

## Health Concerns and the Student with Prader-Willi Syndrome Information for School Staff



Students with Prader-Willi syndrome (PWS) experience some unique health issues. It is important for school staff to be aware of these to ensure that the individual has a safe, healthy educational experience. Health concerns along with their corresponding strategies are summarized below.

Health Concerns	Strategies
<p><b>Altered Pain Threshold – Decreased Pain Sensitivity/High Pain Threshold</b></p> <ul style="list-style-type: none"> <li>▪ Pain may be diminished or absent - even in severe injuries.</li> <li>▪ Fatigue or irritability may be a sign of illness.</li> <li>▪ Increased bruising and swelling is common.</li> </ul>	<ul style="list-style-type: none"> <li>▪ All injuries need to be assessed by an adult.</li> <li>▪ Report all injuries or changes in behavior to the parent or caregiver.</li> <li>▪ Person may require examination by a physician to rule out fracture or other health problem.</li> <li>▪ Apply ice to injuries as needed.</li> </ul>
<p><b>Altered Temperature Regulation</b></p> <ul style="list-style-type: none"> <li>▪ Common to see unexplained high and low temperatures.</li> <li>▪ Little or no fever may be present with illness.</li> <li>▪ Often experience low tolerance to high or low outside temperatures.</li> </ul>	<ul style="list-style-type: none"> <li>▪ Help to ensure student does not overheat.</li> <li>▪ If extreme redness of the face and sweating is noted, move student to a cool area and encourage cool water (flavored water) and/or cooling measures.</li> <li>▪ In colder climates make sure student is appropriately dressed and does not spend a long time outside.</li> <li>▪ If illness is suspected, notify parent. Fever may not be present.</li> </ul>
<p><b>Insatiable Appetite and Low Metabolism</b></p> <ul style="list-style-type: none"> <li>▪ PWS hyperphagia is a life-threatening, uncontrollable genetic drive to eat that is not satiated regardless of the quantity of food consumed. There is no learning to control PWS hyperphagia. It includes preoccupations with food; food seeking, foraging; manipulation, sneaking, hiding, and hoarding food; and eating unusual food-related items (e.g. sticks of butter, pet food, mouthwash, rotten food taken from trash). It causes food-related anxiety that frequently results in dangerous behaviors (e.g. verbal aggression; physical aggression; elopement; burglary; theft; self-injury; lack of regard for personal safety).</li> <li>▪ Hyperphagia puts students at risk for morbid obesity and death from choking, gastric or bowel rupture or necrosis. People with PWS have died from a single food gorging incident.</li> <li>▪ Restricted calorie and carb intake is required. Student with PWS will gain weight on significantly fewer calories than their peers.</li> </ul>	<ul style="list-style-type: none"> <li>▪ Receive/follow prescription from health care professional for calorie-restricted diet.</li> <li>▪ Supervise student at all times, especially when food may be accessible. Monitor trash cans for discarded food items, especially in the restrooms. Keep unauthorized food out of sight.</li> <li>▪ Do not use food as a reward or in classroom activities.</li> <li>▪ It may be necessary to empty garbage cans that contain discarded food.</li> <li>▪ Staff should be trained in the Heimlich maneuver.</li> <li>▪ Avoid class parties that contain food. If unavoidable, ask family for input regarding how to manage food.</li> <li>▪ Treatment and management of hyperphagia consists of restricted access to food and continuous 1:1 supervision at all times. No currently known medication reduces or eliminates this life-threatening symptom.</li> </ul>
<p><b>Severe Stomach Illness</b></p> <ul style="list-style-type: none"> <li>▪ Severe stomach illness has been noted especially in students who have had access to unauthorized food.</li> <li>▪ Symptoms may include abdominal bloating. Pain may or may not be present; instead there may be a general feeling of not feeling well or fatigue.</li> <li>▪ It is rare for a person with PWS to vomit. If vomiting occurs it could signal life-threatening issue.</li> </ul>	<ul style="list-style-type: none"> <li>▪ If symptoms of stomach illness are present, notify parent. If there is known consumption of unauthorized food and student experiences abdominal bloating and/or vomiting, immediately notify parent as immediate hospitalization may be warranted.</li> <li>▪ Encourage the student to share honestly if they have accessed unauthorized food, however admitting to don't anticipate had a binge episode. The student should not be punished if this has occurred.</li> </ul>

<b>Health Concerns</b>	<b>Strategies</b>
<p><b>Maladaptive Behavior</b></p> <ul style="list-style-type: none"> <li>▪ <i>Persons with PWS have problems regulating their emotions.</i></li> <li>▪ <i>Many have difficulty with change or transitions.</i></li> <li>▪ <i>Some exhibit obsessive-compulsive tendencies, low tolerance to frustration, exaggerated emotional responses, and extreme verbal and/or physical aggression.</i></li> <li>▪ <i>Some may take medications to assist with mood stabilization.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ Minimize changes in routine. When they do occur – prepare ahead if possible.</li> <li>▪ Request information from PWCF about PWS Behavior Management Strategies.</li> <li>▪ State what behavior you want to see. Avoid using the words “no” or “don’t” because these spark frustration.</li> <li>▪ Make sure medications are administered at school at the appropriate times.</li> </ul>
<p><b>Osteoporosis</b></p> <ul style="list-style-type: none"> <li>▪ <i>High risk due to hormonal abnormalities and dietary limitations.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ At high risk for fracture – assess injuries for possible sprain/fracture. May require x-ray to rule out fracture.</li> </ul>
<p><b>Increased Sensitivity to Medications</b></p> <ul style="list-style-type: none"> <li>▪ <i>More sensitive to medications that can cause sedation or sleepiness. Metabolism of medication is slower than normal; risk of overdose. Start dose low and increase slowly.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ Be aware of medications that the student is taking that could cause this. Report any problems to parents.</li> </ul>
<p><b>Skin Picking</b></p> <ul style="list-style-type: none"> <li>▪ <i>Common behavior seen in persons of all ages.</i></li> <li>▪ <i>Open sores are common.</i></li> <li>▪ <i>May pick at various openings of body.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ Provide diversion activities – keep hands busy. Do not draw attention to the picking.</li> <li>▪ Encourage liberal application of lotion.</li> <li>▪ Incentive program often needed to keep wound covered.</li> <li>▪ Teach self care of wound if able.</li> <li>▪ Monitor frequent trips to bathroom. Time limits and supervision in bathroom may be needed.</li> </ul>
<p><b>Daytime Sleepiness</b></p> <ul style="list-style-type: none"> <li>▪ <i>Common to see in persons with PWS. Often symptom of sleep apnea.</i></li> <li>▪ <i>May be result of weak chest muscles-poor air exchange.</i></li> <li>▪ <i>Narcolepsy and cataplexy common.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ Physical therapy evaluation for muscle strengthening.</li> <li>▪ Get student up and moving if fatigue is noted.</li> <li>▪ May require a rest time during the school day.</li> <li>▪ May require medication.</li> <li>▪ Assist in communicating problem to health care provider if problematic.</li> </ul>
<p><b>Eye Abnormalities Strabismus and Amblyopia</b></p> <ul style="list-style-type: none"> <li>▪ <i>Strabismus and/or amblyopia result from poor muscle tone in eyes.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ Provide careful attention to this during eye screening.</li> <li>▪ Refer to eye specialist if needed. Vision Therapy may help avoid surgery.</li> <li>▪ If prescribed, ensure student wears glasses and/or patch.</li> </ul>
<p><b>Scoliosis and Other Orthopedic Problems</b></p> <ul style="list-style-type: none"> <li>▪ <i>Common to see scoliosis and other orthopedic deformities in persons’ with PWS.</i></li> <li>▪ <i>May be difficult to detect if obese.</i></li> <li>▪ <i>May require bracing or surgery.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ If suspected, recommend referral to orthopedic specialist.</li> <li>▪ Support and assist with brace if needed.</li> <li>▪ Adaptive measures may be needed for physical education.</li> </ul>
<p><b>Dry Mouth – Dental Problems</b></p> <ul style="list-style-type: none"> <li>▪ <i>Common problems: dry mouth causes thick, sticky saliva; teeth grinding, rumination, cavities.</i></li> </ul>	<ul style="list-style-type: none"> <li>▪ Teach and encourage good dental care and appropriate water intake.</li> <li>▪ Utilize special dry mouth products including over the counter ACT dry mouth toothpaste.</li> <li>▪ Help locate a dentist if needed.</li> </ul>

*For more information about supporting individuals with Prader-Willi syndrome contact:*

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