

# PRADER-WILLI SYNDROME

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## Symptoms or Behaviors

A major concern of PWS is obesity. If not controlled, the obesity can become life threatening and may result in diabetes, hypertension, breathing difficulties, heart disease and skeletal problems.

School staff should watch for the following:

- Food intake
- Weight increases
- Skin picking
- Sleepiness
- Speech defects in articulation
- Scoliosis
- Injury due to high pain threshold
- Respiratory difficulty
- Bruising (tendency to bruise more easily)
- Behavior Problems
- Vomiting (rarely occurs with PWS; if vomiting occurs may indicate a life-threatening illness)

Not all children with Prader-Willi Syndrome are the same. Staff must become familiar with the needs of individual children.



## About the Disorder

Prader-Willi Syndrome (PWS) is a genetic disorder caused by the lack of normally active DNA from the father on the long arm of chromosome 15. In 70% of the individual's with Prader-Willi Syndrome, there is a deletion or unbalanced translocation on the father's chromosome 15. In 25% of the cases, both chromosome 15's may be from the mother, and none from the father. In 1-4% of people affected there is a mutation on the Chromosome 15. This is the only cause of Prader-Willi Syndrome recurring in families. In 1956, doctors A. Prader, H. Willi and A. Labhart described the syndrome, although descriptions of individuals with PWS can be found as far back as the 17<sup>th</sup> century. About 1 in 12,000 to 1 in 15,000 live births result in Prader-Willi Syndrome. Males and females are affected equally and it occurs in all races.

Prader-Willi Syndrome is divided into two distinct clinical stages. Stage one occurs from birth through the ages of two to four years of age. Babies are born with very low muscle tone, and may need to be fed with special nipples or be tube fed. Due to the low muscle tone babies need to be turned when sleeping and most take longer than normal to reach developmental milestones. During the first year or two of life, infants with PWS put on weight slowly. If a diagnosis of PWS has not been made, the infant is labeled as "failure to thrive" producing anxiety for the parents.

The second stage of the PWS occurs around age two to four years. The child with PWS has a sudden increase in appetite and the child consumes everything put in front of them. Diet must be carefully monitored or weight gain is rapid. In addition to increased food intake, fewer calories are needed to maintain an acceptable weight (30-50% less than average). Calories are burned more slowly, therefore food intake must be carefully monitored and an exercise program implemented. A part of the brain (hypothalamus) that determines hunger and fullness is flawed. Individuals with PWS also have short stature, small hands and feet, hypogonadism (small genitalia and delayed puberty), obsessive compulsive behaviors (often picking at their skin/sores and requiring a consistent daily care routine), tendency to be argumentative/oppositional/ rigid, stubborn, temper tantrums, eye abnormalities, speech articulation defects (low muscle tone).

Several methods are used to diagnose the condition. Chromosome analysis identifies some of the cases, while methylation analysis detects almost all cases of Prader-Willi Syndrome. Tests called by the acronyms FISH and PCR (polymerase chain reaction) will identify 99% of the cases when both are given. In addition to the characteristics described distinctive facial features include almond-shaped eyes, small-appearing mouth with thin upper lip and downturned corners of the mouth and narrow face.

Medications and appetite suppressants have had very limited success. Growth hormone therapy has been approved to treat individuals with PWS and appears to increase muscle mass, strength and endurance, allowing for more physical activity.

One of the greatest challenges for families is the supervision of food around-the-clock to control obesity and prevent the complications that occur with it.

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## Educational Implications

Individuals with Prader-Willi Syndrome have IQ's ranging from 40 to 105, with an average of 70. Individuals with average IQ's often have learning disabilities with mixed abilities. Problem areas often include difficulties with attention, short-term memory, reading ability, receptive language, and poor auditory processing.

Children tend to socialize with adults easily but are often more isolated from peers. Concerns, especially at the secondary level include easy access to food, being exposed to unsafe laboratories or workshops, having privacy in changing rooms (especially for boys who are not as physically developed as peers), name calling/hurtful comments, having the opportunity to compete in team games and the opportunity to make close friends.

Evaluation to determine the need for therapies (speech, occupational therapy, physical therapy, adapted physical education) and/or special education is needed throughout the child's school career. A school diet management plan may be part of an overall health plan.

## Instructional Strategies and Classroom Accommodations

There is a strong need for routine, sameness and consistency in the environment. Warning about changes in plans or routines and time countdowns before transitions may be necessary. When a student is stuck on an issue, reasoning with them is not successful. Try redirecting them to another topic or defer the discussion to some other time. Provide external motivation and rewards.

Most students with PWS do better with concrete rather than abstract concepts. Strengths are often seen in areas such as jigsaw and word search type puzzles. Use of visual cues assists the student in carrying out instructions.

If poor short term memory is present the child may appear to have learned a topic one day, only to have forgotten it the next. Repeated revision and consolidation is needed to ensure the topics are fully understood.

Poor emotional control (tantrums, yelling, swearing, aggression and self-injury) can be brought on when the student is stressed. The student needs time to recover and often feels sadness, remorse and guilt. Reassurance is needed. Time out is often a successful strategy. Social skills often need to be taught to the student with PWS.

The student with PWS has an insatiable appetite and will never have control of their craving for food. Students should never be left alone with food. Provide close supervision and make sure all staff are aware of dietary restrictions. Teachers should communicate with parents on alternatives to "school treats". Encourage a daily exercise routine to help maintain weight and prevent fatigue.

People with PWS tend to tire more easily and may fall asleep during the day. Morning is their best learning time and classes need to be scheduled to meet optimal learning times.

## Resources

The Prader-Willi Syndrome Association (USA)  
1-800-926-4797  
5700 Midnight Pass Rd.  
Sarasota, Florida 34242  
E-Mail: [national@pwsausa.org](mailto:national@pwsausa.org)  
Web: <http://www.pwsausa.org/>

The Prader-Willi Connection  
<http://www.pwsa-uk.demon.co.uk/educatn.htm>

### Books:

My Name's Not Willy!  
<http://www.pwausa.org>

Michael and Marie: Children with Prader-Willi Syndrome  
By Valerie Rush Sexton and Debbie Fortin (designed to be read in an elementary classroom)

Prader-Willi Syndrome, A Practical Guide  
By Jackie Waters, Fulton Publishers

Research on Behavior and Development in Prader-Willi Syndrome  
By Elisabeth Dyken

Transition From School to Adult Services in Prader-Willi Syndrome: What Parents Need to Know  
By Julie A Seguin and Robert M. Hodapp

School Nurse's Source Book of Individualized Health Care Plans – Volume II  
Sunrise River Press, c. 2005  
[www.schoolnursebooks.com](http://www.schoolnursebooks.com)