requirement. Individuals do have minor decision-making ability in determining how their calories will be divided among a meal. Although when reviewed by others the calorie count may appear low, the calorie count is low due to unique nature of their metabolism. Individuals with a diagnosis of PWS do not have a typical working metabolic system so they do not metabolize food as the typical population. A small amount of calories above their dietary plan can cause significant weight gain. With the diet plan, the dietician also recommends an exercise plan for the individual. The individual does determine what type of exercise they would like to complete to meet this requirement.
- **Weight:** Each individual in the program is required to have his/her weight recorded at least weekly. It is also reviewed to see if needed that they are weighed before and after any outing they have without Open Options specifically trained employees. Due to their extraordinarily slow metabolism they can gain weight quickly if they have had access to food outside of their dietary plan. There is also a risk of significant illness, injury, or death if an individual was to access a large amount of food. Weights are checked to ensure that the individual is at their expected or typical weight and are not in need of medical intervention, as well as to track their progress towards their goal weight.
- **Access to money:** Individuals with a diagnosis of PWS consistently think about and have a desire to obtain food. When given access to money unsupervised, individuals with PWS often perseverate more on food and will use money to purchase food or food related items. Each individual is given access to the balance of their personal needs account and they are able to use their finances to purchase items they desire with the exception of food-related items. Employees are trained in how best to shop with individuals and to maintain a focus on appropriate items.

- **Time alone:** Due to the nature of the PWS and the perseveration on food and food-seeking behavior, individuals with this diagnosis should have supervision always. With review of an individual's history, which is the best predictor of food seeking, and completion of the "Home Alone Curriculum," home alone time can be used on an individual basis, but never where access to food could occur. Without this supervision, many people with PWS will attempt to gain food through what means they find necessary. Access to and ingestion of large amounts of food presents the risk of intestinal neuropathy (a lack of blood flow to the intestinal tract from too much food in the intestinal tract that causes the tissue to die). This in turn causes infection and disease that would potentially lead to death or severe medical consequences.
- **Opening and Receiving Mail:** As the individual's behavior is often focused on food-seeking, many means are used to try and obtain food or food-related items. For some individuals, this may include convincing family members to send money or coupons for food, or ordering food products online or via mail. Because of this risk, when the individuals in our program receive mail at their home, it is given immediately to the individual but they open the mail with the supervision of employees trained in the management of PWS. Employees review the mail to insure it does not have coupons or food items. If an item is located, it is taken without comment and placed in a secured area and they can have the item incorporated into their scheduled diet. Access to these items could cause serious and potential life threatening medical issues for the individual and unproductive behaviors which can manifest it into concerns that could lead to harm for themselves or others.
- **Inspection of belongings:** When an individual in this program has been with individuals other than those employed by Open Options who has received specialized training, their belongings are checked for food or food related items in a respectful and private manner. If an item is located, it is taken without comment and placed in a secured area and they can have the item incorporated into their scheduled diet. Access to these items could cause serious and potential life threatening medical issues for the individual and unproductive behaviors which can manifest it into concerns that could lead to harm for themselves or others.
- **Limited reading material:** Individuals in this program are restricted from reading items that relate to food. Access to these reading materials could cause unproductive behaviors which can manifest it into concerns that could lead to harm for themselves or others.
- **Limited choice of television programming:** Individuals in this program are restricted from watching programming related to food. Access to these programs could cause unproductive behaviors which can manifest it into concerns that could lead to harm for themselves or others.
- **Limited conversation:** Conversations in these homes are limited and food or food related topics are not discussed. These conversations could cause unproductive behaviors which can manifest it into concerns that could lead to harm for themselves or others.

Each individual could have additional behavioral techniques or restrictions based on their need.

Note that all rights restrictions are subject to due process and should be clearly delineated in the Individual Support Plan (See Section E. Rights of Persons Served).

### Trained Employees

Employees working with individuals with a diagnosis of PWS will be offered additional training, including but not limited to the following:

- Food Management – a training based on the dietary, food preparation, and menu planning needs to assist the individuals to remain healthy and safe.
- Rights, Responsibilities and Restrictions - a training to review the rights and responsibilities afforded to all individuals. The training will include a review of approved restrictions within this program, why these restrictions are in place and how these restrictions are determined for each individual.
- Unique Characteristics of PWS – a training in the unique characteristics of an individual with this diagnosis. This is to include hyperphagia, lying and stealing, obsessive-compulsive behaviors, self-injurious behaviors, physical appearance and impulsivity/aggression.
- Specialized medical needs and supports - a training to discuss the medical issues related to a diagnosis of PWS.
- Life in the Home - a training to make employees familiar with the needs of the specific home or homes in which they are assigned.

### **Continuum of Care Throughout Their Day**

Individuals with a diagnosis of PWS have unique communication skills and interpersonal relationships. This can cause health, safety and communication complications between those involved in the people involved in planning and treatment for the individual. Persons served should have opportunities to participate in various community activities and family activities. These opportunities also provide opportunities to forage for food which poses a danger to their health. The food seeking behaviors and their related dangers are so unusual that day program personnel and employers often underreact to the stated warnings from Open Options program employees. The following steps may be taken to assist the individual to be safe in these environments.

a) Day Program or Employment – The following strategies may be utilized or recommended

- Provide support and documentation to the sites as they request.
- Provide weekly contact with the employees to discuss any episodes of foraging for food. If any are reported, inquire as to how the food was obtained and what the team can do to assist the individual in this environment. Documentation of the contact will be maintained.
- When the individual starts a new program, daily weights will be taken before and after program attendance and documented. If there are no weight fluctuations in the first 90 days this strategy will be evaluated for continuation, and reviewed annually to document if this is necessary.
- Lunch shall be prepared and pre-packed to attend events outside of the group home. The person served will have the option to check their lunch before leaving the home to agree that all dietary guidelines had been met. Any disagreements or errors must be corrected before the lunch leaves the house.
- When the individual starts a new program in most cases the lunch boxes will be carried into the program and delivered to employees within this program. The individual will have safe guards in place per individual need.

b) Visits with family or friends without Open Options support present special safety challenges. The following check-in procedure is implemented following all activities without employees, day visits, and overnight stays outside of the program (not including day programs, school, or work unless specified in the PCP):

- Assist the individual to open bags, pockets, purses, backpacks or baggage immediately upon return from these community outings. If a food item is located, it is taken without comment and placed in a secured area and may be incorporated into their scheduled diet. Checks should be conducted in as private a manner as possible, typically one on one in a private room.
- Weigh the person served and document the weight. For weight fluctuations of 5 lbs. or greater (over their weight before departure), individuals should be monitored closely for signs of stomach and bowel distress. Significant symptoms may include vomiting, complaints of constipation, diarrhea, and long periods of time in the restroom, decrease or increase in appetite and or change in usual pattern of behavior and activities, or any complaints or gestures that would indicate pain or discomfort in abdominal or intestinal area. Vomiting and reports or observations of stomach or abdominal pain must be evaluated by a physician as soon as possible. The dietician should be consulted as to whether dietary changes need to be made when weight has increased. Contact the Registered Dietitian if weight is up or down by 5 pounds or more.
- Once the individual returns home, talk with parents or friends privately (away from the person served) if possible following a visit. This can be helpful in determining behavior and/ or opportunities provided for inappropriate food seeking.

#### c) Community Outings

When the individual participates in an activity in which food may be accessible, support and oversight must be provided to help guide choices and portion sizes. To help the individual within the community, employees must have knowledge of calories levels, dietary guidelines, food preparation and portion sizes. The following guidelines should be used (unless otherwise indicated in the Individual Support Plan):

- Whenever possible, consult resources or references prior to dining out to identify appropriate menu choices in advance. If information is not available, the Registered Dietician should be consulted.
- When the individual is dining out, guidelines need to be set before leaving. The individual and employees need to agree to all guidelines related to food choice, and acceptable portions.
- Provide the individual supervision as outlined in the Individual Support Plan; never leave the individual alone in a food setting.
- To assist the individual to be as comfortable as possible in this setting have any uneaten food removed from the area as soon as possible.

#### Reference Sources contributing to Open Options Procedures for Services for People with Prader-Willi Syndrome:

- Butler, Merlin G., Lee, Phillip D.K. Lee and Whitman, Barbara Y. Management of Prader-Willi Syndrome. 2006
- Goff, B.J. Supporting Adults with Prader-Willi Syndrome in Residential Settings An Expert's Recommendation.
- Gourash MD, Linda M. and Forster MD, Jancie L. Prader-Willi Syndrome Association (USA) presentation "Pittsburg Partnership Food, Behavior, and Beyond. Practical Management for the Child and Adult with PWS."
- Gourash MD, Linda M. and Forster MD, Jancie L. Regulation of Weight in Prader-Willi Syndrome: Theoretical and Practical Considerations. Pittsburgh Partnership. 2009.
- Soyer PhD, Hubert and Hödebeck-Stuntebeck, Norbert. Best Practice Guidelines For Standard of Care in PWS. Distributed and copyrighted by IPWSO 201
- "The Red, Yellow, Green System for Weight Control," The Children's Institute, 2008