

The Young Child with Prader-Willi Syndrome

Physical and Sensory Issues and Recommendations

By Janice M. Agarwal, PT, CNDT

Janice M. Agarwal, PT, CNDT is a pediatric physical therapist who is also the mother of a son with Prader-Willi syndrome (PWS). She is the author of a booklet, *“Therapeutic Interventions for the Child with PWS”* that is available for purchase from PWSA (USA). Website: www.pwsausa.org or call toll free: 1-800-926-4797.

Most children with PWS receive services from a physical therapist for many years. Today many infants and children now receive growth hormone therapy, so the degree of low muscle tone has improved. However many still face physical and sensory issues that require therapy and ongoing attention. Consult with a physical therapist for questions and recommendations about any of the strategies suggested.

Physical and Sensory Deficiencies:

All children with PWS are born with abnormal muscle tone and have physical and sensory deficiencies.

As a result, it is common to see:

- Trunk/upper and lower extremity weakness
- Skeletal abnormalities – scoliosis (lateral curvature of the spine) and hip dysplasia
- Sensory integration problems or deficiencies
- Oral-motor dyspraxia - difficulty in making and coordinating precise movements, which are used in the production of spoken language, which results in severe, persisting speech production difficulties.

SENSORY INTEGRATION PROBLEMS

Vestibular System: The vestibular system provides information on movement, gravity and changing positions (esp. head positions).

Some problems you may see include:

- Inability to use eyes and hands together in a coordinated effort.
- Poor balance. Clumsiness.
- Difficulty paying attention, concentrating and using reasoning
- Doing the same thing over and over again.
- Avoids movement or touch
- Low muscle tone/hypotonia
- Difficulty maintaining alertness; fatigue
- Unpredictable emotions. Difficulty handling changes
- Difficulty organizing self and using self-control
- Poor understanding of relationships to objects in space
- Poor understanding of what is said to them. (Not good at auditory processing)
- Difficulty sleeping.

What You Can Do to Help:

- Rocking, gentle bouncing, slow spinning in one direction
- Rocking in rocking chair or on rocking horse.
- Walking, running, hiking or swimming.
- Bounce on large balls and mattresses
- Rolling
- Roughhousing or wrestling.
- Somersaulting.
- Spinning on swivel chair, “Sit and Spin” toy, scooter board or tire swing.

Calming strategies for vestibular problems include slow, rhythmic, linear swinging or rocking, gentle, slow spinning in one direction. Gentle bouncing.

SENSORY INTEGRATION PROBLEMS

Proprioception: Proprioception input provides an unconscious awareness of our body, its position and its relationship to other parts as well as other people and objects. It helps us know how much force is needed for muscles to contract and move. Receptors are located in all of our joints. It helps calm the nervous system.

Some problems you may see:

- Clumsiness
- Exerts too much or too little pressure on objects
- Tantrums – throws self on ground.
- Affectionate – hugs tightly, sits on laps. Seeks enclosed/tight spaces for boundaries
- Poor writing skills; difficulty coloring in lines and/or stopping activities

What You Can Do to Help:

- Carrying heavier object – books, watering cans, suitcases
- Pushing and pulling items
- Crawling through tunnels/ boxes
- Hanging (from monkey bars)
- Jumping – on trampoline, mattress or air mattress
- Pounding nails; rolling play dough
- Swimming or extra bath time
- “Tug of War” with blankets or ropes
- Karate

SENSORY INTEGRATION PROBLEMS cont.

Tactile: Tactile input provides us with information about light touch, pressure, vibration, temperature and pain. This feedback system helps to develop body awareness and motor planning. It has complimentary protective and discriminative abilities. Tends to detect sensory dysfunction.

WARNING ABOUT LIGHT TOUCH: It is alerting; it may be uncomfortable. It can make a person feel threatened.

Some problems you may see:

- Not as sensitive to cuts, bruises, pain and temperature.
- Does not like having teeth or hair brushed.
- Drops things easily
- Flicks or shakes hands, rubs face or licks/chews on lips
- Picks at skin. Often does not tolerate Band-Aids.
- Needs extra personal space.
- Sloppy eaters and dressers
- The feel of new clothes or tags in them may be intolerable.
- Insects bites make some children crazy – scratch until bleeding.
- Touching activities uncomfortable – finger painting, glue on hands/fingers
- Don't always like tickling or petting

What You Can Do to Help:

BRUSHING AND JOINT COMPRESSION

- Provide DEEP PRESSURE/ heavy massage to the skin
- Obtain a soft brush from a therapist; brush perpendicular to the arm/leg.
- Start with the palm of the hand and go up the arm (like you are painting a wall up and down).
- Gently but firmly compress joint in to themselves.
- Go to the back. Brush up and down, side to side and horizontally.
- Move to the foot and legs.
- **DO NOT** brush – face, neck stomach, chest or genital areas.
- Start with a 2 week trial.
- Determine what behaviors you want to measure; see if there is improvement.
- Do brushing 4-5 times per day. If you see positive changes – continue for 1 month, then reduce.
- This is SHORT TERM TREATMENT – it is not forever.
- May need a “tune up” after times of stress – illness, growth spurts, holidays...

ROLLING

- Use simple wooden rolling tool
- Roll up and down back from neck to bottom.
- Perform 4-5 times per day.
- See improvement – continue for 1 month then reduce.
- Maintain treatment daily

ORAL MOTOR PROBLEMS

Eating is one of the most sensory intensive activity. Oral motor activity – sucking creates a calm, self-regulating state. It supports head, neck and trunk development.

Some problems you may see:

- Difficulty sucking or blowing through a straw.
- Teeth grinding; inappropriate use of tongue or lips
- Poor suck and swallow
- Prefers certain types and texture of foods

What You Can Do to Help:

- Blow bubbles, whistles, party blowers
- Offer variety of food textures and contrasts:
 - Cold or frozen foods – crushed ice chips
 - Warm soups/drinks, oatmeal, cream of wheat
 - Chewy bagels, dried fruits, fruit roll-up, gum
 - Crunchy pretzels, vegetables, apples
 - Sour/tart foods – cranberries, lemon-lime wedges, sour sprays
 - Sucking – use straws, lollipops
 - Tugging/pulling – beef/turkey jerky, licorice

CALMING TECHNIQUES

These strategies may help to relax the nervous system and reduce exaggerated responses to sensory input:

- Warm or tepid bath
- Deep massage, back rub, deep brushing or roller
- Snuggling in sleeping bag, beanbag chair or pillow
- Swinging back and forth; slow rocking, hugging
- Hide out, fort or quiet corner. Reduce noise & light

SLEEPING TECHNIQUES

- Warm bath or shower at bedtime
- Use body pillows or sleeping bag
- Try different types of pajamas – loose/tight, silky/cotton – see what child prefers
- Swaddle infant, heavier weight blanket sleepers.
- Dark blinds or shades to minimize light
- Back rubs, brushing, rolling.
- Clean, uncluttered room.



For more information, contact Prader-Willi Syndrome Association of WI Inc
Phone: (920)733-3077 Website: www.pwsaofwi.org