



## Information for School Staff Supporting the Student Who Has Prader-Willi Syndrome Across Their Life Span

Compiled by Barb Dorn RN, BSN, Outreach Program Director, PWSA of WI, Inc.

<b>Common Characteristics Seen in Students With PWS</b>	<b>Possible Management Strategies</b>
<p><b><u>POOR MUSCLE TONE</u></b></p> <p><b>Excessive Sleepiness</b> Babies with PWS exhibit excessive sleepiness often sleeping for 20-23 hours a day. Parents are not able to feed on demand because the baby often will not cry or communicate hunger.</p> <p><b>Developmental Milestone Delays</b> Due to poor muscle tone, young children with PWS are delayed in their growth and developmental milestones. Even though they proceed in the same order as other children, events such as sitting up, standing, and walking come later. In later years, this also impacts many social/recreational milestones such as riding a bike, roller skating, etc.</p> <p><b>Fine Motor Delays</b> As the student with PWS grows older, many are faced with weaknesses and challenges in their fine motor skills. Daily living skills as well as hand strength activities (cutting, writing) may be affected.</p> <p><b>Gross Motor Delays</b> Students with PWS typically have generalized low muscle tone. This contributes to their delay in many developmental milestones. It also accounts for lifelong challenges with large muscle weakness and coordination problems.</p> <p><b><u>ALTERED NUTRITIONAL NEEDS</u></b></p> <p><b>Feeding Difficulties</b> The number one concern for infants who have PWS during their first 6 months of life is <b>feeding difficulties</b>. Most of these babies have a poor suck with an inability to cry when needing to be fed. They also experience poor weight gain. This is best known as <b>the Failure to Thrive stage</b>.</p>	<p><b>For EXCESSIVE SLEEPINESS</b></p> <ul style="list-style-type: none"> <li>• Provide various forms of stimulation. <b>DO NOT</b> allow the child to sleep all day.             <ol style="list-style-type: none"> <li>1. Awaken baby for feedings.</li> <li>2. Provide physical contact.</li> <li>3. Play/talk with baby.</li> <li>4. Provide bright, colorful surroundings.</li> <li>5. Reposition baby.</li> <li>6. Play music.</li> </ol> </li> </ul> <p><b>For DEVELOPMENTAL DELAYS</b></p> <ul style="list-style-type: none"> <li>• Refer to Early Intervention Program with team of professionals to assist with stimulation activities (OT, PT, Speech &amp; Language clinicians, &amp; Educators).</li> <li>• Teach parents exercises, play and toys that will help enhance progression of motor development.</li> <li>• Empower the parent in helping in child's progress.</li> </ul> <p><b>For MOTOR DELAYS</b></p> <ul style="list-style-type: none"> <li>• Provide school-based therapies — OT and PT for strengthening and overall enhancement of fine motor and large muscle activities.</li> <li>• Assist with cutting and writing activities.</li> <li>• Start with keyboarding.</li> <li>• Modify with art work activities.</li> <li>• Assist with daily living skills — tying shoes, cutting food.</li> <li>• Provide adaptive gym class if indicated.</li> </ul> <p><b>For FEEDING DELAYS</b></p> <ul style="list-style-type: none"> <li>• During the first 6 months the infant may need:             <ol style="list-style-type: none"> <li>1. Gavage (tube) feeding and/or specialized nipples.</li> <li>2. Oral stimulation program — OT or Speech Pathologist.</li> <li>3. Referral to a nutritionist — GOAL is not to overfeed or underfeed.</li> </ol> </li> <li>• Parents may need encouragement to bond with child (cuddling, snuggling), possibly after feeding.</li> </ul>

## ALTERED NUTRITIONAL NEEDS cont.

### **Weight Gain, Preoccupation with Food and Food Seeking**

Around the age of 2, children with PWS start to experience weight gain with a noticeably greater interest in food. In some young children it proceeds quickly to food seeking and stealing. Others move at a slower pace with the desire to seek out food. As they grow older, the craving for food increases. It becomes a life long struggle, and it does not resolve. This situation is compounded in the fact that food is metabolized at a very low rate, and this can cause extraordinary weight gain very quickly if food and diet is not monitored. We typically call this the **Thriving Too Well stage**.

## SPEECH AND LANGUAGE

### **Expressive and Receptive Language Difficulties**

PWS can impede all aspects of speech and language development. Poor muscle tone and incoordination of facial and oral muscles can affect a child's ability to make sounds and form words. When intellectual capacity is impaired, both expressive and receptive language may be affected. Typically, children with PWS have stronger receptive skills.

### **Neuromotor and Articulation Problems**

Poor muscle tone as well cognitive limitations can cause articulation problems as well as a slow rate of speech and poor voice quality. A decreased ability to communicate may contribute to increased frustration in the student with PWS.

### **For WEIGHT GAIN, FOOD PREOCCUPATION, AND/OR FOOD SEEKING:**

- Refer to nutritionist who has knowledge about PWS.
- Initiate low calorie diet with supervision of food in all environments.
- Structure meal/snack times.
- Do not use food as a reward.
- Avoid using food in classroom projects.
- Work with parents in establishing guidelines for treats or eating of extra food while at school.
- Don't leave student alone in areas of food. Be aware of staff candy dishes or other sources of food.
- Supervise in the lunchroom and in all food related areas — vending machines, school stores.
- Allow student to eat in classroom (with peer or friend) if lunchroom becomes overwhelming.
- Discuss any delays in snack or lunch ahead of time (often affected by field trips or schedule changes).
- PRAISE situations where the student did well with handling a food situation.
- Be aware that parents often have to lock their refrigerator and cabinets at varying times (individual) This is a health and safety measure.
- The school nurse may need to monitor weight weekly or more frequently if indicated. Counseling may be needed.
- Develop a plan of dealing with stealing or trading of food. These students should not be punished but a proactive approach should take place.
- Supervise students in hallways or near unlocked lockers. Not only will they steal food, but they may steal money to buy food.
- Find an appropriate vocational placement that does not involve food. (Library, Mail room, Computer work, Sheltered Workshop, Flower or Gardening activity).
- Parents may want to educate peers and classmates regarding this disability to elicit support for the unique dietary needs.

### **For SPEECH AND LANGUAGE**

- Encourage and respond to vocalizations.
- Refer to a Speech Pathologist who can provide:
  1. Augmentative Communication Techniques — sign language, picture/symbol boards. These are usually temporary, but it helps to decrease frustration.
  2. Variety of techniques to improve pronunciation, vocabulary skills, or word formation.
  3. Assistance in learning socially appropriate communication behaviors — eye contact, initiating conversations.

## LEARNING DIFFERENCES

### **Mental Retardation or Learning Disabilities**

Approximately 60% of persons with PWS have mental retardation. 40% have learning disabilities. You will see quite a range in IQs with most students functioning in the borderline area

Some of the LEARNING CHALLENGES these students face include:

#### **1. POOR SHORT TERM MEMORY**

Many students have problems with short term memory processing — especially when the information is given verbally. This is often misunderstood as disobedience because many times the student has been successful at understanding a "piece" of the information that is given to him/her.

#### **2. SEQUENTIAL PROCESSING DEFICIT DIFFICULTY UNDERSTANDING ABSTRACT CONCEPTS**

Students with PWS have a deficiency in the **INTAKE, PROCESSING AND RESPONSE** of information. When a student has a sequential processing deficit, he/she will not learn from "experience". The timing and order gets all mixed up so the "if \_ then" reasoning is absent. This is a key frustration factor for many parents and educators in working with these students. This is also the basis for failure in behavior modification programs.

Other areas of learning impacted by this include:

- Difficulty with reading comprehension problems
- Trouble interpreting and executing multi-step directions
- Difficulty summarizing and restating events
- Challenges in scheduling timelines for completing long term projects

#### **3. PROBLEM SOLVING DIFFICULTIES**

Students with PWS often have difficulty planning or ordering steps to problem solve. They tend to be "Black and White" thinkers. They have difficulty using deductive reasoning.

Some of the LEARNING STRENGTHS often seen in students with PWS include:

- **GOOD LONG TERM MEMORY SKILLS AND GOOD ROTE SKILLS** — Once the student has learned the material, there is typically good retention.
- **MULTI-SENSORY LEARNERS** - Are very much **VISUAL** learners as well as **HANDS ON** learners.
- **STRENGTH IN READING** - May do better with books and tapes.

#### **For SHORT TERM MEMORY PROBLEMS**

- Teach strategies to increase short term memory:
  1. Verbal rehearsal.
  2. Lists — put in writing.
  3. Repetition of directions — "practicing".
  4. Visuals — pictures.
- Provide students with written format for daily announcements.

#### **For SEQUENTIAL PROCESSING DEFICIT AND DIFFICULTY WITH ABSTRACT CONCEPTS**

- Use visual representation of time — calendars, clocks.
- Keep instructions/steps limited to 2-3 steps.
- Assist with summarizing activities.
- Assist with setting up realistic goals and timelines (Make sure they are short term to lower frustration level).
- **BEHAVIOR MANAGEMENT TECHNIQUES** have been found to be most successful.
  1. Practice proactive planning.
  2. Minimize or remove situations or environmental factors that provoke behaviors (especially food).
  3. Emphasize **APPROPRIATE** behavior (reward the good instead of punishing the bad).
  4. Set goals and management to **DECREASE FREQUENCY AND INTENSITY** of problem behaviors.

#### **For PROBLEM SOLVING DIFFICULTIES**

- Rehearse appropriate behavior expectations ahead of time.
- Take advantage of pointing out learning opportunities as they arise in the community.
- Have simple, clear **WRITTEN** rules and expectations.

## **BEHAVIOR**

### **Rigid Thought Process**

People with PWS often need to receive and store information in a very orderly manner. There is a strong need for routine, sameness, and consistency in the learning environment.

### **Perseverative Thinking**

This is the tendency to get "caught" on one issue or thought to the point where it overshadows the main theme of the learning or social event. Perseveration can lead to loss of emotional control.

### **Tenuous Emotional Control**

Any combination of life stressors can lead to emotional "discontrol" as evidenced by tantrums or rages — yelling, swearing, aggression, destruction, and self-injury. Recovery of control takes time and is often followed by sadness, remorse, and guilt.

## **SOCIAL**

### **Difficulty with Peer Interactions**

While children want and need other children and value friends, it may be difficult for them to be exposed to the unpredictability of others for long periods. The need for order frequently translates into fairness issues and comparing themselves to others, often resulting in anger.

## **HEALTH RELATED CONCERNS**

### **Strabismus (Crossed-Eye)**

Due to poor muscle tone, many young children with PWS develop strabismus. With proper following by a pediatric ophthalmologist, these children can have this problem corrected.

### **For RIGID THOUGHT PROCESS**

- Foreshadow changes and allow for discussion.
- Use visuals; put things in writing — lists, schedules.
- Communicate changes in personnel ahead of time.
- Don't make promises you can't keep.
- Give warning of ending ("Ten more minutes").
- Breakdown procedures into concise, orderly steps.

### **For PERSEVERATION**

- Use reflection — have student restate what you said.
- Set limits. "I'll tell you one more time, then we move on to next topic".
- Put in writing; use visuals.
- Avoid power struggles and ultimatums.
- Ignore (if possible).

### **For TENUOUS EMOTIONAL CONTROL**

- Provide positive attention and praise when person is maintaining control, especially in difficult situations.
- Prevent loss of control by encouraging communication and acknowledging feelings.
- Anticipate build up of frustrations and help him/her to remove self to "safe area" where he/she can share feelings.
- Don't try reasoning during times when out of control.
- Have a plan in place if student becomes more violent. Be consistent in your approach in how this is handled.
- Provide positive closure. Don't hold a grudge.

### **For DIFFICULTY WITH PEER INTERACTIONS**

- Provide social opportunities in a small group setting. At times it is OK for them to be alone.
- Preplan outings. Keep time short.
- Provide a "supported recess" — planned activities with a friend.
- Include child in planning activities that are of interest to him/her (board games, puzzles, and computer games).
- Provide social skill classes that emphasize sharing, taking turns and handling of various social issues.

### **For STRABISMUS**

- Refer any student with possible turning in or out of the eye to a pediatrician, family practitioner or pediatric ophthalmologist. This may be detected during vision screenings.
- Assist student/family in developing incentives or rewards for success in following treatment program (patching).
- Advise/educate if surgery is indicated.

## **HEALTH RELATED CONCERNS cont.**

### **Poor Stamina**

People with PWS tire more easily and may fall asleep during the day. Morning is typically their optimal learning time, when energy level is highest.

### **High Pain Threshold**

Although the exact mechanism is unknown, persons with PWS have an altered response to pain. Many individuals will not complain of pain until an infection or injury has become very severe. Close assessment of pain along with listening to caregivers concerns is very important.

### **Lack of Vomiting**

About 2/3 of persons who have PWS, lack the ability to vomit. This has special implications in the treatment of poisoning. Most will not respond to the use of emetics, and treatment must focus on having their stomach pumped.

### **Scratching and Skin Picking**

These two behaviors are often seen in individuals with PWS and may be worse during times of stress. Combined with the higher pain threshold, these behaviors can result in severe tissue damage if not controlled.

### **Diabetes and Scoliosis**

Students with PWS are at greater risk of developing diabetes especially if their diet and weight are not under control. Students may also be at risk for the development of scoliosis.

### **For POOR STAMINA**

- Get person up and moving. Send on errand. Take a walk.
- Schedule high energy, mobilizing activity after lunch.
- Offer items /activities which stimulate large muscles and breathing (balloon blowing, party blowers).
- Provide scheduled rest time.

### **For HIGH PAIN THRESHOLD**

- Assess all complaints of pain.
- Evaluate injuries. Do not utilize pain as a primary source of assessment.
- Listen to caregivers when they report change in behavior, activity level and appetite (however — appetite is rarely affected).

### **For LACK OF VOMITING**

- In the case of poisoning, emetics are rarely effective, and repeated doses may cause toxicity.
- Often must perform pumping of stomach.

### **For SKIN PICKING**

- Provide activities to keep hands busy (coloring, computer time, play dough, hand-held games).
- Provide supervision. Reward and praise for not picking.
- Cover area with bandaide or similar covering.
- Keep nails short. Apply lotion liberally.
- Apply mosquito repellent before any walks or outside activity.

### **For DIABETES AND SCOLIOSIS**

- Be aware of risk factor for diabetes especially if child is obese.
- Be aware of risk factor of scoliosis — visual exam often not reliable due to obesity. Xray confirmation may be needed.
- Work with school nurse in supporting and assisting family in treatment plan if diabetes or scoliosis is present.

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