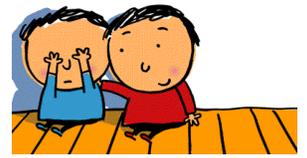




## Caring for the Child with Prader-Willi Syndrome Information for Respite and Child Care Providers



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Prader-Willi Syndrome (PWS) is a complex developmental disability that causes life long challenges. Children are all individuals and are affected in varying degrees. The following is a handout meant to help respite and child care professionals gain a quick understanding of some of the characteristics that many of these young people have in common, as well as some possible solutions. The best strategies however are those that parents will or should share, that are known to be helpful and successful for the person that will receive respite services. It is important to make sure to ask questions and gain an understanding of all aspects of care. Respite and child care providers are a valuable part of the support team that helps parents and families.

<b>Common Characteristics Seen in Children with Prader-Willi Syndrome (PWS)</b>	<b>Possible Solutions and Strategies</b>
<p><b><u>POOR MUSCLE TONE</u></b></p> <p><b>Excessive Sleepiness</b> Babies with PWS exhibit excessive sleepiness often sleeping for 20-23 hours a day. Parents are not able to feed on demand because the baby often will not cry or communicate hunger.</p>	<p><b>For EXCESSIVE SLEEPINESS</b></p> <ul style="list-style-type: none"> <li>• Provide various forms of stimulation. DO NOT allow the child to sleep all day.</li> <li>1. Awaken baby for feedings.</li> <li>2. Provide physical contact.</li> <li>3. Play/talk with baby.</li> <li>4. Provide bright, colorful surroundings.</li> <li>5. Reposition baby.</li> <li>6. Play music.</li> </ul>
<p><b>Developmental Milestone Delays</b> Due to poor muscle tone, young children with PWS are delayed in their growth and developmental milestones. Even though they proceed in the same order as other children, events such as sitting up, standing, and walking come later. In later years, this also impacts many social/recreational milestones such as riding a bike, roller skating, etc.</p>	<p><b>For DEVELOPMENTAL DELAYS</b></p> <ul style="list-style-type: none"> <li>• Ask parents about exercises, activities and toys that will help strengthen and improve muscle tone and motor development.</li> <li>• Make sure to find out what the child is able to do or not do.</li> </ul>
<p><b>Fine Motor Delays</b> As the child with PWS grows older, many are faced with weaknesses and challenges in their fine motor skills. Daily living skills as well as hand strength activities (cutting, writing) may be affected.</p> <p><b>Gross Motor Delays</b> Children with PWS typically have generalized low muscle tone. This contributes to their delay in many developmental milestones. It also accounts for lifelong challenges with large muscle weakness and coordination problems.</p>	<p><b>For MOTOR DELAYS</b></p> <ul style="list-style-type: none"> <li>• Encourage and assist as needed with fine motor activities such as cutting, picking up small objects, coloring and writing activities.</li> <li>• Encourage and assist with daily living skills — tying shoes, cutting food.</li> <li>• Play with games and other things that may help strengthen fingers and hands – play dough.</li> <li>• Engage in play with the young child that encourages use of large muscles – running, swimming, playing on playground equipment, throwing a ball ...</li> <li>• Make it fun.</li> </ul>
<p><b><u>ALTERED NUTRITIONAL NEEDS</u></b></p> <p><b>Feeding Difficulties</b> The number one concern for infants who have PWS during their first 6 months of life is <b>feeding difficulties</b>. Most of these babies have a poor suck with an inability to cry when needing to be fed. They also experience poor weight gain. This is best known as <b>the Failure to Thrive stage</b>.</p>	<p><b>For FEEDING DELAYS</b></p> <ul style="list-style-type: none"> <li>• Follow the instructions of parents on timing, type of formula/foods and use of any special feeding techniques. Some may require feedings through a tube in their stomach. Others may need/use special nipples and/or bottles.</li> <li>• Cuddle and hold the baby after feeding.</li> </ul>

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<p><b><u>ALTERED NUTRITIONAL NEEDS cont.</u></b>  <b>Weight Gain, Preoccupation with Food and Food Seeking</b>            Around the age of 2, children with PWS may start to experience weight gain with a noticeably greater interest in food. In some young children it proceeds quickly to food seeking and stealing. Others move at a slower pace with the desire to seek out food.            As they grow older, the craving for food typically increases. It becomes a life long struggle, and it does not go away. This situation is compounded in the fact that food is metabolized at a very low rate, and this can cause extraordinary weight gain very quickly if food and diet are not monitored. We typically call this the <b>Thriving Too Well stage</b>.</p>	<p><b>For WEIGHT GAIN, FOOD PREOCCUPATION, AND/OR FOOD SEEKING:</b></p> <ul style="list-style-type: none"> <li>• It is important to receive and follow instructions on when and what food the child can eat. After age 4-5, most require a calorie-restricted diet.</li> <li>• Many do best with structured meal/snack times.</li> <li>• Do not use food as a reward.</li> <li>• Avoid situations where food may be present that the child is unable to eat.</li> <li>• Don't leave child alone with food. Be aware of all food sources including purses and/or money or food in cars or vehicles.</li> <li>• Always supervise around all food related areas — vending machines, stores.</li> <li>• Discuss any delays in snack or lunch ahead of time; this may occur with an outing or unplanned event.</li> <li>• PRAISE situations where the child did well with handling a food situation.</li> <li>• Be aware that parents often have to lock their refrigerator and/or cabinets. Some use alarms to prevent the child or young adult from accessing food. Make sure to know where keys are located; how to deactivate an alarm or what the combination for the locks may be.</li> <li>• Know what to do if the child or young adult with PWS steals food.</li> <li>• Keep purses or backpacks out of sight and if needed locked up. Some may not only steal food, but they may steal money to buy food.</li> <li>• Realize that food and food-related activities often become irrational topics and issues.</li> <li>• Preplan all outings, making sure that all snacks and meals are discussed and agreed upon ahead of time.</li> </ul>
<p><b><u>SPEECH AND LANGUAGE</u></b></p> <p><b>Expressive and Receptive Language Difficulties</b>            PWS can impede all aspects of speech and language development. Poor muscle tone and incoordination of facial and oral muscles can affect a child's ability to make sounds and form words. When cognition/learning ability is delayed or impaired, both expressive and receptive language may be affected. Typically, children with PWS have stronger receptive skills – they can understand more than what they can say.</p> <p><b>Neuromotor and Articulation Problems</b>            Poor muscle tone as well cognitive limitations can cause articulation problems as well as a slow rate of speech and poor voice quality. A decreased ability to communicate may contribute to increased frustration in the child with PWS.</p>	<p><b>For SPEECH AND LANGUAGE</b></p> <ul style="list-style-type: none"> <li>• Don't be afraid to talk, sing and read to the child. Any way to model appropriate speech is helpful.</li> <li>• Learn if and what assistive techniques are being used to help the child express his/her needs and wishes. Some may use sign language, picture/symbol boards. These are typically used more with the very young to decrease frustration.</li> <li>• Be a role model by providing assistance in learning socially appropriate communication behaviors — eye contact, initiating conversations, good manners ...</li> </ul>

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<p><b><u>LEARNING DIFFERENCES</u></b></p> <p><b>Cognitive Disabilities or Learning Disabilities</b>  Approximately 60% of persons with PWS have a cognitive disability; 40% have learning disabilities. You will see quite a range in IQs with most functioning in the borderline area. They are however very capable of learning and love to please those who teach them.</p> <p>Some of the <b><u>LEARNING CHALLENGES</u></b> these young people may face include:</p> <p><b>1. POOR SHORT TERM MEMORY</b>  Many young people have problems with short-term memory processing — especially when the information is given verbally. This is often misunderstood as ignoring or disobedience because many times the child or young adult has been successful at understanding a "piece" of the information that is given to him/her.</p>	<p><b>For SHORT TERM MEMORY PROBLEMS</b></p> <ul style="list-style-type: none"> <li>• Be patient and understanding. Keep things simple.</li> <li>• Use and teach strategies to increase short term memory: <ol style="list-style-type: none"> <li>1. Verbal rehearsal – practice what to say ahead of time.</li> <li>2. Write things down.</li> <li>3. Use visuals — pictures.</li> </ol> </li> </ul>
<p><b>2. SEQUENTIAL PROCESSING DEFICIT - DIFFICULTY UNDERSTANDING ABSTRACT CONCEPTS</b></p> <p>Many young people with PWS have a deficiency in the <b>INTAKE, PROCESSING AND RESPONSE</b> of information. When someone has a sequential processing deficit, he/she will not learn from "experience". The timing and order may get confused. The "if - then" reasoning is absent or it gets mixed up. This is a key frustration factor for parents and care givers.</p> <p>Other areas that may be impacted by this include:</p> <ul style="list-style-type: none"> <li>▪ Trouble interpreting and executing multi-step directions</li> <li>▪ Difficulty summarizing and restating events</li> <li>▪ Difficulty understanding time frames. ‘Later’, ‘soon’, or ‘tomorrow’ can be difficult to understand.</li> </ul>	<p><b>For SEQUENTIAL PROCESSING DEFICIT AND DIFFICULTY WITH ABSTRACT CONCEPTS</b></p> <ul style="list-style-type: none"> <li>• Use visual representation of time — calendars, clocks.</li> <li>• Keep instructions/steps limited to 2-3 steps.</li> <li>• Plan ahead.</li> <li>• Minimize or remove situations or environmental factors that provoke behaviors (especially food).</li> <li>• Emphasize APPROPRIATE behavior that you DO want to see. Stay away from the word “don’t”.</li> <li>• Use time frames that the child/young adult understands – “We will go after we eat lunch” ... or “after you make your bed.”</li> </ul>
<p><b>3. PROBLEM SOLVING DIFFICULTIES</b></p> <p>Many young people with PWS often have difficulty planning or ordering steps to problem solve. They tend to be "Black and White" thinkers. They have difficulty using deductive reasoning.</p>	<p><b>For PROBLEM SOLVING DIFFICULTIES</b></p> <ul style="list-style-type: none"> <li>• Rehearse appropriate behavior expectations ahead of time.</li> <li>• Take advantage of pointing out learning opportunities as they arise in the community.</li> <li>• Have simple, clear WRITTEN rules and expectations.</li> </ul>
<p>Some of the <b><u>LEARNING STRENGTHS</u></b> often seen in students with PWS include:</p> <ul style="list-style-type: none"> <li>▪ <b>GOOD LONG TERM MEMORY SKILLS AND GOOD ROTE SKILLS</b> — Once the child has learned something, they typically remember it for a long time.</li> <li>▪ <b>MULTI-SENSORY LEARNERS</b> – Tend to be <b>VISUAL</b> learners as well as <b>HANDS ON</b> learners.</li> <li>▪ <b>STRENGTH IN READING</b> - May do better with books and tapes.</li> </ul>	

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<p><b><u>BEHAVIOR</u></b>  <b>Rigid Thought Process</b>  Many young people with PWS often need to receive and store information in a very orderly manner. There is a strong need for routine, sameness, and consistency in their environment.</p>	<p><b>For RIGID THOUGHT PROCESS</b></p> <ul style="list-style-type: none"> <li>• Prepare for changes and allow for discussion.</li> <li>• Use visuals; put things in writing — lists, schedules.</li> <li>• Don't make promises you can't keep.</li> <li>• Give warning of ending (“Ten more minutes”).</li> <li>• Breakdown procedures into concise, orderly steps. Limit steps to 3 at one time.</li> <li>• Try to keep the routine the same. Don't feel like you should change things.</li> </ul>
<p><b>Perseverative Thinking</b>  This is the tendency to get "caught" on one issue or thought to the point where it overshadows their ability to move on. (S)he may repeat and repeat a word, phrase or thought over and over again. Perseveration can lead to loss of emotional control.</p>	<p><b>For PERSEVERATION</b></p> <ul style="list-style-type: none"> <li>• Have the child restate what you said.</li> <li>• Set limits. “I'll tell you one more time, then we have to move on”.</li> <li>• Put in writing; use visuals.</li> <li>• Avoid power struggles and ultimatums.</li> <li>• Use diversion. Change to another topic you know is of interest to the child/young adult.</li> <li>• Ignore (if possible).</li> </ul>
<p><b>Tenuous Emotional Control</b>  Any combination of life stressors can lead to emotional "discontrol" as evidenced by tantrums or rages — yelling, swearing, aggression, destruction, and self-injury. Recovery of control takes time and is often followed by sadness, remorse, and guilt.</p>	<p><b>For TENUOUS EMOTIONAL CONTROL</b></p> <ul style="list-style-type: none"> <li>• Ask parents what things may cause the child to become upset and avoid those triggers if possible. Make sure to find out what helps to calm him/her.</li> <li>• Clear written instructions are often helpful – especially when it comes to food. If needed, have parents write down what food child/young adult can have.</li> <li>• Provide positive attention and praise when person is maintaining control, especially in difficult situations.</li> <li>• Prevent loss of control by encouraging communication and acknowledging feelings.</li> <li>• Anticipate build up of frustrations and help him/her to remove self to “safe area” where he/she can share feelings.</li> <li>• Don't try reasoning during times when out of control.</li> <li>• Have a plan in place if the child/young adult becomes more violent. Stay calm. Limit verbal interaction..</li> <li>• Have contact information and instructions of what to do if things escalate too much. Carry a cell phone.</li> <li>• Provide positive closure. Don't hold a grudge. Remember – it is the Prader-Willi – not the child who can have these explosive times. Don't take it personally.</li> </ul>
<p><b>Elopement – Running Away</b>  Many people who have PWS have been known to elope or run away. This is often seen when they are very upset. It is also seen when they do not appear to be overtly upset about something. At times, these individuals have been found sneaking away to get food - going into a neighbor's house, going to a nearby convenience store or even sneaking food from garbage dumpsters. In some individuals, it is seen as an attention-seeking behavior. This sort of behavior can be very upsetting and presents a very big health and safety risk for those with PWS.</p>	<p><b>For ELOPEMENT – RUNNING AWAY</b></p> <ul style="list-style-type: none"> <li>• Listen and evaluate why the child may be running and try to address those issues.</li> <li>• Teach them appropriate problem solving strategies. “If you are angry at your friend, you should ...”</li> <li>• If a person with PWS has a pattern of running away, (s)he may require close supervision.</li> <li>• Discuss how this behavior can put him/her into an unsafe situation – being taken by stranger etc.</li> <li>• Foreshadow behavior expectations in a positive manner – “We care about you and want you to be safe so it is important to stay with the group” (Don't say – “Don't run away”).</li> <li>• Know what to do; who to call ... if this behavior occurs.</li> </ul>

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<p><b><u>SOCIAL</u></b>  <b>Difficulty with Peer Interactions</b>            While children want and need other children and value friends, it may be difficult for them to be exposed to the unpredictability of others for long periods. The need for order frequently translates into fairness issues and comparing themselves to others, often resulting in anger.</p>	<p><b>For DIFFICULTY WITH PEER INTERACTIONS</b></p> <ul style="list-style-type: none"> <li>• Often times the respite worker is a main source of providing social opportunities.</li> <li>• Preplan outings. Don't be afraid of "specializing". Be the "movie respite worker". Establish a routine with the outing so that it is a successful experience.</li> <li>• Include child in planning activities that are of interest to him/her (board games, puzzles, and computer games).</li> <li>• Be a role model in demonstrating social issues that are often problematic – sharing, taking turns, losing.</li> <li>• Stay away from crowds until you have a good relationship. These can often be over-stimulating and result in a behavior upset. You may have less control with food &amp; other issues.</li> </ul>
<p><b><u>HEALTH RELATED CONCERNS:</u></b> **Keep a health brochure with you at all times in case you need to seek medical care. Many health care providers need educating and reminding of the health issues for those with PWS. Make sure you know the child's allergies, current medications, physician and hospital preference.</p>	
<p><b>High Pain Threshold</b>            Although the exact mechanism is unknown, persons with PWS have an altered response to pain. Many individuals will not complain of pain until an infection or injury has become very severe. Close assessment of pain along with listening to caregivers concerns is very important.</p>	<p><b>For HIGH PAIN THRESHOLD</b></p> <ul style="list-style-type: none"> <li>• Assess all complaints of pain. Look for redness or swelling. Do not rely totally on pain or lack of to determine if an injury or illness is severe.</li> <li>• Make sure to notify parents if injury occurs or if the child is complaining of pain.</li> <li>• Find out from parents what the child's typical behavior is when ill. Sometimes illness is suspected when there is a change in behavior, activity level and appetite (however — appetite is rarely affected).</li> </ul>
<p><b>Medication Sensitivity</b>            People with PWS may have unusual reactions to standard dosages of medications. They seem to be more sensitive to medicines that can cause sedation.</p>	<p><b>For MEDICATION SENSITIVITY</b></p> <ul style="list-style-type: none"> <li>• Have clear written instructions and an understanding of all medicines you may be asked to give.</li> <li>• Do NOT give any medicines without parent permission.</li> </ul>
<p><b>Gastric Illness</b>            Severe stomach illness has been noted in persons with PWS – especially those who have had a binge eating episode.</p> <p><b>Symptoms:</b> abdominal bloating, vomiting, pain may or may not be present, general feeling of not feeling well. It is rare for a person with PWS to vomit.</p>	<p><b>For GASTRIC ILLNESS</b></p> <p>ALL Cases of vomiting should be reported to the parent. A serious life threatening situation could be occurring. The person with PWS may need to be emergently evaluated by a health care professional.</p> <ul style="list-style-type: none"> <li>• <b>Encourage the child to share honestly if they have had a binge episode. (S)he should not be punished if this has occurred.</b></li> </ul>
<p><b>Lack of Vomiting/Choking</b>            About 2/3 of persons, who have PWS, lack the ability to vomit. This has special implications in the treatment of poisoning. Most will not respond to the use of medicines that induce vomiting and treatment must focus on having their stomach pumped. Many children and young adults eat very fast and do not chew and swallow adequately and slowly. They are at risk for choking.</p>	<p><b>For LACK OF VOMITING/CHOKING</b></p> <ul style="list-style-type: none"> <li>• In the case of poisoning, contact parents immediately and seek their advice on what to do further.</li> <li>• Make sure you know the proper procedure for handling choking for the age child/young adult you are working with.</li> <li>• ALL Cases of vomiting should be reported to the parent. A serious life threatening situation could be occurring.</li> </ul>
<p><b>Body Temperature Abnormalities</b>            Both very high and very low body temperatures have been reported in persons with PWS. This can happen quickly due to illness or environmental temperatures. The body doesn't always regulate itself properly and/or the person with PWS may not use good choices in selection of appropriate clothes. Fever may be absent in spite of infection.</p>	<p><b>For BODY TEMPERATURE ABNORMALITIES</b></p> <ul style="list-style-type: none"> <li>• If person with PWS seems ill and fever is not present, make sure to alert parent or family member.</li> <li>• Discuss appropriate clothing and outside temperature before it is time to wear the clothing.</li> <li>• Limit time out in the heat or in the cold.</li> <li>• Use cooling or heating strategies as needed. – cool water, air conditioning, blankets, heater ...</li> </ul>

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<p><b>HEALTH CONCERNS cont.</b>  <b>Skin Picking – Lesions, Bruises</b>            It is very common for people with PWS to pick at their skin and develop open sores. It may be worse during times of stress &amp;/or boredom. Combined with the higher pain threshold, these behaviors can result in severe open sores if not controlled. Many of these individuals bruise easily. The appearance of open sores and bruises may wrongly lead to the suspicion of abuse.</p>	<p><b>For SKIN PICKING – LESIONS, BURISES</b></p> <ul style="list-style-type: none"> <li>• Provide activities to <u>keep hands busy</u> (coloring, computer time, play dough, hand-held games).</li> <li>• Keep nails short. Apply lotion liberally. This can also be a form of diversion.</li> <li>• Apply mosquito repellent before any walks or outside activity.</li> <li>• Teach and encourage self-care if bleeding from sores is present. Keep gloves handy along with Band-Aids.</li> <li>• Report all incidents where bruising occurs.</li> </ul>
<p><b>Poor Stamina</b>            Children and young adults with PWS tire more easily and may fall asleep during the day. Morning is typically their best time when energy level is highest.</p>	<p><b>For POOR STAMINA</b></p> <ul style="list-style-type: none"> <li>• Be aware that these young people tire easily. It is good to get them up and moving. Send on errand. Take a walk.</li> <li>• Find out if they require a scheduled rest time.</li> <li>• Don't schedule too much. A tired child/young adult is at risk of more behavior problems.</li> </ul>
<p><b>Respiratory Concerns</b>            Many people with PWS may be at increased risk for respiratory difficulties. Low muscle tone, weak chest muscles and sleep apnea (times when a person stops breathing while sleeping) are often seen in many of these young people. Daytime sleepiness and sleep-disordered breathing is common.</p>	<p><b>For RESPIRATORY CONCERNS</b></p> <ul style="list-style-type: none"> <li>• When a person with PWS gets a cold or upper respiratory infection, it is important to get them up and moving and encourage fluids.</li> <li>• Some may use a device called CPAP (Continuous Positive Airway Pressure) while they sleep. Obtain instructions if used.</li> </ul>
<p><b>Dental Problems</b>            Many children and adults with PWS experience dental problems including soft tooth enamel, thick sticky saliva, poor oral hygiene, teeth grinding and rumination (the act of "burping" up food and swallowing it again). It is common to see white crusting on the sides of their mouth.</p>	<p><b>For DENTAL PROBLEMS</b></p> <ul style="list-style-type: none"> <li>• Encourage good oral hygiene at least twice a day.</li> <li>• Report rumination – it could be a sign of gastric reflux problems.</li> <li>• Limit soda pop, fruit juices and other high acid foods.</li> </ul>
<p><b>Diabetes and Scoliosis</b>            Young people with PWS are at greater risk of developing diabetes especially if their diet and weight are not under control. Many may also be at risk for the development of scoliosis.</p>	<p><b>For DIABETES AND SCOLIOSIS</b></p> <ul style="list-style-type: none"> <li>• Make sure to follow the diet and eating instructions that the parents have outlined.</li> <li>• If the person needs to have their blood sugar level checked, make sure to receive instructions on how to use the blood glucose monitoring device. Many young adults can do this independently.</li> <li>• Know what the signs are for low blood sugar – especially behaviors specific to that individual. Know what to do in case of a low blood sugar. It is often advisable to have a source of sugar (out of the person's sight and knowledge).            Follow the treatment plan if a brace must be worn for scoliosis.</li> </ul>

**This is just a short synopsis of some of the considerations in providing respite or child care for the child or young person with PWS. For further information on Prader-Willi Syndrome, please feel free to contact:**



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