

Young Students with Prader-Willi Syndrome – Preschool Years INFORMATION FOR EDUCATORS

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).

The preschool years are an exciting time for little ones as they expand their ability to communicate and learn about the world around them. It is a time of growth and change. These youngsters strive to become independent and expand their problem solving skills. Most love to be leaders and helpers.

This can be a time when many young students with Prader-Willi syndrome (PWS) have minimal observable developmental delays. Speech and language is often the most common problem and challenge. Just like most non-disabled preschoolers, students with PWS may lack emotional and impulse control. It is a time when most students with PWS have more similarities than differences from their peers.

The Learner and the Learning Environment

Most parents and educators report that the early years of students with PWS are filled with many successes. Even though these students usually have about a 1-2 year developmental lag from their peers, they gain enormously from non-disabled peers who act as role models and motivators. Just like all preschoolers, they learn best in a smaller class environment. They also benefit from learning proactive behavior management strategies as well as healthy food choices. During these early years, many gains are often seen with speech and language as well as gross and fine motor skills.

There are broad ranges of learning abilities and learning challenges in students with PWS. A majority of students fall in the “borderline” range of a cognitive disability or mental retardation. Few function at a lower level and some may function in a higher range. Those in the higher range often demonstrate some kind of learning disability.

Learning Styles – Strengths and Weaknesses

Weaknesses and delays are often more pronounced in young children who have a greater degree of poor muscle tone (hypotonia) as well as a lower overall intelligence level. Relying solely on IQ scores at this age is doing the student a great disservice. There are situations where learning weaknesses have lessened once muscle tone and communication deficits have improved. Since many more infants and young children are receiving growth hormone therapy, less significant motor delays are beginning to be seen. Over time, more will be known how this therapy is influencing cognition and mastery of developmental milestones in these younger students.

Learning Strengths and Weaknesses – The Pre-School Student with PWS

<u>Learning Strengths</u>
<ul style="list-style-type: none">• Good Long Term Memory Skills Students with PWS are often good at recalling information that is stored in long-term memory. Educators may not see this initially but with repetition the outcome is often very successful. If information can be channeled from short term into long-term memory, students are able to use this strategy with learning for many years to come.
<ul style="list-style-type: none">• Receptive Language Children with PWS are often better at understanding what is being said than what they are able to communicate verbally. This is especially true of the pre-school child.
<ul style="list-style-type: none">• Visual Learners Most students with PWS are visual learners. Using visually based materials is often very helpful. This includes the use of pictures, videos and hands-on demonstration. This approach is especially true for those with expressive language delays.
<ul style="list-style-type: none">• Good at Puzzles Most students with PWS are very good at putting together puzzles. This can often be an activity that helps them to calm themselves and provide diversion. Even at a young age, they are often successful at completing multi-piece puzzles.
<ul style="list-style-type: none">• Multi-sensory Learners As with many preschoolers, young children with PWS benefit from learning material using a variety of different approaches. Caution must be used, however, in using items that may be edible since these students are likely to lack the control not to eat the manipulative.
<ul style="list-style-type: none">• Reading Skills Many young students with PWS are able to learn word recognition and reading skills at a young age. This may be related to the use of repetition and their strength with long-term memory skills.
<ul style="list-style-type: none">• Social and Friendly Many young children with PWS can melt your heart with their outgoing, sweet personalities. They take much pride in being “helpers” and are often seen saying hello to everyone. Even though they love to be around other children and benefit greatly from this approach, they are often seen engaging in parallel play. Like many preschoolers, they often lack skills in handling unsafe social situations and instruction in this area is needed for many years.
<u>Learning Weaknesses</u>
<ul style="list-style-type: none">• Expressive Language Younger children often experience expressive language delays. This is believed to be a result of poor muscle tone (hypotonia) as well as cognitive delays. You will see varying degrees of muscle weakness in young children with PWS. In some there are significant oral motor problems as well as speech apraxia. Most young children with PWS benefit from the services of a speech and language pathologist who diagnose and treat the problems identified. Motor and speech delays are covered in more detail later in this chapter. Finding a way for the young child to communicate is essential. One of the greatest sources of frustration and behavior escalation at this age results from their inability to communicate.
<ul style="list-style-type: none">• Poor Fine and Gross Motor Skills Because of problems with balance, coordination and strength, students with PWS often are delayed in developing gross motor skills such as running, jumping, climbing, skipping, catching and throwing. In addition, most of these young children also show weaknesses in hand strength. Drawing, cutting and printing are often frustrating activities. Occupational and physical therapists can provide interventions and adaptations that can help to facilitate success.

Learning Weaknesses

• **Poor Short-term Memory Skills**

Many students with PWS have poor short-term memory skills. This is especially true if material is presented to them in an auditory manner. This deficit is often misdiagnosed as inattentiveness or disobedience.

• **Sequential Processing Deficit – Difficulty Understanding Abstract Concepts**

Students who have PWS often have a deficiency in the intake, processing and response of information. When someone has this sort of processing problem, he/she will not necessarily learn “from experience”. If they do learn this, it can often take some time for them to do so. The timing and order gets all mixed up so the “if ... then” reasoning may be diminished or absent. This can be a key frustration factor for many people who support these students. Affected areas include:

- ❖ Trouble interpreting and executing multi-step problems (can easily become overwhelmed when the process seems too big)
- ❖ Difficulty summarizing and restating events
- ❖ Challenges in understanding time

Students with PWS frequently have difficulty planning or ordering steps to problem solve. They also have trouble with deductive reasoning. They tend to be “black and white” thinkers. Keep instructions simple. Assist with abstract concepts and summarizing. Like many students with cognitive disabilities, they have trouble generalizing or transferring what they learn in one environment to another. It is often helpful to use visual representation of time including calendars, “the hands on a clock”.

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The following factors do not impact all students. Remember, all students are individuals with their own personalities, strengths and weaknesses. What strategies work for some, may not work for all. What does not work one day – may work the next. The goal is to maximize success so that learning can be a positive experience for all.

Factors the May Influence Learning	Possible Strategies
<p><u>Fatigue and Daytime Sleepiness</u> Preschoolers with PWS often tire easily and exhibit daytime sleepiness. Because of weak chest and trunk muscles often compounded by obesity, they may not breathe as deeply as they should and may fall asleep. Many have disordered sleeping patterns. These altered breathing patterns can result in sleep disturbances and other respiratory problems such as apnea (the cessation of breathing for long periods of time during sleep).</p>	<ul style="list-style-type: none"> • Morning is typically an optimal learning/working time when their energy level is at its best. In a half-time program, placement in a morning program may be more beneficial. • When drowsiness is problematic, increase activity level – send on errand, take a short walk. • Some require a scheduled rest time.
<p><u>Structure and Routine</u> Most young students thrive on structure and routine. This seems to be especially reassuring to the preschool student with PWS. There is a strong need for routine, sameness and consistency in their environment. They do not handle changes well.</p>	<ul style="list-style-type: none"> • Foreshadow any changes and allow for discussion. With changes, there are often many misconceptions and worries. Keep change to a minimum. If possible, try to incorporate slowly • Show empathy and give positive praise when the student is faced with change. Reward flexibility. • Provide visual representation of change.

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<p><u>Delayed Toileting Skills</u> Most young students with PWS are delayed in their toileting skills. Because of weak muscles and other developmental delays, most have not mastered this skill by the time they enter an early childhood or other pre-school program. The preschooler must be ready both physically as well as emotionally to master this skill.</p>	<ul style="list-style-type: none"> • Work with parents in coming up with a program that best meets this child’s needs. • Avoid using food as a reinforcer • Stickers, party favors and other rewards are often motivators.
<p><u>Food Preoccupation/ Food Seeking/ Uncontrollable Food Drive</u> There is no predetermined age that young people with PWS begin to show food preoccupation or food seeking behaviors. However, parents and teachers often begin to notice a change in a student’s interest or obsession with food-related topics or issues in these early years. Unless calories and weight are monitored, many young children can rapidly gain weight and begin to show evidence of obesity. It is clearly a time when good nutritional habits should be established.</p>	<ul style="list-style-type: none"> • Be alert for signs of food seeking within the classroom and/or the school. • Keep food out of sight. • For those with a stronger food drive, snack items, student lunches and other food may need to be placed or locked in a cabinet or closet. • Educate and inform all school personnel including secretaries, lunchroom staff, bus drivers and custodians about this concern so that candy dishes and other treats can be removed or discouraged. • Avoid using food as a reinforcer for all students. Anyone can have a hidden food allergy. • Incorporate nutritional education and healthy food choices into the curriculum.
<p><u>Compulsive Tendencies</u> Many students with PWS are compulsive in nature. Most need to complete what they start. Unless accommodations are made, it is one characteristic that often causes escalation in anxiety and temper outbursts. Many students rely on rituals or specific ways of doing things. Some also show these tendencies in repeatedly playing with the same toy or drawing something over and over. This compulsive behavior may be used as a means of calming or a way to handle stress.</p>	<ul style="list-style-type: none"> • Start off with less work and add more if you feel the student will be able to complete the work in the allotted time. Most students want very much to please their teacher and feel very proud if they finish what they are assigned and have time for “extra credit”. • Modify worksheets so that only parts of the problems are present. In this way, the worksheet may look very similar to other students but contains fewer problems. • Utilize “strategic timing” for activities you know are difficult for this student to stop. “Strategic timing” is the scheduling of an activity just prior to another activity that you know is motivating to the student. An example might include scheduling computer time before snack, lunch or recess.

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<p><u>Perseveration or Obsessive Thinking</u> This is the tendency to get "caught" on one issue or thought to the point where it overshadows the main theme or learning experience. The student with PWS often will repeat the same question over and over even after they have been given the answer. They can talk about the same topic and get locked in to one issue making it difficult for them to move on to a different subject. It has been described as a “neurological switch that won’t turn off.” Perseveration can lead to increased anxiety and a loss of emotional control. This behavior is often seen in students of all ages.</p>	<ul style="list-style-type: none"> • Once you have answered the question, ask the student to restate what you said. This can validate both your understanding and the individual's understanding of the issue. • Assist in writing it down or provide some sort of visual that helps clarify what you just discussed. • Set limits; bring topic to resolution. "I will only answer this question 2 times. That was number 1”. Avoid power struggles or ultimatums. • Don't give more information than is necessary too far off from the event. Students often obsess about it.
<p><u>Tenuous Emotional Control</u> Persons with PWS often lack effective internal controls to regulate and manage their emotions. Even enjoyable activities can be stressful and offer anxiety. You may see extreme excitement when happy and extreme anger when sad. Any combination of stressful situations can lead to emotional "discontrol". Expressive language problems are often a source of emotional escalation in the younger student. These students seem to be extra sensitive and you may see a build up of emotions. Some students show their frustration by crying, yelling, and possibly lying on the floor and kicking. These are typical tantrum-like behaviors. For other students however, the display of emotion and frustration goes to a higher level. You may see swearing, aggression, destruction or self-injury. Some of these behaviors resemble rage attacks. Once control is lost, it typically takes a period of time before equilibrium is regained. Sadness, remorse and guilt often follow.</p>	<ul style="list-style-type: none"> • It is imperative to find a way for the student to communicate. Some are taught to use sign language or some other communicative means. Speech therapy is very important. • Validate feelings. Use charts and words to facilitate communication. Help the student to learn to use words not behavior when communicating emotions. • Prepare for changes. Discuss these changes in an area where the student is able to share feelings. • Clear up misunderstandings – write things down. Remember -most students are <u>visual learners</u>. • What is the trigger for the behavior? Are there any common factors? Time of day? Place? Certain class? Fatigue? • Learn to identify early signs of emotional distress. Have strategies in place to address issues before things get out of control. • Teach activities that are more appropriate for releasing frustrations. (Ripping paper, going for a walk...) in an area that is appropriate for this. Practice those activities before they are needed.
<p><u>Rigid Thought Process</u> Many students with PWS tend to be black and white thinkers. When life becomes “gray” they tend to worry and show increased anxiety. Terms like “later” or “maybe” can be problematic.</p>	<ul style="list-style-type: none"> • Make sure to follow through on promises. • Foreshadow but do not share information too far in advance in case the situation could change. • Plan for change. It is helpful to have a back up plan if needed... “If it is a sunny day we will walk to the park... If it is a rainy day we will have a special classroom activity.”

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<p><u>Social & Emotional Needs</u></p> <p>Most young children with PWS are very loving and sweet. They, like most preschoolers, are beginning to explore the world of making friends. They may have difficulty with some of the same issues of their peers. Taking turns, sharing and cooperative play are often challenges. It is common to see these young ones engaged in parallel play and have minimal interactions with other students. Speech problems and emotional sensitivity can add to their frustration in dealing with friends. Many of these students are very imaginative and can play for hours in their own make believe world.</p>	<ul style="list-style-type: none"> • Provide opportunities to teach and practice the basics of play etiquette including taking turns and sharing. • Facilitate small playgroups that meet for short periods. Select non-disabled peers to serve as role models. • Use books, stories, role playing and other creative means to help students understand and express their emotions as well as the emotions of others including interpreting nonverbal communication. • Help student to identify their emotions and the emotions of others. Emotions need to be labeled and validated.