

## Prader-Willi Syndrome (PWS) – FACT SHEET

### Toddlers 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 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 children face. They are individuals. They are not all the same.
- Due to advances in research and hormone replacement therapy, many children 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**

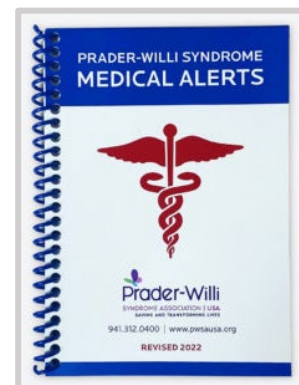
- Children with PWS are beautiful, caring, happy individuals. They can experience times of frustration and have temper tantrums like other toddlers.
- All experience poor muscle tone, which often results in delayed developmental milestones. They may be slower to crawl, walk and begin potty training. However, you will see progress as their muscles grow stronger. Growth hormone treatment helps to strengthen muscles, and many have fewer delays.
- They tend to sleep more and often require more naps and an earlier bedtime. Daytime sleepiness is common. They often fall asleep while eating and riding in a car.
- Some toddlers begin to become more interested in food and all food related activities. For these children, food security measures may need to be started.



#### **Health Concerns**

- Growth Hormone (GH) deficiency is common. There are numerous benefits for a child to begin GH as soon as they complete an evaluation by a pediatric endocrinologist.
- It can be challenging for the infant to gain weight. However, many toddlers start to gain weight more easily. It is important to begin a calorie-controlled diet and monitor the child’s weight daily. A nutritious diet is very important, as well as a more structured approach to snacks and mealtimes. Take advantage of any ways to become educated on preparing low calorie, highly nutritious food. Learn to read nutrition labels.
- Because of their muscle weakness, it often takes a toddler time to master chewing and swallowing. It is important to provide a nutritious high fat diet, especially during the first 2 years. Eating “good fats” (nuts, fish, avocados, seeds etc.) is good for all of us and should be included in moderation for the child with PWS every day lifelong. Talk to your health care provider about the use of fat free milk and other products.
- Infant boys may experience undescended testicles. Discuss with the primary care provider. A referral to a pediatric surgeon or urologist may be needed.
- Temperature regulation problems. They may not run fever when infection is present, and they are sensitive to high or low external temperatures.

- Respiratory issues – hypoventilation (shallow, slow breathing), sleep disordered breathing, and sleep apnea may be seen in some children.
- Orthopedic issues – higher incidence of hip dysplasia and scoliosis. Consultation with a pediatric orthopedic specialist is recommended.
- Some experience eye muscle weakness resulting in cross eyes. Consultation with a pediatric ophthalmologist is suggested.
- Sensitivity to medications – especially those that cause sedation, including anesthesia.
- As teeth begin to erupt, it is important to begin brushing and cleaning the teeth. This also provides oral sensory stimulation. It is suggested that children begin to see a dentist around the age of 1 year or when teeth start to erupt. If severe grinding of teeth is noted, discuss this with your dental professional.
- Children with PWS have an altered pain response. They do not feel pain like other children do. 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.
- Children with PWS are at greater risk for experiencing constipation. 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 by their stomachs emptying 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 the PWSA I USA website.



### **Supporting the Toddler**

- Toddlers with PWS benefit from early intervention services to help prevent or slow developmental delays. It is common for them to receive therapy from an occupational, physical and speech therapist.
- Because they require more sleep, parents must plan for naps, rest periods and earlier bedtimes.
- Keeping them active is very important to help strengthen muscles, enhance bone development and burn calories. Play is the number one way to keep them moving.
- It is helpful for all toddlers to have a predictable schedule, especially when it comes to eating times and patterns. It is also helpful to develop routines for bedtime, play time (exercise) and other activities. This provides emotional security and lessens anxiety.
- Toddlers with PWS may start to demonstrate temper tantrums 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 tantrums.
- Parents often find it helpful to begin 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.
- Continue to take time to enjoy your toddler. Play, talk, and read to them. Take them for walks. Celebrate every accomplishment.

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.