

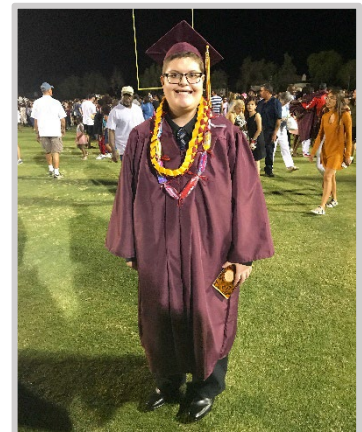
## Prader-Willi Syndrome (PWS) – FACT SHEET School Age Children and Young Adults with PWS

### **Brief Overview of PWS**

- PWS is a very complex, neurobehavioral, genetic disorder caused by an abnormality on the 15<sup>th</sup> chromosome. Most of the challenges these children and young adults face result from a defect in the area of the brain called the hypothalamus.
- It affects both males and females equally and in all races around the world.
- The prevalence range is 1:15,000 to 1:25,000 individuals.
- There are other genetic conditions or health issues that impact the hypothalamus which may require the same support as a person with PWS.
- It is a “spectrum disorder” meaning there are personal differences in the degree of challenges that these individuals face. They are individuals. They are not all the same.
- Due to advances in research and hormone replacement therapy, many children and young adults with PWS have experienced significant health and developmental benefits from growth hormone.
- There is no cure however, ongoing research continues. We are gaining more knowledge and understanding to help prevent and manage many of these challenges. There is hope for a cure.

### **Common Features You May See**

- Children with PWS are beautiful, caring, happy individuals. As they grow older, some face challenges with controlling their emotions. They can experience times of anxiety, frustration, and behavior escalations.
- As the child with PWS grows older, many become more obsessed with food and food-related activities. Many require food security strategies and supervision around all food sources. For most, food security measures, including locking up food, need to be started and maintained.
- Many continue to experience daytime sleepiness. They may require rest periods and an early bedtime.
- Most students require assistance with learning through special education services. They may also receive occupational, physical and speech therapies. Many also qualify for transportation services.
- Many older students lack age-appropriate social skills and benefit from social skills instruction and coaching.



### **Health Concerns**

- Growth Hormone (GH) deficiency. Persons with PWS of all ages benefit from Growth Hormone therapy. This treatment helps to strengthen muscles and helps with increased metabolism and bone health. They should complete an evaluation by a pediatric endocrinologist. It is never too late to begin this treatment.
- Even if the person is receiving GH treatment, it is important to begin or maintain a nutritious, calorie-controlled diet and monitor the child’s weight daily. A more structured approach to snacks and mealtimes often helps to lessen anxiety. Take advantage of any ways to become more educated on preparing low calorie, highly nutritious food. Learn to read nutrition labels. Keep food out of sight.
- Temperature regulation problems. They may not run fever when infection is present. They are sensitive to high or low external temperatures. Some may not wear clothing appropriate for the weather and need guidance.

- Respiratory issues – hypoventilation (shallow, slow breathing), sleep disordered breathing, and sleep apnea may be seen in some children. Some may require Continuous Positive Airway Pressure (CPAP) therapy. Prompt assessment and treatment of upper respiratory infections is often needed.
- Orthopedic issues – higher incidence of hip dysplasia and scoliosis. Consultation with a pediatric orthopedic specialist is recommended. Many require bracing and sometimes surgery for scoliosis.
- Sensitivity to medications – especially those that cause sedation, including anesthesia, as well as medications used to treat mood stabilization and diarrhea.
- Dental care is very important. Dry mouth, grinding of teeth and poor oral hygiene is often seen in children and adults with PWS. It is a life-long struggle. It is important to have the individual brush their teeth at least twice a day. Daily flossing is also recommended. It is suggested that they see a dentist at least every 6 months. If severe grinding of teeth is noted, this should be discussed with your dental professional.
- People with PWS have an altered pain response. They do not feel pain like others. It can make it challenging to know if they have a serious injury. It may be necessary to have them evaluated by a health care professional when they have a more severe injury or have continued complaints of pain or discomfort.
- Individuals with PWS are at greater risk for experiencing constipation. This is often a lifelong health concern. It is important to encourage a diet of high fiber foods along with water/fluids. Monitor their stools (poop). If a problem is identified, discuss this with their primary care professional.
- Many children with PWS do not like to drink water. Water is very important. If a child refuses to drink water, many flavor it with lemons, cucumbers or other fruits or vegetables. Avoid artificial sweeteners.
- It is rare for anyone with PWS to vomit. This can indicate a serious health problem. It may be caused when their stomach empties too slowly. All cases of vomiting should be evaluated by a health care professional immediately.
- Keep the [PWSA I USA Medical Alerts booklet](#) handy or access the [GI Chart](#) on PWSA I USA's website.



### **Supporting the School Age Student and Young Adult**

- Most students with PWS benefit from special education services as well as other therapies to help correct or prevent further muscle weaknesses, speech and learning challenges. Some can go on to post high school education including 2- and 4-year colleges.
- The older student with PWS may benefit from pre-vocational assessments, skills, and opportunities.
- Keeping them active is very important to help strengthen muscles, enhance bone development and burn calories. Playing, walking, swimming and/or other physical activities should be done daily.
- It is helpful for all persons with PWS to have a predictable schedule, especially when it comes to eating times and patterns. It is also helpful to develop routines for bedtime, exercise, and other activities. This provides emotional security and lessens anxiety.
- Persons with PWS may start to demonstrate more behavior challenges when they face frustration or other stressful situations. Food, fairness (they want things to be the same as others) and unplanned change are three common areas that often cause behavior escalations. In some cases, these behaviors can be severe. Some may be diagnosed with mental health conditions. In those cases, individuals should be evaluated by a mental health professional for possible medication treatment.
- Parents often find it helpful to use consistent behavior management strategies. Acknowledging their feelings; providing a “quiet area” to settle down; as well as finding a way for them to share their feelings are often utilized. Minimizing change or preparing for change can also be helpful.

For more detailed information about PWS, contact PWSA I USA at the email [info@pwsausa.org](mailto:info@pwsausa.org), the website [www.pwsausa.org](http://www.pwsausa.org), or phone number (941) 312-0400.