

# The International Journal for Direct Support Professionals

## PWS: Living Beyond the Initials

By: **Melonie Sampson**

Prader Willi syndrome (PWS) is a disability that has its fair share of challenges; however, it also has its fair share of successes and opportunities as long as individuals get the support and understanding that they need. When individuals with PWS get the support they need, they can start a jewelry business, graduate high school, maintain their health and weight, find and enjoy lasting relationships, and enjoy life to the fullest.

Mike is a person with Prader Willi syndrome who has lived in multiple group homes from the age of 15. When Mike first moved to a group home, he went to live in a home with other people who did not have PWS and did not need the same supports as he. This was very difficult for Mike as he would eat food he wasn't supposed to. He was not at a healthy weight, and he felt angry and anxious when other people were having food he wasn't able to. He was worried all the time about whether or not he would be safe at activities he was participating in or if there would be food distracting him from enjoying himself and getting the most from it. Mike felt like he wasn't successful and he wasn't enjoying his life. After moving a few times between the ages of 15 and 20, Mike found himself living in a home that practices food security and had access to other people with PWS, as well as staff who understood the special supports he needs as someone with PWS. Mike is now volunteering and participating in a self-advocacy group, has completed classes towards his goal of getting a high school diploma, spends quality time with his family, enjoys summer camp for a couple of weeks every year, has a girlfriend, and is looking forward to finding employment.

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Here are some tips on supporting someone with PWS:

### 1. **Know and understand what Prader Willi syndrome is:**

PWS is a rare genetic syndrome affecting approximately 1:15,000 births; it is discovered through genetic testing via a special blood test. There are many symptoms and traits of PWS, some common symptoms for infants are known as "floppy baby syndrome" along with difficulties feeding. As a toddler, commonly between the ages of two and six years of age, unregulated appetite or hyperphagia or being hungry all the time may begin, along with the possibility of other symptoms and physical characteristics, one of the most common being obesity.

Other common behaviours like foraging or looking for food constantly, eating large amounts food, difficulty managing your emotions, becoming aggressive, and skin picking, to name a few, may also be present at different developmental stages throughout their life. People diagnosed with PWS may also have a developmental disability or a learning disability. There is no specific cause of PWS; it is caused by different malfunctions or lack of functions of chromosome 15 for which there is no cure.

It is important to recognize and understand that PWS can be a life-threatening condition. If someone with PWS listens to their minds and bodies telling them they are hungry and are able to eat all the time, they will overeat. This will lead to many medical conditions with serious consequences to their health, wellbeing, quality of life and prevent them from achieving their goals.

## **2. Know what Prader Willi syndrome isn't:**

PWS is not the 'end of the world,' although it has its challenges, it is very much manageable. It isn't a reason for the community to not support them, for people not to respect them, or their wants and needs, for them not to try and do their best, and it isn't a reason for them not to be able to go into the community and have meaningful and valuable experiences.

## **3. Food security or food safety:**

Helping to create an environment that is free of food or food triggers can help to increase the enjoyment of day-to-day life and activities. Out of sight, out of mind. When the environment is food safe, individuals with PWS have less stress, less anxiety and less to worry about while they enjoy meaningful activities and working on their goals. Food security is a support strategy that helps them focus their energy on activities rather than on acquiring food or trying their hardest not to take the food despite their brain and body telling them to. Trusting you have food safety or security and that the environment and the people around you are going to be supportive and respectful when it comes to food, makes individuals feel comfortable, accepted and able to be themselves without any added challenges.

At home, prevent food from sitting around the house, like fruit baskets or sweets, being left out on the countertops in the kitchen, and even put compost food waste bins in a cupboard that is locked. Avoid pictures of food as decorations and have a ventilation system to help remove the smell of food during and after cooking. Lock the pantry or cupboards where dry food items are stored, as well as the fridge. While locking food up may seem extreme, people with PWS often regard this as supportive rather than restrictive. In the same vein, support staff, as well as all who love and support someone with PWS, need to enjoy their treats and snacks in private, not around someone who is unable to enjoy them with you.

## **4. Menu Planning:**

Work with a dietician and doctor to develop a healthy appetizing menu that meets all of the individual nutritional needs, including three healthy meals and two to three healthy snacks. Spread them throughout the day from the time you wake up, remembering to leave time for digestion before going to bed. Planning for two or three weeks at a time

can help prevent the stress of trying to plan something at the last minute and will help with grocery shopping. Using a system like the Red Yellow Green Nutritional Guide as recommended by the Ontario Prader Willi Syndrome Association, will make planning and decisions easy to understand for the supporters and the individuals and, when changes are needed, will provide advice on the best alternatives. Remember to always include lots of water, as individuals with PWS don't often enjoy drinking water. There are ways to be creative and make water more appetizing without adding sugar or calories, such as using a water diffuser to add fruit or cucumber slices for flavour, adding lemon or sugar-free fruit drinks and even decaffeinated coffee or flavoured tea; just remember – do not add sugar, milk or cream.

#### **5. Community access:**

Plan community activities or events that do not specifically involve food, such as going to the park, museum, library, bowling, etc. This is not to say there will not be food triggers or distractions at these activities, but being clear about the purpose of the activity will help with focus and enjoyment.

When planning an activity that involves food, such as going to a restaurant – plan, plan, plan. Choose the restaurant and look up the menu, nutritional values, calories and ingredients online. Plan the individual's menu in advance, write it down, provide a copy to them, and encourage them to bring it with them as a reminder for themselves and for those supporting them in what is going to be a stressful environment. Discuss and plan for how they will handle anxiety and distractions, like when the server offers them items not on their menu or items that were not ordered, such as free refills, larger servings or additional side dishes. Plan on leaving the restaurant once the meal is finished and not 'hanging around' where there are likely going to be many food triggers that will increase anxiety and stress levels the longer they stay there.

#### **6. Teach Coping Strategies:**

Coping strategies can help individuals manage in situations where their needs are not being met. As a person with PWS, it is crucial to learn that sometimes it is important to not listen to their body, especially when it comes to hunger. Just because there is food, and they may feel like they need to eat it, doesn't mean that they should. Some other skills they can learn and practice when it is difficult are walking away, asking for help, distracting yourself and, if possible, leave the situation. Additional helpful skills are learning some breathing and mindfulness exercises for when they are feeling anxious, and planning ahead so they know what to expect and are not surprised or overwhelmed.

It is also important to be connected to other people with PWS, who understand what they and their families are going through. It is useful to be connected to organizations and services that understand PWS and are able to provide supports that are specific to PWS. Being around people who understand them, their challenges and who need the same type of supports as them will be comforting, and they will be less likely to feel as though they are standing out from the crowd, and more as if they really belong.

## **7. Use smaller plates and cups:**

'We eat with our eyes.' Using smaller plates/divided plates and multiple cups for water, coffee and sugar free drinks will be pleasing to the eye and help make them feel as though they are eating more than they really are by making it look like more than it is. Spread the food out and add extra nonfat and nutritious foods like pickles, vegetables and salad, but beware the dressing – do not add extra salad dressing or thick sugary condiments like ketchup.

Another strategy is to try to avoid mixed foods like sauces and chili's or sandwiches but when possible, serve all the individual components of the food separately as individual items, again making it seem like it is much more than it is. For example, you can have one sandwich OR two slices of bread, 3 slices of meat, one slice of low fat cheese, lettuce, two slices of tomatoes and mustard.

## **8. Have an enjoyable and active schedule:**

Having a daily routine that includes meal times, exercise and activities will help individuals focus their energy and thoughts on things other than food. Be sure to include all the things they like to do such as, bowling, movies, work, school, games, or visiting with friends and family. Be sure to include some of the more challenging or non-preferred activities such as daily exercise, medical appointments and weight monitoring, mixing them in amongst opportunities to do their favourite activities to make them easier to follow through with and to handle. Ensure they have activities they really enjoy doing and create opportunities for them to do them as often as they can. Find creative and exciting ways to exercise so that it doesn't feel like a chore or become boring.

Downplay the meal times and food involvement in their daily routine and activities. Remind and encourage them to focus on participating and enjoying the activity and the people who are doing it with them – friends, family or supporters. Discuss with them and let them know that food or eating alone does not mean that they're having fun. Spending time with the people you care about, laughing, sharing, supporting and having experiences with is having fun. Discuss that, although sometimes there is food involved, they need to try to realize and remember that it is not the most important part.

## **9. Be consistent:**

Follow the plan and schedule as it has been outlined, don't make changes. Structure, knowing what to expect and trusting in this knowledge helps to reduce anxiety and fear. When changes are needed, do your best to make them as far in advance as possible, discuss the change in detail with the individual and come up with a new plan. Make sure to clarify what is going to happen, how or if it will affect the menu, how long this new plan will last and clarify that, once this change is over, things will go back to the old schedule and routine. When making long term changes, make them slowly, one change at a time, ensuring that the change was successful and taking time to adjust before making the next one. Focus on the benefit and success that change will bring about, not the reason for the change, especially if it may not be a positive reason such as a menu change because of weight gain or other medical needs.

### **10. Be inclusive:**

'Nothing about me without me.' Make sure to include the person in all planning. Feeling like they have some control over their life makes them happier to participate in it. There are many things about PWS that they cannot control and this can be frustrating. Being sure to include them with the things that can be controlled will help them feel happier and confident.

Help create and find opportunities to be included in events with family, friends and especially with other people with PWS who understand and appreciate what they are going through (and where there is not likely to be a lot of food around).

Include the individual's family, friends and important relationships by teaching them about PWS and the type of supports that are important for success. This is important so that they will be able to recognize and understand when and how the individual may need them to support them, and by helping avoid difficult and maybe even embarrassing situations before they happen. Talk about approaches and strategies that are helpful to the individual. Do they like to be told things in private? Does using humour help when approaching them? Do they appreciate reminders? Do they use a calendar to track their day? Do they need time to think about their choices?

### **11. Never underestimate:**

Never underestimate the abilities of someone with PWS. The hand is faster than the eye. Opportunities for food are around us all of the time, even if you are working hard at avoiding them. If there is food to be had, then they will have it, even from places that may seem to the rest of us as unappetizing, unpleasant, and maybe even embarrassing. On the flip side, with a little ongoing support, they will achieve their goals and maintain a healthy weight.

### **12. Appreciate and praise success:**

Celebrate every success no matter how small. If one pound is lost – celebrate. If one extra lap is done – celebrate. Use positive praise and feedback as reinforcement. Do not focus on the lack of success. If a pound isn't lost, don't worry about it, celebrate that none were gained. If an extra lap isn't done, just encourage them to try, try again next time. Never use food to celebrate successes and never take away food as punishment for lack of successes. Appreciate how difficult it is to be part of a community that focuses so much of its time and energy on food, when you are not able to part of it.

Prader Willi Syndrome is a disability which comes with very specific support needs. However, with emotional and physical supports to meet those needs, understanding from friends, family and staff, as well as opportunities to grow and develop, they are able to flourish. It's very possible for PWS to stand for Person Who Succeeds. I know it's true, because I've seen it over and over again.

## Resources:

Ontario Prader Willi Syndrome Association, [www.OPWSA.com](http://www.OPWSA.com)

Foundation for Prader Willi Research Canada, [www.FPWR.ca](http://www.FPWR.ca)

The Children's Institute, Pittsburg, [www.amazingkids.org/pws](http://www.amazingkids.org/pws)

## About the author

Melonie has been working in the Developmental Services field for 20 years, and has been dedicated to supporting people with PWS in a treatment home at Vita for the past eight years and, more recently, in the community as a volunteer for the Ontario Prader Willi Syndrome Association Board of Directors. Melonie has a passion for working with people with disabilities and especially prides herself in working with people with rare and/or difficult to understand diagnosis, as well as their families, friends, staff and communities.

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