

The Six-Month Update Booklet

We want to make sure that as your child grows and changes, you have quality, up-to-date information available to you that will assist you and your care team in making the decisions and choices necessary to help your child be the very best that he or she can be. In this E-Book, you will find a collection of printable articles that cover many topics relating to PWS in the growing child. This collection should help you address many of the new situations you will face in the coming months. We hope this collection is just what you need to carry you into the next stage of life with your child.



Anakin Timmons, living with PWS


Prader-Willi
SYNDROME ASSOCIATION | USA
SAVING AND TRANSFORMING LIVES



Greetings!

Can you believe it has been six months or more since you first reached out to PWSA | USA for support or information regarding your child with Prader-Willi Syndrome? We sincerely hope that the materials you received and the connections you have made through PWSA | USA have been helpful as you've begun to walk this life-long journey with your child. Because PWS is a multi-phasic syndrome, sometimes it seems just as you've settled in to a comfortable pattern, everything changes again! We want to make sure that as your child grows and changes, you have quality, up-to-date information available to you that will assist you and your care team in making the decisions and choices necessary to help your child be the very best that he or she can be.

In this E-Book, you will find a collection of printable articles that cover many topics relating to PWS in the growing child- From oral care to preschool choice, this collection should help you address many of the new situations you will face in the coming months. Please feel free to print this amazing collection and store it in the Package of Hope box that you received, for convenient access. We hope this collection is just what you need to carry you into the next stage of life with your child.

And....We would love to hear from you! Please don't hesitate to call or email if there are other topics of concern that you would like covered in the collection. Let us know how you liked the Package of Hope, how you are doing with your Parent Mentor, and anything else you would like us to know about your experience with our programs and services. Your opinions today will affect the experiences of new parents in the future as we continue to expand and improve our New Parent Support programs.

It has been a pleasure to "meet" you and your child, and we consider it a great honor to be allowed to be a part of your lives as they grow and progress. Please remember you can call our main number, (941-312-0400) or contact myself or any of our family support team at the information below if you ever have questions or concerns. Look for more information coming from PWSA | USA in the future!

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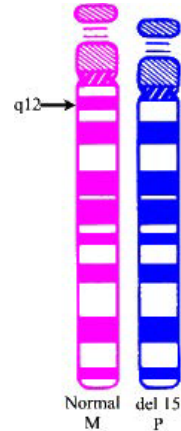
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THE GENETICS OF CHROMOSOME 15

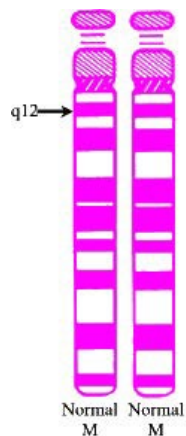
Deletion - 70%

A deletion including the q12 band (arrow) of chromosome 15 is contributed by the father and observed in about 70 percent of the subjects with Prader-Willi syndrome. The imprinted 15q11-q13 chromosome region includes genetic material (DNA) that is normally active (paternally expressed) on the father's chromosome 15 and inactive on the chromosome 15 inherited from the mother. Therefore, the paternally expressed genes from this region of chromosome 15 are missing in the deleted region of chromosome 15 and Prader-Willi syndrome occurs.



Laboratory Testing

This typical common deletion of chromosome 15 can be seen at the microscope level and detected with high resolution techniques. The deletion is confirmed by fluorescence in situ hybridization (FISH) using DNA probes from chromosome 15.



Maternal Disomy or UPD - 25%

In approximately 25 percent of subjects with Prader-Willi syndrome, both chromosome 15s are inherited from the mother (maternal disomy or UPD) and no chromosome 15 is present from the father. The missing chromosome 15 from the father which contains the active or paternally expressed genes required for normal development causes Prader-Willi syndrome in individuals in which both chromosome 15s are inherited from the mother.

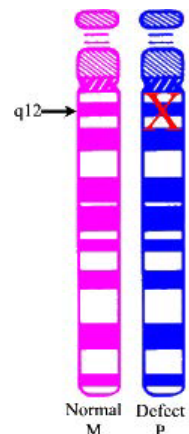
Laboratory Testing

Genetic testing is required to identify maternal disomy 15 or UPD using DNA obtained from the parents and the person with Prader-Willi syndrome.

Polymorphic DNA markers from chromosome 15 are studied to identify whether both chromosome 15s are from the mother.

Imprinting Defect - 5%

A few individuals with Prader-Willi syndrome (up to 5 percent) may have an imprinting defect inherited from the father of the chromosome 15 involving the q11-q13 region. This imprinting defect will not allow the normal expression of genes that are active on the chromosome 15 from the father and needed for normal development. Prader-Willi syndrome then occurs. Rarely these imprinting defects on the father's chromosome 15 may be inherited with a potential 50 percent risk for having additional children with Prader-Willi syndrome.



Laboratory Testing

To identify imprinting defects on chromosome 15 requires specialized laboratory techniques available in only a few genetic laboratories conducting research in Prader-Willi syndrome.

FIRST SMILES: DENTAL TIPS AND TRICKS

People who have Prader-Willi syndrome have decreased yet thick and sticky saliva, which makes them prone to cavities and increased tooth wear. Limiting intake of sugary foods and regular brushing and flossing contribute to a child's dental health, as well as regular visits to the dentist. Your partnership with the dentist will help ensure healthy teeth. Proper dental care begins before a baby's first tooth appears. Below are some tips to keep kids' teeth healthy and strong beginning at birth.



Before teeth appear

- Wipe gums after feeding with gauze, a soft wet washcloth or finger toothbrush pads. This will help get rid of the sticky coating called plaque that can cause tooth decay.
- Getting your baby used to having his mouth cleaned as part of his daily routine should make it easier to transition into tooth brushing later on, too.
- For children who are fed via feeding tube, oral health is just as important to keep the mouth clean.

When teeth first appear

- As your child's teeth start to appear, brush teeth twice a day with water and a SOFT-bristle toothbrush.
- Gently brush on the inside and outside of each of your baby's teeth, as well as her tongue to dislodge bacteria that can cause bad breath.
- Schedule first dental appointment within six months after the first tooth erupts, or by their first birthday, whichever comes first.

3-5 years

- Start using fluoride toothpaste at age 3. Use only a pea-sized amount on a SOFT-bristle toothbrush. Make sure your child spits it out after brushing.
- Start visiting your dental team every 6 months.

6-9 years

- Begin flossing as soon as the teeth touch.
- Parents should assist with brushing until the child is able to practice good oral care independently. Typically, assistance is needed until early middle school. Teach and encourage the child to do it, with the parent going over the areas missed.

TIPS AND TRICKS

- Only use a SOFT-bristle toothbrush to avoid wear of teeth and gums.
- Always provide supervision with toothpaste use to prevent the child from overeating it. Toothpaste in small amounts will not hurt you.
- Children should limit sugary and sticky foods and drinks to protect against tooth decay.
- For school-aged children, tooth brushing after lunch may help aid in keeping teeth clean. As your child's permanent teeth grow in, the dentist can help seal out decay by applying a sealant to the back teeth, where most chewing occurs. This protective coating keeps bacteria from settling in the hard-to-reach crevices of the molars.
- If your water supply is not fluoridated or if your family uses purified water, ask your dentist if fluoride supplements should be considered.
- Ask for your dentist's advice on when and if to start using mouthwash. If swallowing is problematic, a Q-tip can be dipped in mouthwash and applied to the child's teeth.



GETTING CREATIVE

- Oral health care is just like the other personal care routines you help your child with. It takes patience and sometimes creativity. Try to use the same time, place, and position when possible, to create a daily dental routine. Start with small steps, until the child gets used to you working in their mouth.
- If tooth brushing causes distress, experiment with positioning, times, places, distractions, and rewards:
 - A small child may be cradled in the arms or brushing can be done standing behind the child with the head supported or from a seated or kneeling position.
 - Sometimes it takes two people to brush teeth one to brush while the other distracts, holds hands, or encourages.
 - Brushing teeth in the bath may be easier.
 - Distractions such as music, singing or their favorite television show can be used.
 - Reward schemes (Tooth brushing charts can be downloaded online, and incentives put in place to encourage cooperation).
- Let your child pick out their toothbrushes, so that they get excited about brushing their teeth. You can also let them choose new toothbrushes monthly to keep them interested. There are many with movie and cartoon characters to choose from. Making it less of a chore and more like a fun task will make your kids more likely to brush regularly.
- Toothbrushes and floss holders can be adapted to fit your child's specific needs. For instance, enlarge the toothbrush handle with a pencil or bicycle grip. Specially adapted toothbrushes are also available for purchase online or try a battery-operated toothbrush for toddlers or older.
- Get kids in the habit of flossing. Flossers are available in bright colors and even animal shapes to keep kids interested.

- Keep in mind, the tooth fairy loves to bring special toothbrushes and toothpastes in new flavors, which kids love!

HELPFUL PRODUCTS

- For dry mouth symptoms, Biotene dry mouth product including toothpaste, mouth spray, gel and gum can help stimulate saliva flow. A humidifier at night can be used to keep the air full of moisture, which can also help alleviate dry mouth symptoms.
- Phillips Sonicare For Kids toothbrush for kids ages 4-10 utilizes sonic technology, featuring two kid-friendly power levels, two brush head sizes and a Kid Timer to help kids reach the recommended two-minute brushing time.
- Brush DJ: Perfect for older children, this free app will play random music from your phone for a set period of time in order to keep you brushing longer. The app also reminds you about dental checkups and when to replace your toothbrush.
- Twooth® Timer is the award-winning, mom- invented tooth brushing timer to help children & adults brush their teeth for the recommended two minutes.
- Countless downloadable tooth timer apps for kids are available to download with many characters and themes to choose from.



DRAWING BLOOD IN CHILDREN WITH PRADER-WILLI SYNDROME

Individuals with Prader-Willi syndrome may have difficulties with blood draws. The hypotonia and increased subcutaneous fat may make finding landmarks more difficult for the phlebotomist. These are some hints that may help.

If your child has had difficulty in the past with blood draws, warn the lab tech that an experienced person will be needed because your child is a “hard stick.” The usual guideline is that the phlebotomist should try twice. If he or she is unsuccessful after two tries; then someone else should try. Do not be afraid to ask up front for the most experienced individual. That individual may not be present at the time, so depending on what labs are needed, you might ask if the blood draw can be postponed until that person is present.

In most children’s hospitals, the phlebotomist would have the most experience with little children. On occasion the parents might wish to ask for a neonatal nurse to come down for the draw. In smaller hospitals, the parents might be able to request a pediatric/ newborn nurse, who may have more experience than the phlebotomist, and the hospital may be better able to comply with that request.



- Request a prescription from your doctor for EMLA cream to help numb the area. EMLA should be placed on the skin 45 minutes prior to lab draw.
- Give your child plenty to drink several hours before the visit to the hospital/doctor. If the weather is cold, bundle them warmly on the way for the blood draw. Ask for warm packs to place on the blood draw site.

FLU SEASON IS HERE – WHAT SHOULD WE DO?

by Janalee Heinemann



Should we stop growth hormone treatment when our child has a respiratory problem?

There is no evidence that stopping GH during respiratory illness would have any benefit or not. (Jennifer Miller, M.D.) Although GH has been mentioned to be contraindicated in cases of severe obesity and respiratory compromise, it does not mean that GH will worsen the symptoms of common respiratory illnesses; therefore, I agree with Jennifer, GH should not be discontinued. (Moris Angulo, M.D.)

Should my baby get the influenza vaccination? What age should he/she start?

The immunization rate in kids is about the same each year. It is down in adults, so it is important that both the parents and children get vaccinated. CDC recommends vaccine after 6 months of age especially in individuals at risk. all individuals with PWS would fit in the high-risk category and should be vaccinated. There are circumstances where the individual needs two vaccinations delivered 4 weeks apart. (James Loker, M.D.)

All infants starting at 6 months of age are eligible for the flu shot. they will need a 2nd vaccine 1 month later as this is the first year they are being vaccinated. Younger infants can have some environmental protection if all adults with whom they come in contact receive the flu vaccine each year. As long as vaccine supplies are available, the vaccine should be given. This is different than the RSV Synagis monthly for selected infants. (Marilyn Driscoll, M.D.)

Insurance is denying paying for Synagis to prevent RSV. Aren't our infants more at risk for RSV due to their increased respiratory risk? Shouldn't insurances be required to pay?

Synagis is indicated for the prevention of serious lower respiratory tract disease caused by respiratory syncytial virus (RSV) in pediatric patients at high risk of RSV disease such as infants with broncho-pulmonary dysplasia (BPD), infants with a history of premature birth (≤ 35 weeks gestational age), and children with significant congenital heart disease. All other use would be

considered off label. Synagis costs about \$1,000 per shot every month during RSV season. Insurance companies do not like to pay for an expensive medicine that is used off label. It is possible that if the cost comes down, we may see more off label use of the medicine. It would take studies that would show not only safety of the shot but also a significant benefit to convince drug companies to lobby FDA to include PWS in the indications. With the small numbers of

infants with PWS it would be difficult to demonstrate a significant benefit. We need to keep an eye on this, but in the meantime, we need to emphasize good hygiene in all infants with PWS. RSV is spread by contact. Parents need to wash their hands and use alcohol cleanser to reduce not just RSV but all infections in infants with PWS. (James Loker, M.D.)

RESPIRATORY PROBLEMS IN PWS

by James Loker, M.D.

Several recent articles continue to show that individuals with Prader-Willi syndrome are at risk for respiratory problems. Problems of central hypoventilation/apnea and obstructive apnea in Prader-Willi syndrome have recently been investigated.

Central hypoventilation is a disorder of decreased breathing rate or depth, particularly during sleep. This usually causes problems with daytime sleepiness and if significant can cause problems with elevated blood pressure in the lungs. Individuals with Prader-Willi syndrome may be at increased risk for this due to decreased muscle tone and mass, excessive obesity, and possibly decreased neural drive for breathing. Studies have shown some individuals with Prader-Willi syndrome have decreased depth and rate of breathing.



Central apnea means the complete cessation of breathing during sleep. There are several studies that show an alteration in the response of some individuals with Prader-Willi syndrome to chemicals that would normally increase breathing. Both receptors in the body and the area of the brain that is involved with breathing are being investigated. The clinical significance of central apnea is still under investigation.

Obstructive sleep apnea is well known to occur in Prader-Willi syndrome as well as in other syndromes with hypotonia (poor muscle tone) such as Down syndrome. It is seen in 2% of the normal pediatric population as well. This results when the individual is trying to breathe while asleep, but due to obstruction in the airway no air enters the lungs. The obstruction can occur anywhere from the nose to the small airway passages in the lungs. These individuals usually have loud breathing and snoring associated with periods of quiet where no air movement is noted. Untreated obstructive apnea can have serious complications including death.

Other problems that can cause respiratory difficulties in the young can be chronic stomach reflux and aspiration. Although the lack of vomiting is felt to be prominent in Prader-Willi syndrome, reflux has been documented and should be investigated in young children with chronic respiratory problems. Individuals with obstructive apnea are at more risk for reflux as well.

The American Academy of Pediatrics has recently set forth guidelines for diagnosis and management of obstructive sleep apnea. The guidelines suggest that all children be screened with history of snoring or other evidence of airway obstruction. Your physician may wish to obtain a sleep study if there is excessive sleepiness, significant obesity, or before surgery. In those individuals with a positive history, a sleep study is performed where breathing patterns, heart rate, oxygen levels and air movement are recorded. If the test is positive, further evaluation may need to be performed to individualize the treatment. The primary treatment as suggested by the guidelines would include tonsillectomy and/ or adenoidectomy or CPAP (Continuous Positive Airway Pressure), where the individual wears a mask at night to keep the airway open.

Frequently obstructive and central apnea may occur in the same patient. This is probably true in the majority of individuals with Prader-Willi syndrome with respiratory problems. Both obstructive and central apnea can be evaluated by a sleep study.

In summary, individuals with Prader-Willi syndrome are at risk for respiratory problems, most commonly obstructive apnea. If any child has symptoms of obstructive apnea, a sleep study should be obtained. The role of central apnea in Prader-Willi syndrome is under investigation.

A special thank you for their contribution to this article goes to:

Dan J. Driscoll, Ph.D., M.D.

Merlin G. Butler, M.D., Ph.D.

David M. Agarwal, M.D.

THINKING AHEAD—THE POWER OF EARLY INTERVENTIONS



There is a wide consensus that early interventions for children with developmental disabilities can result in significant improvements in cognitive, academic, and social outcomes. This is certainly true for children with PWS. The PWSA | USA publication, “Therapeutic Interventions for the Child with PWS,” states:

The first years of life are a critical time in a child’s development. All young children go through the most rapid and developmentally significant changes during this time. During these early years, they achieve the basic physical, cognitive, language, social and self-help skills that lay the foundation for future progress. These abilities

are usually attained according to predictable developmental patterns. Because children with Prader-Willi syndrome face delays in all areas of motor development, Early Intervention is highly recommended (p.6).

For children with PWS, basic early intervention services include physical therapy, speech and language therapy, and occupational therapy. As a parent of a young child with PWS, we highly recommend connecting with professionals who can help you implement – at home and school – these critical early intervention services. We also encourage implementing early practices and strategies in the home to help manage the challenges of weight and behavioral issues which we often see in children with PWS. These practices will help you build a strong foundation of support for your child as they grow and mature. Here are some tips to get you started:

Healthy Eating Habits

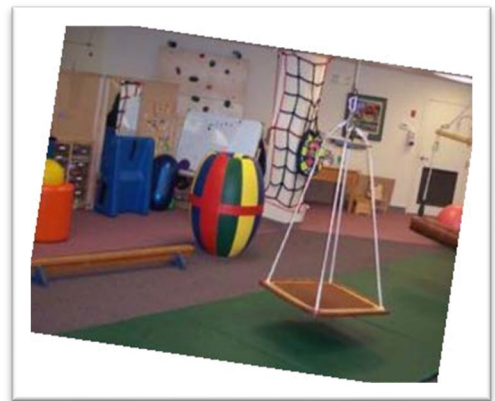
Caring for a child with PWS is more than simply about counting calories. It is also about making sure your child is consuming healthy calories that will contribute to more healthy development. Fortunately, earlier diagnosis and education can make it easier for children:

- To adapt to a specialized diet at a younger age.
- To accept that food is available only at certain times.
- To learn about making healthier food choices.
- To help with age-appropriate cooking activities to learn about calories, portion sizes, and preparation of food. *Note: Depending on a child's food drive, this might not be an option for every child.*

This can all be an important part of the strategy to prepare a family – including siblings – for what may need to be done to keep a child with PWS safe as they grow into adulthood.

Although it may be necessary or helpful to lock food in the future, early education and behavioral management are critical components of early intervention for a child with PWS.

Professionals like Dr. Jennifer Miller and others are constantly enhancing and advancing our understanding of what healthy eating habits look like for a child with PWS. So, with the support of your child's medical team, keep informed about the latest recommendations for how to implement healthy eating habits for everyone in your home. Over the long run, this will help your child to appreciate healthy eating and optimize their ability to be part of their own well-being. It will also better prepare a child for food related modifications (such as locking a refrigerator, cabinets, etc.) should those be necessary in the future.



Behavior Management

Children with PWS are most successful behaviorally when a positive behavioral strategy is developed and consistently employed. This is a tried-and-true strategy that is effective for people with PWS of all ages. A positive behavioral strategy uses incentives and rewards to move a person successfully through the day by mixing preferred and nonpreferred activities.

For example:

If Tommy, who has had a problem getting up and ready for school in the morning, is able to accomplish this task in the designated half hour time period he will receive a sticker. If he earns 10 stickers during the week, he will be able to pick a movie he wants to watch.

In this example, Tommy's desire to watch a movie of his choice is used to motivate him to successfully accomplish a non-preferred activity – getting ready for school on time. You can find many helpful resources on positive behavioral strategies by searching online. If you are working with a counselor, social worker, or school professional they also might be able to assist you with ideas. And don't forget to contact PWSA | USA for helpful behavioral resources (video and written) for supporting people with PWS. The more you know the better your plan will be! Implementing the concept of home positive behavioral support early in your child's life will prepare them for similar support programs at school and other settings. It will also help you to be a resource to help other professionals understand what kind of behavioral support works best for your child.

The Good News: It Works for Everyone!

Remember, healthy eating habits and positive behavioral supports can work for everyone in the house – including parents and other children. So, you have nothing to lose by doing some research and experimenting with what works best for your home in these crucial areas. In the long run we believe it will help you to create a healthier and happy family!

Evan Farrar

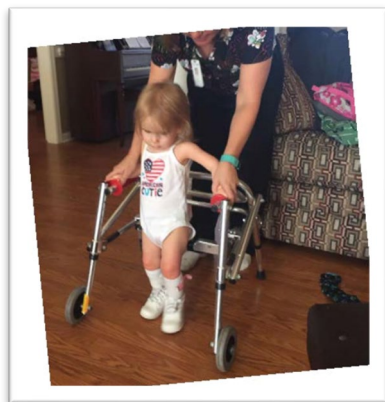
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Clinical Advisory Board

THE YOUNG CHILD WITH PRADER-WILLI SYNDROME: PHYSICAL AND SENSORY ISSUES AND RECOMMENDATIONS

by Janice M. Agarwal, PT, CNDT



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:

- 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 repeatedly
- 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
- Bouncing 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:

- include slow, rhythmic, linear swinging or rocking, gentle, slow spinning in one direction.
- Gentle bouncing.

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 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



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
- Insect 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 joints into 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 activities. 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

FIRST STEPS TO PRESCHOOL

There are many things you can do to make sure your little one is ready for the big “P” (preschool), and many of them can be started much earlier than you think! This simple “to do” list will help you get a head start on getting your little Einstein ready to take the preschool world by storm.



Give your little one opportunity to interact with other children from an early age.

Cooperative play, taking turns, and sharing are difficult for nearly all small children, but more so for those who have had less time to practice these important skills. And remember, this is just practice time! Don't expect your toddler with PWS to become a Master of Social Interaction!

It takes years to hone and refine these abilities.

Encourage your child to interact with their environment.

Little ones with PWS often have delays in both fine and gross motor skills. Many activities that little children love are great ways to practice those skills! From finger-painting with applesauce on their highchair tray, to dropping clothespins into a jar (with supervision, of course- these are “mom-and-kid” activities), to crawling over and around a pillow maze on the floor, every interactive activity encourages movement, curiosity, and dexterity. Ask your child’s PT/OT for some fun suggestions to wake up little minds and bodies!

Maintain an organized and predictable schedule whenever possible.

It does not have to be based on the clock, but a predictable “order of operations” relieves anxiety and gives your little one a sense of consistency and security. Keep big changes to a minimum, and always give some age-appropriate warning and explanation when they are unavoidable. But it’s important to note that occasional, small switch-ups in routine can actually be good. Flexibility is a crucial life skill that you can begin building now!

READ to your child daily, even when they seem too young to understand.

This is a wonderful opportunity to learn dozens of valuable skills; listening, naming (pictures, letters, etc.), reciprocal conversation, predicting (what do you think will happen next?) rhyming, print recognition, etc., etc., etc.!! A recent study showed that children who are read to from infancy have better language comprehension, more expressive vocabulary, and higher cognitive scores as young as two years of age!

If your child is not in daycare, begin leaving them with a trusted adult for short periods of time (occasionally).

Knowing that you ALWAYS come back is especially comforting to a young child. Some parents even invent a special goodbye/hello ritual that is the same every time you leave and return. (a special kiss, a crazy handshake, a chosen phrase, etc.). This ritual can help comfort your child and reassure them that although you may leave them for a while, you always return. When started at an early age, this ritual can provide a reminder of your dependability through an entire childhood of comings and goings!



CHOOSING A PRESCHOOL

Choosing the best preschool for your child with PWS can seem like a challenge, but with these key points to consider, it may be easier than you think.

Always remember that your little one is a child first, and a child with PWS second. Many of the criteria you would use to choose a preschool for a typical child are the same as those you would use to choose a program for your young child with PWS. A warm, friendly staff, a child-friendly, welcoming atmosphere, and a structured, yet flexible environment are positive indications - But, before you sign up, here are some questions you may want to ask:

- What is the child to teacher ratio in my child's class/level?
- Is my child's teacher certified? Does she have special needs training or certification?
- Are there Support Professionals available if my child needs extra help? (PT, OT, Speech, paras, or aides, etc.)
- Does my child's classroom have a sensory area, or is there a sensory room in the school?
- What is the system for parent-teacher communication?
- Is this school willing/able to provide and enforce a no food classroom environment for my child?
- Is there a designated person to assist with toileting/diapering needs?
- Does my child's classroom have a "quiet area" that my child can access if they become fatigued or over stimulated?
- Does the school or teacher have knowledge of or experience with PWS (or are they willing to learn)?
- What does this school consider the goal or purpose of the preschool experience?

Never be embarrassed to ask questions- and take notes! As you ask these questions, you will clarify and define your own hopes and expectations for your child's preschool experience, as well as receive tangible criteria for choosing the program that is just right for your family, and your child with PWS.

Young Students with Prader-Willi Syndrome – Preschool Years

INFORMATION FOR EDUCATORS



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 are 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

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.

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.

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.

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.

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.

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.

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.

LEARNING WEAKNESSES

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.



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.

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

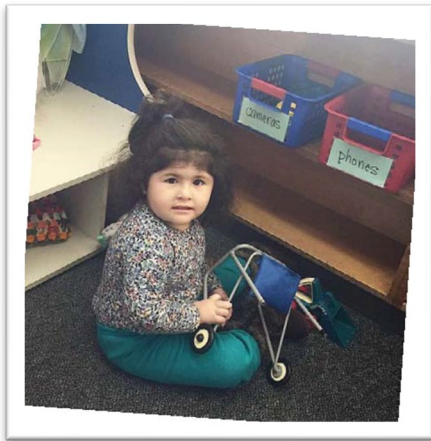
timing and order get all mixed up so the “if ... then” reasoning may be diminished or absent. This can be a key frustration factor for many people who support these students.

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”.

YOUNG STUDENTS WITH PRADER-WILLI SYNDROME – PRESCHOOL YEARS



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.

Fatigue and Daytime Sleepiness

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).

Possible Strategies:

- 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.

Structure and Routine

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.

Possible Strategies:

- 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

Delayed Toileting Skills

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.

Possible Strategies:

- 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

Food Preoccupation/ Food Seeking/Uncontrollable Food Drive

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.

Possible Strategies:

- 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

Compulsive Tendencies

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.

Possible Strategies:

- 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.

Perseveration or Obsessive Thinking

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.

Possible Strategies:

- 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.

Tenuous Emotional Control

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 buildup 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.

Possible Strategies:

- 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 visual learners.
- 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.

Rigid Thought Process

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.

Possible Strategies:

- 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 backup 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.”

Social & Emotional Needs

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.

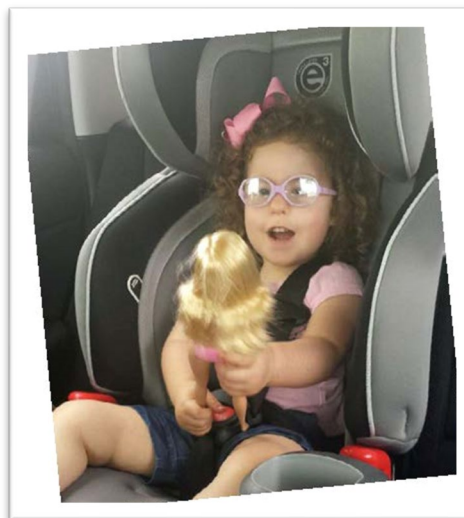
Possible Strategies:

- 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.

CAR SEAT SAFETY

by Todd R. Porter, MD, MSPH, CPST

Despite significant progress in preventing unintentional motor vehicle occupant injuries and fatalities, our society still has a way to go in shortening the gap between child passenger safety laws and best practices. “It is important to note that every transition (from rear-facing, to front facing, to booster seats, to seat belts) is associated with some decrease in protection; therefore, parents should be encouraged to delay these transitions for as long as possible.” [Pediatrics 2011; 127:788-793]. Muscle tone is defined as the amount of tension or resistance to movement in a muscle. Given that children with Prader-Willi syndrome have low muscle tone, they are even more susceptible to the mechanical forces on the body during a motor vehicle crash, and therefore will benefit from delaying each transitional stage (as outlined below) for as long as possible.



Stage 1: Rear Facing

The American Academy of Pediatrics, Bright Futures, and Safe Kids recommend that all children from birth through age two ride rear-facing until they reach the highest height or weight allowed by their car seat manufacturer. This recommendation is based in part on research which showed that children ages 12-23 months of age were five times less likely to be killed or seriously injured when rear-facing, compared to forward facing. Even greater side impact protection was noted in the rear facing direction. Facing toward the rear helps keep the child's spine in line by spreading the crash forces evenly over the entire head, neck, torso, and pelvis. In the forward-facing position, the child's disproportionately large head size, and low muscle tone, lead to increased forces on the weaker neck muscles, ligaments, and spinal cord.

Also note that bulky clothing, including winter coats and snowsuits, can compress in a crash and leave car seat straps too loose to properly restrain your child, leading to increased risk of death or injury. For optimum safety, dress your baby or toddler in thinner layers of clothing and tuck a coat or blanket around the buckled harness straps if necessary.

Selection:

Infants and toddlers may be restrained in the rear facing position using either an Infant Carrier or Convertible style car seat until they reach the upper weight or height limits set by the manufacturer. The best car seat is one that fits your child, fits your vehicle, fits your family budget, and one that you will use correctly every time.

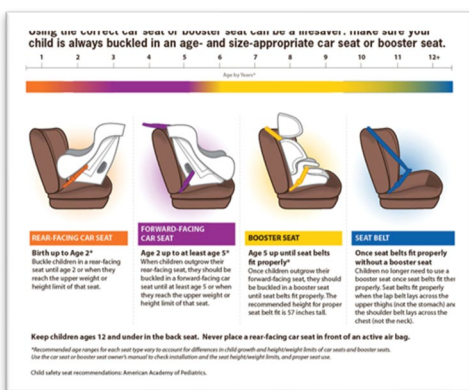
Stage 2: Forward Facing

Once the child is over two and exceeds the maximum rear-facing weight and height limits set by the car seat manufacturer, they can be turned to the forward-facing position. Research shows that in children four and under, forward facing seats reduce injury by 80% compared to seat belts alone.

Selection:

A child can be restrained in the forward-facing position in a Convertible Car Seat, a Forward-Facing Harness Seat, or a Combination Seat with Harness. Safety advocates recommend keeping the child restrained in the forward-facing harness restraint as long as allowed by the car seat manufacturer. Maximum weight limits on harness system seats range between 45 and 85 pounds. Parents of children with PWS who have not reached the appropriate height to safely graduate to a belt positioning booster seat should strongly consider a higher weight approved harness seat. A list of higher weight harness seats is available at <http://www.hmhb-mt.org/docs/BoosterAlternatives.pdf>

Stage 3: Belt Positioning Booster Seats



A booster seat uses no harness; it uses the vehicle's lap and shoulder belts only. When using a booster seat with no integrated harness, be sure the seat belt (including the shoulder belt) is properly buckled and positioned. Safety advocates recommend that once children outgrow their forward-facing harness seat as defined by the car seat manufacturer, they should use a belt positioning booster until they can pass the "5 Step Test". (If you answer "no" to ANY of these questions, the child must remain in a booster seat. If the child can pass the 5 Step Test in some vehicles, but not others, the child must remain in a booster seat

in those vehicles that do not pass.)

1. Does the child sit all the way back against the automobile seat?
2. Do the child's knees bend comfortably at the edge of the automobile seat?
3. Does the shoulder belt sit between the child's neck and shoulder?
4. Does the lap belt stretch across the tops of the thighs (not the tummy)?

5. Can the child remain seated in this position for an entire car trip, both while awake and asleep?

The ability to pass the “5 Step Test” generally occurs when the child is nearing 5 feet tall. It is generally recommended that children continue to use a booster past age 8 and possibly to age 12 or older. If your child with PWS is not able to pass the 5 Step Test but has exceeded the weight recommendation set by the manufacturer of their booster seat, please contact a child passenger safety technician trained in restraining children with special health needs.

Why Aren't Seat Belts Good Enough?

Booster seats work by positioning the lap and shoulder belts over the bony structures of the child's pelvis and shoulder, and not the vulnerable abdomen and neck. Research shows that premature graduation into a poorly positioned vehicle seat belt can cause head, spinal cord and abdominal organ injuries. Children ages 4 and up who are restrained in a belt positioning booster seat are 45% less likely to be injured in crashes than those restrained in a vehicle seat belts (this increases to 68-82% less likely to sustain injuries when the crash involves a side-impact). An Australian study found that children 12 and under were 7 times more likely to suffer spinal cord injuries when restrained in a seat belt alone, when compared to children over age 12.

Selection:

Booster seats come in many styles; backless, high back, combination, etc. While there are many different brands of booster seats, not all of them give equal fit and performance. The Insurance Institute of Highway Safety maintains a list of booster seats in four separate categories- Best Bets, Good Bets, Not Recommended, and Check Fit... This list can be accessed by going to www.iihs.org/iihs/ratings/child-boosters. Please review this list before considering your next booster seat purchase.

Stage 4: Adult Seat Belt

Once the child passes the 5 Step Test, they are ready to use the vehicle lap/shoulder belt system in that vehicle. It is well proven that when used correctly, seat belts reduce the risk of fatal injuries to front-seat passengers by 45% and the risk of moderate to critical injury by 50%. The CDC currently recommends that children remain as rear seat passengers until they are over age 12 and able to use a vehicle seat belt restraint system correctly. Research shows that restraining a child in the rear seat reduces the risk of injury by more than 40%.

Installation

Sadly, studies show that approximately 80-90% of all child safety restraints are improperly installed. Safety advocates strongly recommend having your child restraint inspected by a NHTSA trained Child Passenger Safety Technician whenever you purchase a new car seat, or transition between any of the above-mentioned stages. To find a Child Passenger Safety Technician in your area, visit www.seatcheck.org.

STRESS MANAGING STRATEGIES

by Lisa Graziano, M.A., MFT

We know it intuitively, but new research by PWS experts Janice Forster, M.D., Linda Gourash, M.D. and Marjorie Royale confirms it: stress is significantly higher among parents and other caregivers of people with PWS than it is in the population at large. It's imperative, then, that we utilize strategies that will reduce and manage our high levels of stress so that we have adequate energy to sustain a calm and neutral response to our child with PWS, so that we are emotionally available to all of our children and spouse, and so that we stay healthy and enjoy the same high quality of life we work so hard to provide to our family. Such is the focus of this article. We tend to think of "stress" as an evil to be avoided at all costs. Stress, however, is an unavoidable fact of life. Just as sad, fearful, or worrisome events cause stress, so too do exciting and wonderful things: marriage, new home, new job, major purchase, pregnancy, births, beginning school, graduating. It isn't, then, that we must keep stress away, but rather that we must learn effective ways to reduce and manage it.



Physiological Reactions to Stress

Stressors cause physiological changes in the body. When the brain perceives a stressor, it first signals the release of the chemicals epinephrine and norepinephrine, and then the release of cortisol and other hormones. The body is put on heightened alert and prepares for a change in behavior (i.e., fight or flight). After the stressor is gone, the body returns to its pre-stress state.

Chronic stress, such as is typically experienced by care providers of persons with PWS, sensitizes the stress system, making it more responsive to stress. The result is the system over responds to new stressors, trapping us in a vicious cycle of perceiving any new stressor as SEVERE, to which we're more likely to attempt to manage it utilizing a familiar, but often unhealthy, stress-reducing strategy, which ends up placing more stress on our body, and on it goes. The long-term potential outcome? Depression, anxiety, hypertension, eating disorders, a tendency to colds and other infections, ulcers, cardiovascular disease, gastrointestinal diseases, migraine headaches, asthma episodes, some cancers, and even an acceleration in the aging process.

Emotional Reactions to Stress

Some people are perpetual Worry Warts who tend to feel more anxious the calmer things become. Others always seem to remain unruffled even in the midst of an obviously sinking ship. While everyone has an emotional reaction to stress, the degree of stress felt is individualized and dependent upon various factors including the complexity of our care taking responsibilities, the quality of our support network, the demands of our daily responsibilities, and our physical

health. Whether you're a Worry Wart, Cool as a Cucumber, or somewhere in between, your first reaction to a stressor will probably be consistent and marks a starting place to begin improving your stress management techniques: Do you tighten up, experience back pain, tummy aches, headaches? Get irritable, impatient, or angry? Do you experience a steady stream of negative thoughts? Tend to cry, ignore, or avoid the problem, or just give up? Do you reach for a cigarette, drink or use some other drug? Eat when you're not hungry? None of these stress managing strategies is necessarily inherently "bad" or unhealthy in and of itself - unless it's the only strategy used to manage stress.

How to Manage Stress

There's actually a lot we can do reduce and manage stress. Below is a list of various stress management strategies. Some strategies won't be a good fit, some may work only occasionally, and some will work very successfully. Give yourself a pat on the back for each one you already utilize and consider adding a few more to your stress-managing cache.

- **Controlled Breathing:** One of my favorite in-the-moment stress reducing techniques is slow, measured breathing. Breathe in normally through your nose and exhale slowly out of your mouth to a count of four. Relax your shoulders. Repeat 4-5 times and notice your mind and body begin to relax.
- **Thought Management:** Another effective in-the-moment technique to use during those "I-can't-take-this-anymore-or-I'll-explode" moments are to softly tell yourself to "Stop. Breathe. Relax. It's ok. I will get through this" or "Stop. Breathe. Relax. There's nothing I can do about the traffic. I'm doing the best I can." Keep things in perspective. Maintain realistic expectations. Don't "sweat the small stuff." Remind yourself that a calm journey is far more important than a punctual but stress-filled destination. Guided Imagery can be used anywhere you have even two minutes: control your breathing, relax your body, and imagine scenes that are relaxing and calming. Other long-term tools include meditation, yoga, and Tai chi.
- **Time Management:** My son's Karate teacher tells his students that the best defense to an attack is to move out of the way. The same principle can apply to stress management: the best defense to stress is to avoid it! Prioritize. Avoid over scheduling whenever possible. Don't over commit or overextend yourself - or your family members. Give yourself plenty of time to transition or travel from place to place. Building in more transition time will place less stress on everyone.
- **Personal Boundary Management:** Having a child with PWS challenges us to think ahead and anticipate potential problems. Acknowledging our own personal boundaries and then assertively (not aggressively) protecting them is healthy, even and especially when interacting with our kids. It's ok to share that today you're feeling extra tired and therefore need extra quiet time. It's ok to tell your friend that you really can't help out today because you've made other commitments (to yourself!). When at all possible, avoid or limit the amount of time you spend with people who stress you out. As Clint Eastwood advises, know your limitations. If you're not good at calmly helping your child with her homework, have someone else do it.
- **Exercise & Diet:** One of the best long-term strategies against stress is to give yourself regular physical activity. Exercise releases wonderful chemicals in your brain that restore and heal the mind and body. Fifteen to twenty minutes a day for 3-5 days a week will make a huge difference in how you feel and how you manage your stressors. I know,

twenty minutes a day of anything focused on just you are a luxury, but we need to create pleasant daily physical activities for our child/adult child anyway! Incorporate a daily walk or wake up 20 minutes earlier in the morning or go to bed 20 minutes later so that you can work out to an exercise DVD. When you get creative, you'll find plenty of opportunities for physical activity. And of course, maintaining a healthy diet gives your brain and body the fuel they need to manage stress better.

- **Sleep:** Adequate, consistent sleep is vital to a healthy body and makes it much easier to manage stressors. For many people, especially those whose child awakens several times during the night, it is very difficult to get the recommended seven to nine hours of sleep necessary to recharge the brain revitalize the body. But without adequate sleep, we are less able to calmly manage even the tiniest stressor.
- **Support:** If you're lucky enough to have a good support system, use it! People with a healthy support network consisting of have fewer stress-related symptoms and manage stress better.
- **Therapeutic Intervention:** It takes intelligence to recognize when we've done all we can and it's just not enough, and it takes courage to reach outside ourselves to ask for help. If you don't have the luxury of a good support system and you're feeling overwhelmed with stress, find a good professional. Many psychologists, psychiatrists, social workers, marriage, and family therapists, even some clergy are trained in stress management techniques. Counseling can be brief or longer-term depending upon the need. When in the midst of a stressor we tend to forget any potential new management options, so until they're automatically incorporated into your daily life it may be helpful to post some of these stress-managing options on your frig or in your car as an in-the-moment reminder. I wish you successful stress-managing strategies that carry you through the holidays... and throughout your lifetime!



Saving and Transforming Lives!

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