

Students with Prader-Willi Syndrome INFORMATION FOR RELATED SERVICES PROFESSIONALS

NOTE: The following are excerpts from the publication, *The Student with Prader-Willi Syndrome: Information for Educators*. This publication in its entirety is available through PWSA (USA).

It takes a team of professionals to help students with Prader-Willi syndrome (PWS) overcome and learn to deal with the challenges they face. Most students require services from Speech, Occupational and Physical therapists. The School Nurse may also be involved to help students with health concerns as well as educate school personnel about health issues. Because of high family stress in managing many aspects of PWS, the School Psychologist and/or a Social Worker may also be a part of a student's educational team.

Speech and Motor Delays – The Need for Occupational, Physical and Speech Therapy

Children and young adults with PWS experience both fine and gross motor delays as well as speech and language challenges. These problems are evident at birth and may be present in varying degrees throughout the individual's life. Generalized hypotonia (poor muscle tone) is often pronounced at birth. An infant typically exhibits very weak, floppy muscles. Most have a weak cry and require special feeding and arousal interventions. Because of these concerns, the infant with PWS is delayed in speech and motor developmental milestones. Early intervention services including speech, occupational and physical therapies should be pursued. Assessments, treatment plans, and ongoing interventions must be very individualized to meet each student's needs.

Most children and young adults with PWS have problems with speech and language as a result of poor muscle tone and cognition. Hypotonia affects oral motor structures that result in problems with speech formation as well as feeding. These muscle weaknesses may result in articulation deficits. Many children are being diagnosed with a specific motor speech disorder called speech apraxia. These students have difficulty sequencing the motor movements needed to execute speech movements. Most children with PWS suffer more delays in expressive language with minimal problems receptively. A child's cognitive level can also impact their ability to understand, process and communicate their wants and needs. A delay in cognition can manifest itself in difficulty for the student to problem solve and use complex grammar and vocabulary. It is important to find a way for these students to communicate. In the early years, many behavioral concerns stem from frustration in not being able to communicate their needs. As the child grows older, muscle tone tends to improve. Often speech and motor problems or delays continue. Therapy often targets incorporating language skills into social, work and other life skill areas. They also benefit from learning strategies that will help them with nonverbal communication.

Low tone, decreased strength, poor motor planning as well as delayed fine and gross motor skills are very common in children and adults with PWS. It is common to see a child struggle with writing, cutting and other fine motor tasks. In the early years, they may face challenges in balance, coordination and strength. Physical and occupational therapy services help the student develop, strengthen and refine their physical abilities to participate in classroom activities as well as recreational opportunities. Many students find success with new assistive technology including the computer. Therapy intervention should target incorporating physical activities that can be enjoyed and utilized throughout the students' life.

Health and Wellness – The Need for Health Services

Students with PWS may face challenges in maintaining health and wellness. Knowing and understanding health issues as well as utilizing strategies that will assist with nutrition and exercise can facilitate wellness. Many of these students also have mental health issues and concerns. The chart summarizes some of the health issues faced by students as well as suggestions that will assist in their management. Additional suggestions are also provided in the age-grade information for educators.

The Student with PWS - Health and Wellness

Health Issue	Strategies
Increased Food Drive/ Low Metabolism – High Risk for Obesity Related Problems The cause for the varying degrees in food seeking behavior is a dysfunction in the hypothalamus as well as a decreased growth hormone production. Initially you may begin to see a food preoccupation. Typically it advances to food seeking and/or stealing. Students can experience a very rapid weight gain. Associated problems include Diabetes II and hypertension.	- Receive prescription from health care professional and implement a calorie-restricted diet. - Supervise and monitor all food sources. Keep food, purses and money out of sight. (Money buys food) - If needed, weekly weight monitoring by school nurse. - If indicated, assist and refer parent to a nutritionist.
Behavior – Emotional Problems Many children and young adults with PWS exhibit difficulty in regulating their emotions and behavior. This is especially true in handling unplanned changes. Most thrive on consistency and routine and can become easily upset when disruption occurs. Some exhibit obsessive-compulsive tendencies and have trouble when unable to complete tasks in a specified time frame. Exaggerated emotional responses may be seen. Behaviors can range widely. Some may have few problems while others may escalate to rage-like episodes. Proactive behavior management strategies are often successful. Many students require services from a mental health professional. Some may require medications to help them with these problems.	- Foreshadow changes and allow time and “safe area” for student to share emotions. - Teach and model use of words and/or pictures in sharing emotions. - Teach and emphasize what behavior you want to see. Avoid using word “don’t” <u>Examples:</u> <u>DO SAY:</u> “David, you need to use a quiet voice.” <u>DON’T SAY:</u> “Don’t yell” <u>DO SAY:</u> “Susie, keep your hands to yourself.” <u>DON’T SAY:</u> “Don’t hit.”
Decreased Pain Sensitivity/ High Pain Threshold Many students with PWS lack the typical response to pain, which may mask signs of infection and/or injury. A person with PWS may not complain of pain until the infection is severe. It may also be difficult for the individual to identify the location of the pain. This can make it difficult to assess for illness or injury.	- Assess all injuries carefully. - Report concerns to the parent or caregiver and find out how the student exhibits illness or pain, as the student may show other signs when becoming ill. Increased fatigue and irritability are often reported.
Increased Bruising Even though blood testing is normal, many persons with PWS bruise easily. Therefore it is important to note the cause and location of any bruises that occur during the school day.	- Report and/or document all bruises that may be caused from a school-related incident. - Apply ice and pressure when injuries result in bruising.
Altered Temperature Regulation Unexplained high and low body temperatures are common. Little or no fever may be present with severe infections. There may be a quick response to high outside temperatures.	- Make sure student does not become overheated. - If illness is suspected, the parent or caregiver should seek medical attention.

The Student with PWS - Health and Wellness

Health Issue	Strategies
• Skin Picking Although the cause is not clearly understood, many children and adults show signs of skin picking and scratching. Open sores are common. These open sores are often mistaken as cigarette burns. Insect bites are often sources of initial lesions. There are some children and adults who pick at various body openings.	• Provide diversion activities – “keep hands busy” - coloring, puzzles, going on computer, writing, hand-held games • Monitor frequent trips to bathroom. Time limits may be needed. Supervision may be required. • Teach the student basic first aid if bleeding occurs. Apply bandages or other coverings. • Encourage liberal application of lotion – provides diversion. Moisturized skin is more difficult to tear and/or open. • Apply insect repellent prior to walks or other outside activities.
• Daytime Sleepiness It is common for students with PWS to exhibit daytime sleepiness. This may be a result of altered breathing patterns in the hypothalamus. Poor muscle tone, weak chest muscles and upper airway obstruction may also contribute to this. If snoring, sleep walking or persistent morning headaches occur, he/she should be evaluated for obstructive apnea (a breathing disorder in which a person stops breathing for prolonged periods while sleeping). Some students respond well to a change in activity – sending them on an errand, taking a walk... Some require a short afternoon rest. There are also therapies that may be tried to alleviate apnea.	• Get student up and moving – send on errand, take a walk, jump on a mini trampoline. • Some require short nap • Assist in communicating problem with health care provider and see if referral to a pulmonologist (lung specialist) is needed.
• Strabismus (Cross Eye) Because of generalized poor muscle tone, many children with PWS, especially younger ones may be at risk of developing strabismus (cross-eye). Glasses, patching or surgical correction may be needed.	• Provide careful attention during eye screening. • Refer to an eye specialist if needed. • Make sure student wears eyeglasses if needed. • Assist with eye patching if indicated.
• Scoliosis and Other Spinal Problems Scoliosis (lateral curvature of the spine) as well as kyphosis (“hump back or hunch back”) can occur in children and young adults with PWS. It is often difficult to detect. Braces and surgery may be indicated.	• If suspected, student should be referred to an orthopedic specialist. • Support and assist if brace is needed.
• Dental Caries Many with PWS are at increased risk of developing dental caries (cavities). Thick, sticky saliva as well as poor tooth brushing skills may contribute. It is also common to see teeth grinding and rumination. Dental sealant and fluoride treatments have been helpful at decreasing cavities.	• Teach and encourage good dental care. • Assist in locating a dentist if needed.
• Osteoporosis Because of hormone abnormalities and dietary limitations, persons with PWS are at high risk of developing osteoporosis at a young age. This can put them at higher risk for fractures and/or spinal problems.	• Therapies often focus on hormone, calcium and vitamin replacement. • Weight bearing exercise (walking) should be encouraged.

The Student with PWS - Health and Wellness

Health Issue	Strategies
• Early or Incomplete Onset of Puberty The average developmental milestones in most children and young adults with PWS are typically delayed by 1-2 years. In most cases, sex hormones (testosterone and estrogen) are low. Early pubic hair and some other signs of precocious puberty may be seen. Puberty is usually late and often incomplete. Most young women do not menstruate or have irregular periods. Fertility is extremely rare. Both sexes do respond to hormone therapy. Most young people with PWS want very much to bear children and become parents. Care must be given in providing accurate sex education.	• Work with parents in educating the student about changes in their bodies and the need for increased attention to hygiene.
• Severe Stomach Illness Abdominal bloating, pain and vomiting may be signs of life threatening stomach problems, which are more common in persons with PWS than in the general population. Rather than localized pain, there may be a general feeling of “unwellness”.	• If a student has these symptoms, he/she should see a health care professional immediately. • Monitor for signs of constipation or other bowel problems. Work with parents and health care provider on encouraging a high fiber diet with increased intake of water.
• Lack of Vomiting Children and young adults with PWS rarely vomit. The presence of vomiting may indicate a serious problem and should be evaluated by a health care professional. Using syrup of Ipecac in the case of poisoning is often ineffective and not recommended. Other treatment options are often needed.	• In the case of accidental ingestion of poisons, make sure parent shares medical brochure with health care provider, which informs him/her of this issue.
• Increased Sensitivity to Medications It has been noted that many people who have PWS have an increased sensitivity to many medications. This is especially true with medicines that can cause sedation or sleepiness. Some people with PWS have experienced water intoxication (a serious problem resulting from too much body water). Taking medication that alters the body’s water levels or taking medicine that causes increased thirst and increased fluid intake seems to be the most likely cause of this.	• Be aware of the medications the student may be taking and report any side effects to the parent or caregiver. • Consult with health care provider if indicated.

Growth Hormone Therapy and the Student with PWS

Research and knowledge have increased on the benefits of growth hormone therapy in children and adults with PWS. This hormone affects many body systems. It increases bone density and prevents osteoporosis. It increases muscle mass, which helps burn calories; increases strength and endurance; and possibly prevents other health issues related to poor muscle tone. Research has shown that growth hormone therapy decreases sleep-disordered breathing and lowers blood lipids. More young children and young adults with PWS are receiving growth hormone therapy. Students are often taller and leaner. Many are more active. Some health problems are being prevented or minimized. So why don’t all children with PWS receive this therapy? As with any treatment, there may be individual situations that contraindicate the use of growth hormone. It is not yet a standard of care for all children with this disability. Therapy involves a daily injection and families may not feel comfortable with this. Some insurance companies will not pay. And there are some families and some physicians who are uninformed.

Complex Behavior & Family Issues – The Need for Psychological & Family Support Services

The constant need to monitor and manage complex behavior in students with PWS can cause extreme stress for many families. Families have varying skills and resources. Some require a great deal of support in behavior management – some do not. Some may be connected to support resources – others may not. School districts and communities also have varying resources. Depending on the family and each school's situation, educators may be called upon to assist families in obtaining parenting skills as well as helping them to connect to human service resources. Social workers, guidance counselors and/or school psychologists are often key people to provide information and support to families in need. The school psychologist is also instrumental in working with the educational team in developing successful behavior plans that can be used at home as well as in school. The psychologist is often the key person in conducting testing that helps identify specific learning and intellectual needs.