

The Child with Prader-Willi syndrome: *Birth to Three*



*Pictured: Michelle Spring
and Audrey (living with PWS)*

By

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With a Parent's Perspective by Cinda Ball

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With a Parent's Perspective

by Cinda Ball ©1999 and 2004

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The children featured in this publication have Prader-Willi Syndrome.

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The Child with Prader-Willi Syndrome: Birth to Three

Revised 2011

Introduction

The first three years of life of children are filled with exciting and dramatic social, emotional, and physical development. While the pace of development of these skills varies from child to child, the dramatic social, emotional, and physical growth that occurs during these years causes them to be known as the “Wonder Years.” Infants and young children with Prader-Willi syndrome (PWS) likewise demonstrate significant developmental achievements in these early years that bring tremendous joy to their families. As these are such critical years of development, we would like to elaborate on some of these achievements. This booklet is intended to help parents as well as extended family members, Early Intervention workers, therapists, physicians, and other care providers of children from birth to three with Prader-Willi syndrome. We will provide an overview of these years with recommendations for optimum care and resources for more in-depth focus on specific areas.

Our goal is to provide parents and care providers with important information so that you have a better understanding of how Prader-Willi syndrome may impact your child, what interventions are available to help you maximize his or her overall development and natural talents, and how you may better prepare for your child’s and your family’s future, all while feeling more confident, optimistic, connected, and supported.



A Parent's Perspective

by Cinda Ball 1999

My husband and I were told that our daughter had been diagnosed with Prader-Willi syndrome when she was 15 days old, the same day we brought her home from the hospital. For the first few weeks, we spent time searching for information about PWS on the Internet and telephone. Our fundamental questions were, "What does the syndrome mean?" and "What can we do for our daughter?" As we searched, we found reams of articles on the syndrome, but surprisingly little regarding what to do. So, in the following paragraphs, I will describe what we are doing to help our daughter.

As my husband and I sought to understand PWS and figure out how to best care for our daughter, we were faced with several major decisions. We needed to choose a pediatrician, figure out how to get her to gain weight, and combat the low levels of activity. Next, we searched for specialists. After seeing three and discussing their advice, we developed a basic approach to Avery's care. Our game plan includes therapy, stimulation, human growth hormone treatment, visits to specialists, and a lot of normal childhood activity.

Choosing her Physicians

Before our daughter was born, we had interviewed and chose the pediatrician in our neighborhood. After Avery's birth we talked to the doctors at the hospital, received recommendations for, and interviewed a couple of pediatricians with backgrounds in developmental delays and neurology. In the end, we chose our neighborhood doctor because of his openness, great common sense, and availability. He also assisted us by frequently weighing Avery, providing emotional support, tracking down information, and assessing the opinions of other specialists.

Feeding

Initially, our primary concerns were feeding the baby and trying to get her to gain weight. In the hospital she had been tube fed and then slowly taught how to suck from a nipple. We decided to keep her on the same feeding schedule that the hospital used every three to four hours.

For the first three months tactics included supporting and moving her jaw, moving the bottle in her mouth to stimulate a suck, tapping the bottom of the bottle, moving her to keep her awake, unwrapping her blanket or uncovering her feet, and playing music. We were aware of the need to balance ensuring that she received adequate amounts of milk without wearing herself out and burning up more calories than she consumed.

In the beginning, we also had a logbook on the counter in the kitchen, where we wrote down a target amount for her to drink for the day and how much she actually drank at each feeding. We also weighed her every two days. If she did not gain weight for two weigh-ins, we knew that she now needed more milk each day to continue to gain weight, and we would increase her target amount of milk. This way we had a good idea of how much she needed to drink to gain weight.

Activity

To address the low levels of arousal and activity, we held Avery as much as we could while she was in the hospital. We also massaged her and talked to her during feedings, diaper changes, and whenever she was awake. For the first four or five months I did range of motion exercises (moving baby's arms, legs, joints, etc.) a couple of times a day. Counting and singing while I moved Avery was fun and gave us an activity to do together in the newborn phase when there are not too many choices for play. We used black and white shapes, a mobile, and a Gemini playmate for visual stimulation and early swatting and reaching. Although our daughter did not develop at the rate of an average child, we found great excitement in each new movement and task she accomplished.

Visits to Specialists

Soon after Avery came home from the hospital, we started searching for specialists. In hindsight we were intrigued with specialists who were practical and proactive. Although the specialists discussed similar information about PWS, their approaches to early action were dramatically different. One recommended watching the growth chart and starting growth hormone when weight and height lines crossed. The second doctor told us to love and care for our child and not do anything special other than some therapy until age two to four when feeding and behavior issues become apparent.

After these two appointments we felt as if we knew more about PWS, but still had not been given any practical advice about how to help our daughter now. The “Birth to Three” booklet was the first literature we read that described actions we could take to possibly improve the outcome for our child and the impact we could have with early intervention. When we spoke to Dr. Robert Wharton and the team he works with in Boston, we found the most proactive group that focused on what we can do now to improve future outcomes. Most regrettably, Robert Wharton, M.D. has since passed away.

We Develop Our Approach

After reading about PWS and visiting these specialists, my husband and I developed an approach to PWS and our daughter’s care. The thought behind our approach is one of planning for the worst and hoping for the future. While there are no magic bullets to cure PWS, there are many suggested actions to take that will not hurt our daughter but may help, such as therapy, nutrition planning and extra stimulation. For example, we are expecting her to have trouble with gross motor skills and speech development and are providing physical therapy and growth hormone medication now. These actions will benefit our child. As for hoping for the best, we believed Dr. Wharton and his colleagues when they told us that early diagnosis, early intervention, and treatments such as growth hormone appear to have created a second generation of children with PWS. Believing in his viewpoint has freed us from the psychological burden of knowing about the old outcomes of the syndrome and freed us to do what is most important – focus on our daughter and spend all of our moments with her productively.

What we do not know includes what characteristics of the syndrome our child will manifest, what is the power of early diagnosis and early intervention, what are the potential outcomes for our child, and what advances in the medical field will bring. From what we do know, there are clues about what to plan for, and from what we don't know, there is endless and tremendous hope that what we are doing will improve our child's life.

The Plan of Action

Based on our approach of planning for the worst and hoping for the best, our plan of action for Avery's care that includes therapy, stimulation, human growth hormone treatment, and regular family activities has taken shape. The role of the parent in therapy is fourfold: arranging the therapy, spending time coordinating and managing the social worker and therapists, participating in the therapy, and carrying over the therapy to our everyday care and play.

Early Intervention (EI) contacted us the day after we brought Avery home from the hospital and assigned us to an agency. The agency planned an at-home evaluation when Avery was seven weeks old that included four evaluators, our pediatrician, and us. During the evaluation, they tested reflexes, body movements, and responses to people, objects, and sounds. Based on the evaluation, we met with the agency and Early Intervention to make an individual Family Service Plan, or IFSP. The IFSP included an hour per week of oralmotor and speech therapy, feeding therapy, occupational therapy (OT) and physical therapy (PT). At about nine months we increased OT and PT to twice a week.

To make our daughter's time with the therapists as productive as possible, I spent time talking to our social worker and coordinating with the therapists. The therapists and I occasionally discussed what their goals were for Avery and what she would be trying to accomplish. Our three therapists talked once in a while and made brief notes in a logbook to coordinate the care and share observations about Avery's development.

I'll give an example of a situation where being close to the daily therapy and managing the situation helped Avery. After the first two months, our physical therapists started spending more time talking about and doing "healing work" instead of standard physical

therapy. Still being new at this, I was unsure about what a physical therapist should be doing with a five-month-old with low tone. By contacting our social worker, we were able to determine that we were not receiving the proper physical therapy and our therapist was changed. The therapists are fantastic and a tremendous resource, but in the end, it is the parents who are handling the child most and are ultimately responsible for the physical and emotional development. So, I do watch and ask the therapists about their work and handling and get their advice regarding what we can be doing at home. Then, as time permits, Avery and I have our own play sessions and incorporate the positioning and activities that the therapists use. Later we show the key things to Dad, babysitters, and grandparents. We spend as much time at floor play as possible and minimize downtime like stroller and car seat time. We do have those “sitting days” and I try to offer some kind of stimulation, even if it doesn’t include physical activity.

To work on speech development, we do oral-motor activities to help develop and use all of the proper muscles. Over time, this has included rubbing Avery’s face with a towel and using an Infantent finger toothbrush and Nuk baby toothbrushes to stimulate the gums, tongue, and mouth. Because a speech problem is likely, including speech delay and/or childhood apraxia of speech (often also referred to as speech dyspraxia), one strategy is to develop alternative methods of communication. We have made a photo book with pictures of familiar objects, pets, and people that we look through and talk about. In addition to hearing about signing with babies from the speech therapists, a book entitled, *Baby Signs* by Linda Acredolo and Susan Goodwyn was very informative. Other good sign language resources include *Talking Hands*. A Sign Language Video for Children by Small Fry Productions, and the website www.signwithme.com. Helpful information about childhood apraxia of speech or dyspraxia can be found on the website www.apraxiakids.org.

Sometimes it seems odd to spend so much time and effort trying to stimulate a child while many parents are trying to calm their children with baths and quiet voices. Stimulating takes creativity and persistence with a child who is less apt to respond. Some things that have helped us have been word and gesture games (with lots of hamming it up and overacting), singing, music, toys that make noise, and physical play. As a first-time mother whose nursery rhyme and song collection was limited to “Old McDonald’s Farm” tapes of children’s music and books with words and finger plays have been invaluable, particularly in the very early days when my daughter’s responses were minimal and play options were limited. The Gimini playmate was useful from months two through seven or so, when Avery was looking, learning to move against gravity, swatting, kicking, and rolling. Light toys with texture, fabric swatches of different textures, and rattles were big hits. One of our favorites is our weekly music class that has tapes and books for home use. We also play with instruments like bells, rhythm sticks and a tambourine. The music also seems to prompt more arousal and physical movement for Avery.

Watching a niece and a friend's child who are the same age as my daughter has given me new ideas for introducing more stimulation. If there are activities that these children enjoy doing that Avery cannot initiate on her own because of the gross motor delays, then I try to create the experience for her. For example, Avery can't get into the kitchen cabinets on her own, so I filled a container with plastic items, bottles, nipples, and pacifiers for her to play with. She also cannot get into the plants or flowers on her own yet, but occasionally I take her to them and let her explore and rip them apart.

Growth Hormone

At the time our daughter was born, one of the biggest decisions a parent of a child with PWS faced was whether to start using human growth hormone (HGH) and when to start.

After discussing it with our doctors, we read a lot of the available information, which at the time was not much. At that time we had two options: do not start HGH and possibly regret that decision five to ten years later, or start HGH and accept the possible unknown future side effects along with the benefits. For us, it was worth the unknown risk to start the treatment. We were particularly anxious to see the benefits of an improving muscle-to-fat-ratio and the potential impact of decreasing appetite and increasing the amount of calories burned. Today, there is sufficient evidence to support that growth hormone treatment can improve the health and quality of life of children with PWS. The questions that remain are largely individual ones - how early to begin treatment, and whether there are good reasons to stop treatment or not to use growth hormone in a particular child.

We started giving our daughter daily injections of growth hormone when she was six months old. The improvements in her tone and strength were noticeable within a month. She progressed to sitting independently quickly. While motor planning, like learning how to push to sit and using knees to transition to standing was difficult, her strength continued to improve. At two years old, her problem-solving and motor planning skills are much more fluid and she is walking and climbing vigorously. The HGH may have increased her attention, level of alertness, energy, and stamina. So far, we are extremely happy with the impact of the human growth hormone.

The Way It Is

We meet with a pediatrician specializing in PWS and an endocrinologist quarterly, and occasionally we see a psychologist. We have met with a speech therapist, physical therapist, and a nutritionist. It has been extremely helpful to talk to these specialists to get new ideas, ask questions, and get an indication of how Avery is doing. It has also been a good way to teach our regular therapists more about the specifics of PWS and Avery's situation.

I would have to imply by all this talk of therapy and play sessions that our life is clinical and scheduled. We do a great deal of "normal" childhood stuff like having play dates, visiting cousins and grandparents, swinging and walking, playing peek-a-boo, and going to birthday parties. Our other favorite daily activity is petting the cats.

Having an approach that my husband and I have discussed at length and worked out has made it easier for us to accept the fact that our daughter has and will continue to have Prader-Willi syndrome. It has helped us to be more relaxed about the daily childcare and more accepting of the curve balls. We know that whatever the outcome for our daughter is, we will probably never know if the causes lie purely in the genetics, the early stimulation, the therapies, or the HGH. Yet we feel better knowing that we have done all we can. So, as we plan for the worst, we have high hopes for the best for our daughter.

A 2004 Update

Key Principles to Help Your Child

Diane Spencer, formerly with Prader-Willi Syndrome Association | USA, notes it is natural and normal for parents to worry about the future of their newborn child. As we talk with the parents of children who have been newly diagnosed with PWS, a reoccurring question is "What does the future hold for my child?" None of our children come with guarantees so no parent really knows what the future holds. We suggest that with research and new management techniques, children with Prader-Willi syndrome can achieve beyond our expectations. The following letter, written five years after the previous article, instructs new parents how to spend their "worry energy" on some truly beneficial activities from the very first day that new little baby comes home with you."

Dear Parents of a Young Child with Prader-Willi Syndrome:

This letter is written to honor the memory of Dr. Robert Wharton. Dr. Wharton taught us four key principles for helping a young child with Prader-Willi syndrome:

- *Stimulate your baby*
- *Use Human Growth Hormone medication*
- *Follow the Food Rules*
- *Provide lots of Socialization*

These are the four key pieces of advice that Dr. Robert Wharton gave to my husband and me regarding our daughter. It is our relationship with Dr. Wharton and his advice that has transformed our lives. With Dr. Wharton's help, we went from fearing that our baby would have mild to moderate cognitive deficits, become obese, and have severe behavior problems, to contending with some learning disabilities, watching what our child eats and limiting some of her behaviors. Dr. Wharton was a developmental pediatrician who devoted much of his work to helping children with Prader-Willi syndrome. He had a knack for focusing on what could be done to help the child's quality of life. He cared for the children and families like no other and excelled at simultaneously comforting and motivating the parents.

Through his work with and devotion to PWS, Dr. Wharton developed strong beliefs about the syndrome. He believed that early diagnosis, human growth hormone treatment and early intervention brought a "Second Generation" of children with PWS. He thought that children who were born with PWS were lacking in survival skills: poor suck, weak cry, reduced movement. Dr. Wharton identified some key areas where intervention by doctors and parents would make an enormous impact on the child and his or her capabilities and happiness.

Stimulate

Dr. Wharton told us that he did not believe that mental retardation necessarily was a part of PWS. He believed that it was a result of the insufficient caloric intake due to the poor sucking instinct and a lack of stimulation due to the sleepiness and low arousal. He believed that if the babies received the proper number of calories and were awakened by the parents and caretakers and stimulated, that the level of cognitive deficits could be reduced or eliminated. (Research conducted in 2006 appears to indicate that morbid obesity prior to the age of four years can lead to cognitive impairment.)

Dr. Wharton noted that babies with PWS often do not “give back” the same way other babies do by smiling, giggling, and asking for more. This lack of response in babies can make many parents feel that their parenting is inadequate and that they are not bonding with their baby. He counseled that the low response is part of the syndrome but does not mean that the child is not bonding or loving. He suggested that parents need to be sensitive to the more subtle feedback given by their child with Prader-Willi syndrome and persistently and consistently interact with, stimulate, and play games with their child.

Human Growth Hormone

Dr. Wharton was an early advocate of human growth hormone medication (HGH). He advised us to give it to our daughter and we started her at 6 months of age. In addition to the obvious benefit of increasing the average height of the child, and Dr. Wharton believed even more strongly in the added benefit that HGH improves the ratio of fat to muscle imbalance. By improving the fat to muscle ratio, it helps to control weight, gives the child more strength and endurance, and helps close the developmental gaps between children with PWS and their peers.

Food Rules

Dr. Wharton said, “Your child lacks the ability to feel full. It is your job to give your child another very consistent signal that she is full. “He thought that children should be kept slim, because as soon as fat cells develop, they might be playing a role in signaling to the brain that the child is still hungry. The formula he gave us was 9 to 11 calories per cm of height. We developed Food Rules that we stick by. Here they are:

Food Rules Goal: To establish disciplined eating habits. It is not that one cookie will lead to obesity, but rather the establishment of habits and strict discipline that is critical.

- All snacks and meals should be eaten at the dinner table when at home
- Snacks away from home should be placed in a “snack bag” and be of a fixed quantity
- No extra snacks should be given to the child in order to keep the child quiet

- Provide all food on the plate or bowl at the beginning of the meal
- Do not provide second helpings unless the total amount of food to be served is divided into two servings
- If our child asks for more, calmly tell the child he/she is all finished eating, and then create a distraction or excitement to transition to the next activity
- No tasting off of other plates
- No tasting or snacking when child is present, unless one is sitting with her and eating from a plate
- For birthdays, special events, or cooking exercise at school, establish the rules you will always follow, and always follow them. For example, you may decide that your child may have one cupcake or cookie, preferably a small one, and no seconds.
- Make fewer exceptions and the child will ask for fewer exceptions

Our three other children also follow these food rules (although they do not follow the 9 - 11 calories per cm). The food rules have benefited us all. There is no begging in front of any candy display, no hassles over meals.

Socialization

Dr. Wharton and his wife, Dr. Karen Levine, believed that ultimately many of the children with PWS were not happy because they were naturally social children but did not socialize well. He suggested that children with PWS need extra help to learn to engage socially with their peers. Dr. Wharton and Dr. Levine told us about “floor time”, a way to play with children, engage, and teach them while you’re playing. He suggested a good resource for learning about floor time is the book, “How to Raise Children with Special Needs” by Stanley Greenspan. Getting your child together with other children, having more children if it’s a possibility and prompting your child to engage with other children are all things that can help lay the groundwork for more fulfilling and healthy socialization later in life.

Conclusion

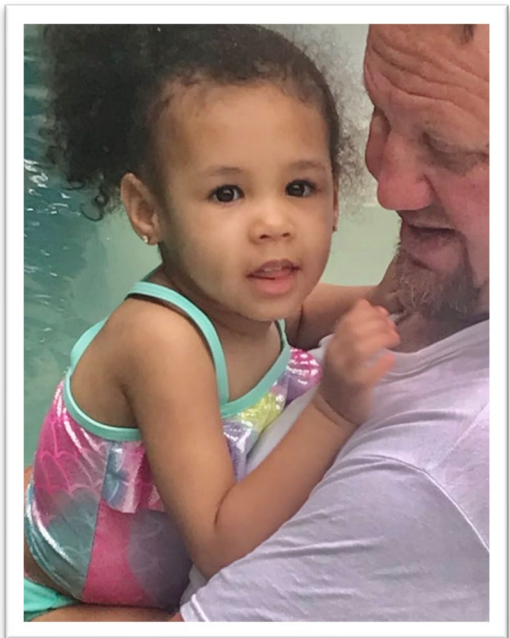
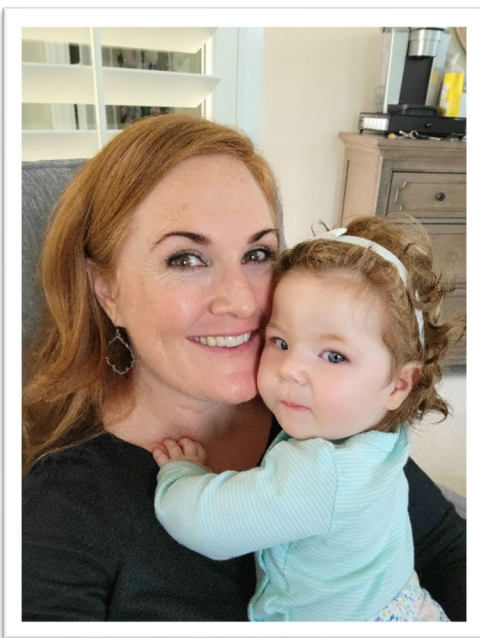
So, the bad news is our children have Prader-Willi syndrome and will have it for every day of their lives. The good news is there are steps you can take that will help your child. For us, over the past six years, helping our daughter and doing everything that we

could do to help her develop and be more capable and happier made us feel better. Somehow, doing for your child cuts through the denial, creates acceptance of the diagnosis, and brings great pleasure to the present and hope for the future.

This letter is really a tribute to Dr. Wharton. We were blessed to have known him for almost six years and we want to spread his words and advice that we believe have been critical to improving our daughter's life. We hope that sharing this information can improve the lives of other children and bring hope to their families!

All our love and luck for a bright future, Another family with a child with PWS.

Please read the PWSA | USA Clinical Advisory Board Consensus Statement on Growth Hormone dated 2009, and the article titled, Consensus Statement/Clinical Practice Guidelines on PWS in the November 2008 issue of the Journal of Clinical Endocrinology and Metabolism, both available from the office of the PWSA | USA.



Chapter 1

Pregnancy and Birth

Pregnancy

Prader-Willi syndrome is a congenital disorder, which means that the signs and symptoms exist because of a problem during early fetal development, namely at the time of conception. There is nothing that either parent did or didn't do at any time in their lives, however, that caused their child to have Prader-Willi syndrome. PWS is simply a part of the human condition and dates back to at least the 16th century.

The earliest recognizable indicators of PWS can usually be detected during pregnancy. Amniocentesis tests do not typically look for Prader-Willi syndrome, but PWS can be diagnosed in utero if it is specifically tested for. Almost all mothers, but especially those for whom the child with Prader-Willi syndrome represents at least a second sustained pregnancy, voice concern during the pregnancy that the child's kicking is not strong, and that the child does not seem to be active. This lack of forceful kicking is usually detected by the mother in the middle trimester and lasts for the remainder of the pregnancy.

In the last trimester additional more subtle differences may also appear. First, while the normal position in utero for a fetus prior to birth is headfirst, in approximately 20 percent of cases of children with Prader-Willi syndrome their position is bottom first or feet first, which is known as "breech." A smaller number of children also may have become stuck in an unusual position whereby their face or their brow is the presenting part at delivery rather than the crown or top of their head. Significantly decreased muscle tone and strength is what causes both the lack of strong kicking as well as the infant's not turning into the usual position for a delivery.

The vast majority of women go into spontaneous labor between 39 and 41 weeks of pregnancy. It is not infrequent that due to the lack of the baby's movement in utero, an obstetrician will want to deliver the baby prior to the due date, often by Cesarean section. When a pregnancy lasts longer than 42 weeks, the child is called postmature. In approximately 20 percent of cases, the pregnancies of woman carrying a child with PWS are in fact prolonged by at least 2 weeks. Therefore, mothers frequently may not go into labor themselves at the expected date of delivery, and instead obstetricians may need to initiate labor either through medication or by performing a Cesarean section.

It is worth reiterating that although the earliest signs of Prader-Willi syndrome can be detected during pregnancy, no activities, medications, drugs, type of work, stress, or other factors related to either parent's behavior have caused their child to have Prader-Willi syndrome.

The problems associated with PWS are caused by a spontaneous and random occurrence involving the child's 15th chromosome pair. Symptoms are due to a lack of expression of genes on a region of the paternally inherited chromosome number 15. The specific region is designated 15q11-q13. Genetic material in this region of chromosome 15 is expressed differently if inherited from the mother or father due to a normal process of genetic modification called "imprinting." In Prader-Willi syndrome, the paternally "imprinted" genes are not expressed due to one of three possible genetic alterations. Deletion of the 15q11-q13 region on the chromosome inherited from the child's father is the most common form, occurring in 70% of all cases. Uniparental Disomy, or UPD, accounts for 25% of cases, and occurs when the baby inherits both copies of chromosome 15 from the mother and is missing the paternal genes on Chromosome 15. The result has the same effect as a Deletion. An Imprinting Defect makes up the remaining 5% of cases. Further testing and genetic counseling are recommended for families who have a child with an Imprinting Defect and who wish to have more children.

The Newborn

As stated above, the newborn with Prader-Willi syndrome continues to have characteristics that have been noted in utero, namely diminished motor skills and arousal. The traditional concept of Prader-Willi syndrome suggests that the hypotonia or diminished motor tone is the most characteristic feature of the neonate. Diminished arousal and alertness should also be considered a "classic" or characteristic sign.

Frequently infants are given poor Apgar scores, a simple method to quickly assess the condition of a newborn immediately after birth, due to their apparent lethargy, poor muscle tone, and general lack of vigorous respiratory efforts. With respect to arousal, the newborn with Prader-Willi syndrome usually demonstrates a lack of alertness unusual for infants having just experienced the trauma of birth. Newborns with PWS characteristically do not cry following delivery nor do they demonstrate auditory alertness or visual attention. Instead, they typically lie on the examining table asleep. They are generally uninterested in and not capable of feeding.

Because of the dramatically decreased tone and arousal, infants with Prader-Willi syndrome are sometimes incorrectly diagnosed as having cerebral palsy or as having undergone a hypoxic injury (lack of oxygen to the brain) as a result of a birth trauma, or are thought to have some form of muscular dystrophy, or benign hypotonia. The application of these misdiagnoses has several implications for children and families.

First, by providing an incorrect diagnosis, a medical evaluation for the correct diagnosis may not be forthcoming, leading to an unfortunate delay receiving important medical and therapeutic interventions. Secondly, parents may look to errors in the birth process as the cause of or contributing factor to the problems they see in their infant.

Lack of muscle tone with diminished strength are additional significant features of infants with Prader-Willi syndrome. If allowed, infants with PWS would tend to sleep through the night from delivery and maintain this pattern of increased sleep for the first six months of life. Infants are generally described as having a “rag doll” feel. This diminished tone, together with decreased arousal, contributes to sucking that is often not initially sufficient for successful feeding. Caregivers must awaken the infant for regular scheduled feedings, night and day. By the end of six months, however, infants generally begin to demonstrate increased periods of alertness and attention. It is likely that their level of arousal, or how awake they appear, while gradually increasing from the newborn period, will still be somewhat diminished. In addition to the features of low muscle tone and poor arousal, there are other features in the newborn that assist with diagnosis. First, although characteristic facial features may be too subtle to detect or may not be present at all, some individuals with PWS may have almond-shaped eyes with upslanting palpebral fissures (the separation between the upper and lower eyelids) that make the eyes look angled slightly upwards. They may also have a narrow forehead and a triangular shaped mouth. Additional features can be a high arched palate, strabismus (see section on strabismus), and a pale complexion. Boys will generally have a small penis and an undeveloped scrotum without the presence of descended testicles. This latter situation is known as cryptorchidism. This feature of hypogonadism will be more difficult to demonstrate in females who may reveal small, underdeveloped labia majora.

Infants with PWS are frequently diagnosed as “failure to thrive” because they often exhibit difficulty sustaining adequate weight gain. This difficulty with growth is associated with insufficient caloric intake caused by feeding difficulties and does not represent a metabolic disorder in the child. Infants generally demonstrate no appetite for their first several months. They tend not to awaken at night for feeding, nor do they cry during the day if they are not fed, or if a bottle is taken away prematurely. Waking to feed, alerting to visual and auditory stimulation, and crying because of discomfort require a level of alertness or arousal that the young infant with PWS does not yet have. These responses are generally described as “primitive responses” and “survival skills” as they alert parents to feed or otherwise respond to the infant. Their absence in the infant with PWS makes the parents’ role even more challenging since they must schedule all feedings, recognize and respond to their baby’s subtle behavior cues, and maximize their baby’s briefer periods of alertness.

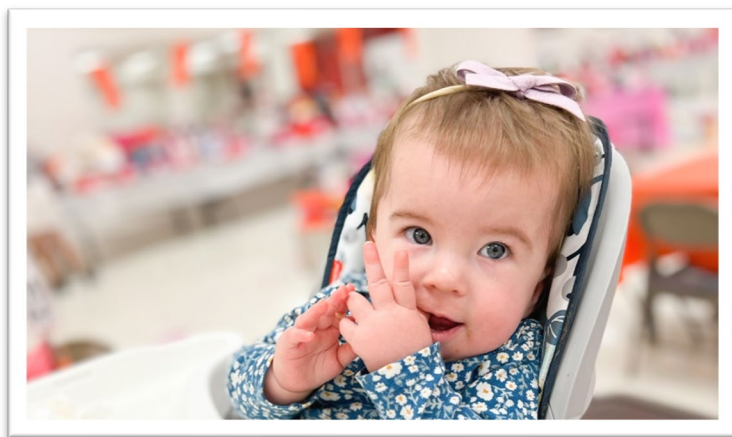
In general, infants should be fed every 2 to 4 hours. Parents have found various strategies that can help keep their baby more awake and alert during feedings: gently scrunching the cheeks with your fingers and thumb to “awaken” the cheek muscles, unwrapping the baby from the blanket or unzipping the onsie to let the air awaken him, tapping the bottom of the feet, tapping the bottom of the bottle, and/or gently move the bottle around in the mouth. Special bottles and nipples, such as the Haberman, can make it easier for the baby to suck. Parents have found other nipples helpful as well, such as NUK silicone Orthodontic slow flow nipples or Gerber Nuk silicone orthodontic or slow flow nipples.

Bottle feedings can take a great deal of effort because muscle strength is generally weaker, and babies often have difficulty coordinating their suck, swallow, and breathing. A great many calories are expended simply during the feeding process. In order to take in more calories than are expended during feeding, a general rule of thumb is to limit the feeding process to no more than 45 minutes. Some pediatricians may recommend a calorie-boosting formula to augment breast milk or formula.

A high or cleft palate can interfere with feeding. The Haberman Feeder is a bottle and nipple system especially designed for impaired sucking ability. Many babies with PWS also appear to experience Gastroesophageal reflux disease (GERD), a condition in which the stomach contents (food or liquid) leak backwards from the stomach into the esophagus (the tube from the mouth to the stomach). A baby with reflux may be reluctant to feed due to discomfort and may benefit from treatment measures to reduce the reflux symptoms, including holding techniques and/or medications (See PWSA | USA’s booklet Nutrition Care for the Infant and Toddler with PWS).

If the baby is still not taking in sufficient calories for adequate growth, it may be necessary to use a nasogastric tube, or NG tube, to feed the infant until he or she has developed a sufficient suck. NG Tube feedings can occur alongside bottle feedings in order to ensure that the infant is receiving the proper caloric intake and is not tiring or burning off more calories than he or she is consuming. Another feeding option that may be used with infants who have continued difficulty bottle feeding is a Gastric tube, or G-tube. A G-tube requires a surgery to insert a tube directly into the infant’s stomach. Breast milk or formula is then poured directly into the tube. The G-tube will remain in place until the infant has developed a sufficient suck. Parents frequently report that they have the greatest success with a Mic-key button feeding tube.

Infants treated with recombinant human growth hormone generally improve their suck at a faster rate and experience greater success bottle-feeding than infants who are not treated with growth hormone. Some babies, though not many, are even able to breast feed. In addition, some families have found that the use of supplements, such as Coenzyme Q-10, L-Carnitine and Creatine help improve levels of alertness, energy and feeding.



Chapter 2

Early Intervention

As early as possible within the first few weeks of life, a referral for therapy services through the State's Early Intervention agency should be made. Early Intervention is a set of therapy services that are federally funded, through the Department of Public Health, for infants 0-3 years of age who have a developmental disability, such as Prader-Willi syndrome. Part H of the Individuals with Disabilities Act, (originally enacted as Public Law 99-457, The Education of the Handicapped Act Amendments), calls for a "statewide, comprehensive, coordinated, multidisciplinary, inter-agency program for all handicapped infants and their families." The Act further states that programs establish "developmental services... to meet a handicapped or toddler's developmental needs in any one or more of the following areas: physical development; cognitive development; speech and language development; psycho-social development; or self help skills." In short, it is your baby's legal right to receive intervention to help with his/her developmental difficulties.

Early Intervention Programs (EIP) are organized by area, much as school systems are, and enrolling in Early Intervention is similar to enrolling in Kindergarten. You can refer your child yourself or ask your pediatrician to refer. You can obtain the telephone number of the EIP in your area by calling your State Department of Public Health office. Following your call, the program will conduct a visit with you and evaluate your child to determine what services you and your child need. You and the EIP staff together then develop what is called the "Individual Family Service Plan" (IFSP), which is the program's contract to provide services for your child.

In some parts of the country local Early Intervention Programs will provide therapy services in the home on at least a weekly basis. In other areas, services are much less frequent and EIP programs will need to be supplemented by therapies funded through other sources, such as private health insurance. It is important for you the parent to be involved in your child's therapies to the greatest extent possible so that you can incorporate the beneficial therapy activities into daily life.

The Early Intervention “core therapies” for almost every child with PWS are Occupational Therapy, Physical Therapy, Speech and Language Therapy, and Social Skills Therapy. Other early intervention therapies can include Feeding Therapy, Oral Motor Therapy, Infant Stimulation Therapy, Music Therapy, Aquatic Therapy, and Behavior Therapy. The types of therapies, their frequency, and duration should be tailored to meet the individual needs of the child.

One to Six Months

The first six months of life for infants with PWS will almost always demonstrate significant positive developments with respect to sleep and arousal, their ability to sustain themselves nutritionally, motor development, and social interaction. Socially, by six months the child will appear increasingly bright, alert and visually attentive. The distant or sleepy appearance of the newborn will have likely improved, although it may not have resolved completely.

The excessive sleep behavior, a feature which leads to infants being described as “good babies”, generally resolves by six months such that the infant has increasingly longer periods of awake and alert time. Somewhat increased nap and/or sleep time, however, may be a persisting feature even into adulthood, although this symptom tends to decrease with growth hormone treatment. It may appear at this time that when awake, infants are fully alert and attentive. The increased attention and arousal brings opportunities for happy reciprocal baby play with parents and siblings.

Muscle tone will continue to gradually improve. The use of growth hormone typically promotes additional improvement in tone. Evidence of this increased tone can be seen in the child’s suck which, in almost all infants with PWS by six months of age, will be sufficient to sustain their nutrition and adequate growth.

The newborn’s apparent lack of interest in food usually resolves during the first six months and a more typical appetite develops. It is imperative that care providers understand the importance of helping the child achieve and maintain a healthy weight and not feed more than the appropriate amount of calories needed for the baby to grow and develop. This is especially important if the child has typically been underweight or appeared frail. Parents may generally rely upon the good advice of your regular pediatrician in regard to amount and types of foods to feed your baby, bearing in mind the importance of providing healthy fats, such as whole milk or soy milk, for good brain development. In general, babies under 1 year should have breast milk or formula.

Whole/soy milk should only be introduced at or after 1 year (some parents have found cow's milk to be problematic, causing too loose stools or diarrhea. Using soy, almond, or some other non-dairy milk product often resolves these intestinal issues).

Tasks

Infants with PWS develop attachments to their parents, siblings, and other family caregivers in the same ways as all babies and thrive on loving attention, cuddling, eye contact, play, smiles, and baby talk. Several small adaptations can ensure that infants with PWS have a full range of play experiences. For the young infant, an Occupational Therapist should provide therapy to improve overall strength and sensory input and integration. As the child approaches six months and older, the Occupational Therapist will focus on improving fine motor skills. If not already working with the child, a Physical Therapist should be introduced to the therapy regimen to begin to focus on the development of large or gross motor skills.

It will be beneficial to purchase lightweight toys (e.g., small, light rattles rather than bigger, heavier ones) that the infant can manipulate easily in spite of motor weakness.

Children with PWS will benefit from early intervention services provided by an occupational therapist who is well trained in sensory-integration therapy. Play therapy which incorporates the use of various textures, movements, body positions, sights, sounds, smells, and tastes will help improve the baby's overall sensory system. Two books on the subject of sensory integration are *The Out-of-Sync Child*, and *The Outof-Sync Child Has Fun*, by Carol Stock Kranowitz, M.D.

Due to low muscle tone, the infant will likely become easily "scrunched down" in some baby seating equipment. Prop the infant up with rolled towels or cushions such that he can sit and look around comfortably as he plays.

With respect to feeding, there are some simple activities a speech therapist can share with you to help improve your infant's mouth and tongue (oral-motor) coordination.

- First, you can begin mealtime with playful exercises called pre-feeding stimulation. Provide deep, firm touch and stimulation to the muscles in the cheeks, lips and tongue to help "wake up" muscles and develop strength and coordination.

- You can also use music with a rhythmic beat to help increase your child's arousal and rhythmic sucking pattern. Most children's music tapes and videos will work.
- Choose a nipple that helps your infant use his/her lips and tongue well but does not make him work too hard for the milk or formula. As previously mentioned, the bottle and nipple system called the Haberman Feeder has been quite successful for a number of families. For more information or to order the feeder, contact the Medela Company at (800) 435-8316 or www.medelabreastfeedingus.com
- Finally, give your child support at his cheeks and jaw so he or she can get a good seal.
- Read the booklet, Nutritional Care for Infants and Toddlers with Prader-Willi Syndrome available from PWSA | USA.

Babies with PWS eventually learn to develop techniques to communicate their wants and needs in essentially the same manner as all babies. Parents may, however, need to pay closer attention to the subtleties of their baby's communications and may need to work a bit harder to capitalize on, and even extend, the baby's awake and alert periods. You will likely need to awaken your baby after providing an adequate amount of time for napping or overnight sleep so that you and your baby can play. Babies learn through play and sensory exploration. The time you spend playing and providing opportunities to experience different kinds of sensory input is critically important to your baby's cognitive, social, emotional, and physical development. Typical games like peek-a-boo help develop your infant's attention and social interaction. When you play, try tickling your baby each time you say "boo" and use lots of intonation in your voice. Your infant will be able to hear, feel, and see the routine of "peek-a-boo." Help your baby shake light-weight rattles, grab crinkle-sounding toys, touch different fabrics, experience different textures, etc.





Chapter 3

Infant to Toddler

Six Months to Two Years

The child with PWS continues to achieve important social and developmental gains during this time period. Socially, the child will progress from just being socially alert to longer periods of social interactive play. Once the infant is alert for long enough to comfortably interact, eye contact and responsive smiles will emerge. The lower muscle tone generally impacts the muscles of the face and often makes early smiles more subtle. The child will continue to demonstrate his strong attachment to parents by showing interest in the parents' activities, need for parental attention, joy in testing parents with games of drop the spoon (or cup or plate!), hide and seek, and occasional displeasure at being left with less familiar adults. Some children may appear to lack some of the features associated with separation anxiety, and can appear quite content to spend prolonged periods in solo play. The ability to sustain themselves in play is occasionally viewed by families with some concern, however this characteristic seems to be simply a temperamental style often associated with PWS and is not associated with deficiencies in emotional development, intelligence or ability to learn new things.

The timing of success in achieving motor and speech milestones varies from child to child, and is usually accelerated with the use of growth hormone treatment. For example, the majority of children with PWS will walk independently between 18 and 30 months of age, although rarely, some children will not walk independently until they are three or four years old. These time frames are usually decreased with the use of growth hormone. Even without growth hormone treatment, however, muscle tone continues to improve over time, though continues to remain diminished irrespective of growth hormone treatment. Children with PWS demonstrate the same will to achieve developmental accomplishments as other children, namely the ability to communicate and move around their environment. However, motor accomplishments are more difficult to attain due to diminished muscle mass and tone, and difficulties with motor

planning skills (global praxis), which is the coordination between the brain and body to plan, sequence, and execute the steps involved in physical movement. All achievements that require improvements in strength, including holding up their heads independently, sitting, crawling, standing and ultimately walking will develop with time and practice, although generally later than seen in typical children.

Strength in the trunk and extremities will develop by placing increasing demands on those muscle groups. For young children this is accomplished gradually through supporting positions that require increasing amounts of strength. Strength alone, however, will not be sufficient to enable children to be successful with their motor milestones. Just as children require memory for such tasks as learning letters and numbers and remembering faces, so too do children require a memory for certain coordinated sequences of movements. Children with PWS frequently require additional practice for the learning associated with planning multiple-step motor activities. Their ability to make their bodies accomplish such tasks as crawling and walking requires sustained repetition for an activity to be truly learned and become automatic.

Expressive language (speech) is frequently the most impaired aspect of development in individuals with PWS. It generally lags behind receptive language (understanding) which develops in tandem with the child's cognitive development. The reason(s) why there are frequently problems with expressive speech are poorly understood but are undoubtedly due in part to oral motor difficulties (i.e., difficulty making the muscles in the mouth and tongue work together to formulate sounds and words). Many children with PWS are diagnosed with a specific speech disorder called Childhood Apraxia of Speech, also sometimes called Developmental Apraxia of Speech, Developmental Verbal Dyspraxia, or Oral-motor Speech Disorder. Speech delay and/ or dyspraxia often cause frustration for both the toddler and the family. In addition, the diminished expressive skills, plus delayed motor skills, make accurate developmental assessment of the child difficult and frequently leads to an underassessment of the child's cognitive level and potential. The actual cognitive level of the child is most apparent from what the child is able to understand rather than what the child is able to say or do with his or her body or hands.

Achievements in play in the first two years generally include the same types of play observed in other children. The usual baby activities such as peek-a-boo, smiling into the mirror, waving bye, and learning how to activate rattles and other noise making toys will all emerge during this period, although in some children they may emerge a few months later. Some baby toys will be too heavy for the child to manipulate but the infant and toddler will easily be able to manipulate lighter toys. The mild delays sometimes apparent in this period can be due to motor delays, longer time spent sleeping, and lowered arousal level and for some children, general developmental delay. Rate of development in this period, however, is not necessarily predictive of later rate. Delays in development of play activities do not necessarily indicate that the child will eventually demonstrate global developmental delays.

Appetite, apparently absent in the first few months of life, begins to appear with the development of oral motor skills sufficient to generate a strong suck. For the remainder of this time period (six months to two years) there are unlikely to be any features of hyperphagia, or the excessive appetite. Instead, children demonstrate the normal range of hunger and typical variety of tastes and interest in food as seen in other children. An increased interest in food or the hyperphagia symptom may appear, however, earlier than age two years. It is important to note that an early expression of the hyperphagia does not necessarily indicate that this symptom will be more severe later in life.

Children's sleep patterns will also change following the first six months of fairly sustained sleepy periods. Children will not only have increased periods of being awake, but their arousal, or level of alertness, will also increase. Growth hormone and supplements may also improve their level of arousal. In spite of this change, however, some children may fall from wakefulness into sleep rapidly and dramatically on some occasions, particularly while eating a meal, as well as at times where the child is experiencing a decrease in direct stimulation and attention (e.g., riding in a car). It is currently theorized that for some toddlers, falling asleep while eating may occur because of the increased pleasure the brain experiences while eating.

Some children with PWS develop constant strabismus (when the eye deviation or turning occurs all the time), intermittent or alternating strabismus (when the eye turn might be observed only occasionally), myopia (difficulty seeing far away), or amblyopia ("lazy eye"). Treatment for these eye symptoms can include patching, visual exercises, eye glasses, or surgery. More recently, some parents report having great success treating eye problems with Vision Therapy. Vision Therapy is performed by an optometrist who is specifically trained in Vision Therapy. More information about Vision Therapy can be obtained online or from an optometrist. A consultation with a pediatric ophthalmologist is recommended by age six months, and thereafter as recommended by the ophthalmologist, optometrist and/or pediatrician. It is worth noting that ophthalmologists often discount the merits of Vision Therapy, while parents with children whom have undergone Vision Therapy typically report good success.

A pediatric urologist should be consulted if your son's testicles have not descended into their scrotal position (cryptorchidism) by age one year. Some urologists suggest a five week course of HCG (human chorionic gonadotropin) treatment to pharmaceutically stimulate the testicles to descend. Though success has been reported by some families, the overall success rate of HCG treatments to permanently descend the testes is fairly low. Parents often report, however, that HCG shots have helped to increase the size of their son's penis and testicles, and make subsequent surgical intervention easier. One temporary side effect that should be noted is the premature and temporary growth of pubic hair. Patients who fail HCG treatment should undergo a common surgical treatment called an orchiopexy. Most pediatric urologists recommend an orchiopexy no

later than age eighteen months. Prior to any surgery, the surgeon and anesthesiologist should be informed that precautions must be taken when using anesthesia and other medications with an individual with Prader-Willi syndrome, especially a child. Information about anesthesia and medication use can be obtained from the Prader-Willi Syndrome Association | USA.

There have been occasional reports of epilepsy or seizures in individuals with PWS and a suggestion that infants with PWS may be susceptible to seizures, particularly febrile seizures caused by increased body temperature. While several systematic population surveys have failed to mention epilepsy as a specific feature of PWS, epilepsy is associated with other chromosome 15q11-13 defects. Parents who suspect their child may be experiencing seizure activity should consult a pediatric neurologist.

Tasks

To increase your child's capacity for social interaction, experiment with ways to increase your baby's excitement level. See what makes him look at you and widen his eyes happily. Try increasing your own emotional output, with happy excited facial expressions, interacting in "large type" yet in slow enough motion so your baby can respond to you. Try pretending to sneeze, with a long ("haa--haaa-haa") buildup before the funny "--CHEW" part. Try physical tickling games and rough and tumble games with facial expression and words ("Iiiii'm gonna GET you!") that build up in excitement. The more animated you are, usually the more animated and interactive your baby will be. Pause after playing excitedly and see if your baby will do something to indicate that he wants you to continue (eyes widen or smile, or searching your face, or waving his arms). Once you and your baby have some familiar games, just begin to start to play one and see if he or she will continue it. This is the beginning of initiating, which is often very challenging for young children with PWS.

Early Intervention services during this period work with the infant to help with the development of motor and speech skills and also serve as consultants to parents. For younger infants, Early Intervention may take place in the home. As children become increasingly social, around two years of age, the child can attend an Early Intervention playgroup in which speech and motor skills can be practiced in a fun, playful context with other children.

Speech and Language Intervention

Speech Therapy should begin in the first year of life, well before the child is able to actually speak, to minimize the child's frustration by helping him gradually learn to communicate wants and needs. The speech therapist should initially work on developing oral motor skills. In the second year of life, strategies to enhance verbal language development as well as on alternative augmentative forms of communication should be emphasized. At this age, we often use gestures with "silly" animated faces to introduce words to children. Adults naturally use many gestures and exaggerated sing-song intonation when talking to babies. These natural gestures such as waving "hi" and "bye", pointing, and exaggerated intonation are especially beneficial for the child with PWS. The use of gestures provides the child with PWS with an additional modality to facilitate attention and interest in speech and teaches him some ways to communicate before he has the ability to form words.

As the child grows older, speech therapists should be watchful of the characteristics associated with speech dyspraxia. Because speech dyspraxia is now considered common in children with PWS, it is generally recommended that the speech therapist treat the child "as if" he has speech dyspraxia. If the child has an undiagnosed and untreated speech dyspraxia, speech therapy is frequently very frustrating and unsuccessful. A nice resource for information about this specific speech disorder may be found online at www.apraxia-kids.org.

Songs, rhymes and finger plays are wonderful activities to help your child develop his ability to coordinate oral movements, sequence sounds, and learn new vocabulary words. The rhythm and repetitive nature of songs makes it fun and easy. When you first introduce songs, encourage your child to participate by clapping or imitating the gestures only. Then they can begin to "hum" or sing along. You can pause during the familiar and repetitive parts of the song to encourage your child to participate. Try, for example, the familiar favorite The Wheels on the Bus. Your child can gesture for "round and round." You can sing "the wheels on the bus go round", .. pause... to let your child join in. There is a wonderful compilation of songs on a CD titled, Time to Sing! produced by a speech language pathologist especially for kids who have speech problems. The CD, which features traditional songs that are played more slowly making it easier for kids sing along, may be purchased through the www.apraxia-kids.org website.

The use of some formal manual signs (e.g., American Sign Language) and picture communication systems are extremely beneficial to augment what the young child can communicate. It has been found that pairing teaching of augmentative communication such as gestures, signs and pictures while teaching verbal language actually helps children learn to talk sooner than when no augmentative systems are taught. Important early signs to teach include “more”, “all done”, a pointing gesture, “yes” and “no” gestures, and family names. Photos or sketches of daily and important events (e.g., family members, baby-sitters, trip to the zoo) are helpful, too. These photos can be placed in a book so you and your child can turn the pages, label pictures, and talk together about familiar people and places, or create a schedule with them. Eventually the child will begin to use the pictures as one means to request as well as comment about specific people or events. Pictures of favorite toys can be used so your child can point to the picture to make his choice while learning to say the name. Other signs or pictures to be taught will vary depending on what your child wishes to communicate.

Occupational Therapy Intervention to Improve Fine Motor Skills

Occupational therapy, to help improve fine motor skills such as grasping, pointing, pinching, etc. should continue. Therapy should focus upon improving the child’s sensory integration system, which involves the brain’s processing and organization of all sensory input including touch, taste, movement, body awareness, the pull of gravity, sight, and sound.

Physical Therapy Intervention to Improve Gross Motor Skills

Physical therapy in this period should emphasize activities to strengthen specific large muscle groups important for improving posture and gross motor skills. Placing your child on his or her tummy will help strengthen the arms, neck and shoulder muscles. Reaching for toys while lying on the stomach and putting weight on the forearms will also help strengthen these muscle groups as well as facilitate shoulder stability. Assisted reaching activities in sitting, assisted pull to sit during diaper changes and gentle resistance during rolling are activities that can be incorporated into play activities at home with songs and toys. As children with PWS often have difficulty organizing their bodies to perform fine and gross motor activities, therapy should be designed to give the child the idea of how to move his/her body to change positions.

Some children may need special equipment in order to achieve success at gross motor skills. Infant car seats may need to be adapted with rolls for better head control and adaptive seating may also be required to facilitate developmentally appropriate fine motor and other developmental activities. For more information about Physical Therapy, please read *Physical Therapy for the Child with Prader-Willi Syndrome* by Janice Agarwal, PT, available from PWSA | USA.

Play and Socialization Skills

At some point between one and two years the child will begin to enjoy pretend play such as with a doll or toy car or stuffed animals. This type of play is important to encourage as it will help language and communication develop more quickly.

Books are also an important activity for developing attention, listening skills, communication, and speech. Just as with the singing, establishing familiar favorites with predictable routines will facilitate your child's participation. In the classic, familiar favorite *Good Night Moon*, your child can join in by waving bye-bye or gesturing for "good night." Encourage your child to say the names of the pictures as you point and say "good night" or just say "night night" if that's easier. You and your child will quickly establish a library of favorites that will be a fun part of a daily routine and an important part of development.

Feeding and Mealtime Routines and Schedules

Regarding feeding during the six-to-twelve month period, the infant's continuing challenge of low muscle tone and oral motor coordination may make it difficult to transition from pureed baby food to chunky foods. Encourage your child to explore appropriate oral toys such as teething toys and his hands in his mouth. This is an important developmental experience for your child's oral motor and sensory skills. This activity helps to develop the movement patterns for chewing. In addition, make the transition from pureed to textured foods slowly. Pair new items and textures with familiar favorites. You can texture little by little. For example, gradually increase the amount of infant rice cereal so it has moist lumps. The benefits of infant rice cereal are that it is bland, has low risk in terms of causing an allergic reaction, is iron fortified, and is easy

to adjust the consistency, although some parents have found that it can cause constipation. Rice cereal should not be fed through a bottle. Children should receive food sufficient to meet their nutritional needs. As some children may be recovering from a period of poor weight gain experienced in the preceding period, parents may have a tendency to try to achieve “catch up” nutrition to ensure adequate weight gain. No child should be encouraged to eat past their own natural appetite, should one exist, however, or consume more calories than is healthy. The amount of food offered should be established in consultation with the child’s pediatrician, and the types of foods offered should be nutritious and contain healthy fats necessary for healthy growth and development. Answers to frequently asked questions about nutrition and feeding infants and toddlers with Prader-Willi syndrome can be found in the PWSA (USA) publication, Nutrition Care for Children with PWS, Infants and Toddlers by Janice Hovasi Cox, M.S., RD. and Denise Doorlag, OTR.

Children with PWS will benefit from the foundations of good parenting approaches appropriate for children in general with some additional strategies that are specific to Prader-Willi syndrome. Establishing structure and routine in daily life is important for all children and particularly important for the child with Prader-Willi syndrome. Structure and routines eliminate the unknown, reduce anxiety, and help children feel safe and secure. Continue to build upon the routines established in infancy. Children with PWS thrive in an environment that is rich with structure, routine, and predictability, so continue the feeding schedules with mealtime and snack schedules, nap, playtime, and bedtime schedules.

Two to Three Years

During this period, exciting gains continue in the areas of socialization, communication, and ambulation. Many children with PWS will begin walking and talking in this period. Play becomes complex, with toddlers beginning to play pretend and to imitate their parents and siblings. Children in this age range will begin to enjoy looking at books and television or videos (welcome to the age of Barney!).

Although typically less so when treated with growth hormone, toddlers will generally be late to attain developmental milestones such as sitting up, crawling and walking. Therapies begun in infancy should continue throughout this phase of life. Occupational therapy should continue to address fine motor control and sensory integration issues. Physical therapy should continue to focus upon increasing muscle strength and improve coordination and motor planning skills. During these first few years of life, the infant’s

mind will likely be “ahead” of his body and he may be ready to learn more about his environment than his body will “allow.” Parents can assist by helping the child explore, maneuver, and have fun with his environment. For example, using your hand over his hand, help your child pick up the blocks, stack them, and knock them down. Speech and language therapy should continue to help improve articulation, word acquisition and language skills. Social Skills Therapy should begin during this phase of life to help the child participate in more cooperative play.

During this period mild behavioral differences may begin to emerge. While children with PWS at this age seem to generally have a reasonable eating pattern, there are often subtle appetite differences. Some children may show increased interest in food and/or may have mildly elevated appetites. Some children with PWS instead of exhibiting the typical fussy toddler eating behavior instead may be described as “serious eaters.” Although most toddlers will not search for food when none is in view and will not tantrum if they do not receive what they believe to be sufficient amounts of food, most will finish most or all food on their plates and eat most if not all foods offered to them. The appetite of toddlers with PWS may vary from one day to the next and they may have clear likes and dislikes. The more indiscriminate eating pattern of toddlers with PWS probably reflects the beginning of the hyperphagia – insatiable appetite – stage. The excessive appetite typically begins sometime around the age of 2 years. However, it is not infrequent that parents will report their toddler’s appetite became extremely high overnight and they were sure the “PWS Appetite” had begun, only to find that in the next day or two their child’s appetite was back to its status quo. These temporary spikes in appetite may simply reflect a growth spurt. Care should be taken to ensure that calories provided to the child are adequate, not too few and not in excess of what is needed for healthy growth. It is recommended that the parents begin to increase their focus to teach their child healthy food choices and amounts and develop healthy eating habits.

It is during this period that families should begin implementing the Principles of Food Security developed by PWS specialists Janice Forster, M.D. and Linda Gourash, M.D. Families and care providers of someone with Prader-Willi syndrome can significantly improve everyone’s daily life by adhering to PWS Specialists Linda Gourash, M.D. and Janice Forster’s, M.D. Principles of Food Security: No Doubt; No Hope; No Disappointment. No Doubt about when meals and snacks will be served, plus No Hope that extra food can be obtained equals No Disappointment that extra food was not obtained, and it is disappointment that generally fuels a behavior problem.

The tantrums and behaviors typically associated with the “terrible twos” are also seen in children with PWS during ages two to three years. Other children with PWS do not experience behavioral difficulties beyond the norm during this period. For children with PWS who do exhibit increased behavioral difficulties, these may be caused by frustration related to their difficulty to verbally communicate. As the child’s verbal skills improve, especially with the aid of speech therapy, this type of behavioral difficulty lessens.

One of the parts of the brain that is impacted by Prader-Willi syndrome regulates the control of emotions. Most children with PWS have a very low tolerance for frustration and can quickly experience a loss of emotional control. In addition, most children with PWS experience higher-than-normal levels of anxiety which leaves them feeling vulnerable to changes in plans and unable to calmly respond to unexpected situations. Parenting techniques that incorporate and emphasize positive reinforcers, consistent routines and structure are beneficial for all children, and absolutely critical for the child with PWS. Children with PWS respond very poorly to criticism and punishment, but respond extraordinarily well to positive praise. Structure and routines create an overall sense of predictability and safety. It is important to establish consistent routines and schedules, including consistent bedtime, naptimes, and wake up times; consistent snack and meal times; consistent therapy and play times; etc. Consistency in terms of parental behavior is equally important. The adage, "Say what you mean and mean what you say" provides a helpful guideline. For example, if you tell your child, "You will need to stop in five minutes and clean up your toys" then you must help the child stop and clean up in five minutes. Or, if you tell your child he or she may only eat one more item, then don't allow your child to eat beyond that one item, especially if the child proceeds to tantrum. Give positive attention for successful behaviors, and ignore or gently limit negative or detrimental behaviors. Brief (not more than a minute) "time out" periods in a safe area such as a playpen for difficult tantrums will be helpful for the child as well as for the parent. Further, increasing ways for the child to express him/herself will diminish behavioral challenges. Introducing simple signs such as "more", "all done", and "no" will help children who do not yet have words for these concepts.

Providing calm and consistent structure and limits now will help reduce tantrum behavior in the future. Remember, though, that all young children tantrum on occasion - it is a child's "job" to push limits, and it is a parent's job to teach the child what those limits are.

Many children with PWS exhibit excessive-repetitive behaviors which can cause concern for their parents. Common behaviors include pulling at or playing with strings, repeatedly putting things in containers and then taking them out, collecting and needing to carry a large number of toys such as dolls or stuffed animals, and lining up items or toys. Unless these behaviors begin to interfere with the daily life of the child with PWS, they probably don't need to receive intervention, other than to limit the amount of time the child spends doing them. Gently gain your child's attention, and encourage a transition to some other activity.

Socially, children with PWS, like other children at this age, are particularly charming and delightful to be around. All children up to the age of about three years engage in parallel play, meaning they "play" by themselves while sitting next to a playmate who is also playing by himself. At this age, young children with PWS will interact and play

appropriately with other children and adults, and play with typically developing children will generally be quite successful. After the age of about three years, typically developing children begin to shift into cooperative play, which incorporates more complex and sustained interactions with other children. For various reasons, including problems with speech problems and/or auditory processing delays, some children with PWS may need assistance to transition from parallel to cooperative play. Play dates and opportunities for more one-on-one social opportunities, including therapeutic play groups, may be beneficial to the social development of the three year old. For children with significant motor delays, play in smaller groups will be beneficial to avoid getting “trampled” by large groups of more mobile children.

Social interaction and symbolic play can be expanded through highly animated fun play sessions with you. Some toddlers with PWS are apt to play alone unless you get them to play with you. Pick toys or activities your child already likes (e.g., bubble play, Sesame Street or Barney figures, etc.) and make the play very interactive. Add pretend elements from the child’s everyday life (e.g., going in the car, being at the park, etc.) to act out in play. Get the child as involved as he can be as you play. The goal here is first and foremost to have fun, and along the way, for your child to learn about turn taking, pretending, and about emotions and sharing of agendas, all of which will be helpful later in conversational skills, behavioral regulation, and friendships. The book, *The Child with Special Needs*, (Addison Wesley, 1998) by Stanley Greenspan, M.D. has many helpful tips about this type of play he calls “floor time.”

Some children, especially those not treated with growth hormone, continue to appear more sleepy or lethargic during this period. This may be due to a combination of lower levels of arousal and also low muscle tone which causes the child’s face and posture to appear more like someone who is sleepy. Growth hormone medication, Creatine and/or L-Carnitine Fumarate supplements may help reduce excessive daytime sleepiness. Many children with PWS exhibit underlying sleep disorders which interfere with quality, restful sleep. A sleep study is recommended for all children with PWS which assesses for potential obstructive and/or central sleep apneas, brain wave activity, respirations, sleep stage architecture, and slow wave sleep. Narcolepsy is occasionally diagnosed in persons with PWS and can be managed with medication.

The rate of learning in young children with PWS varies, with some children developing generally like their age mates with the exception of speech and motor areas and others showing delays in all areas. Although verbal expression as well as gross and fine motor skills will still exhibit some delay, careful developmental evaluation can help provide a better understanding of the child’s learning capabilities. The best indicators of a child’s level of cognitive development is the words the child understands, and how the child plays with pretend toys such as dolls, cars, or trucks. In this period a careful evaluation of a child’s learning strengths by a developmental psychologist or an educational

therapist will be helpful to determine at what level a child is thinking. The person evaluating the child with PWS must be skillful in the assessment of young children who have speech and motor delays. This professional can also supply suggestions regarding helpful activities to enhance learning.

Therapeutic Interventions

Infants and children with PWS can benefit from regular play dates and playgroup experiences in addition to special therapies and early intervention. Through these experiences toddlers learn social skills, language skills, develop early friendships and have lots of fun! Differences in motor or language skills amongst groups of toddlers are generally easily accommodated in small playgroups.

Early Intervention services are very important in this period and should include physical therapy, occupational therapy, speech therapy, social skills play groups, and a developmental educator. Additionally, an Early Intervention play group provides a beneficial learning and social environment. In addition to providing direct therapy, therapists should also serve as consultants to the parents, teaching parents how to incorporate the therapeutic interventions into playful activities to do at home between sessions. It is the impact of these many parent-child interactions and play periods that reinforces and supports the progress made in each therapeutic hour of therapy, and facilitates the child's development.

Physical therapy will continue to be important to assist with strength, endurance, coordination, postural stability, and motor planning development. Due to low muscle tone, children with PWS tend to have excessive foot pronation. Corrective dynamic ankle foot orthotics (DAFOs), or SureStep orthotics, or soft inserts, heel cups or UCBs (University of California Berkeley inserts) can assist in earlier and more stable walking. Additionally, weighted down toy grocery carts or baby carriages can be used for stability during walking. Additional information about physical therapy issues and interventions is available from PWSA | USA.

Occupational therapy will help improve hand strength, finger strength and dexterity, motor planning, coordination, and should address sensory integration issues. For children not treated with growth hormone, and sometimes even for those who are, lightweight toys are easier for play and cognitive stimulation. Use toys that teach the "cause and effect" concepts, such as lightweight rubber squeaky toys and Busy Boxes. Provide assistance if the buttons and knobs are too difficult for the child to activate. Toys can be attached by very short ties to the highchair or play table for the child who is not yet mobile.

There are several important goals for speech therapy during this period. First, for children with significant oral motor problems, working with the child to enhance control of the muscles of the mouth and tongue, through a variety of exercises and games, will help with articulation and pronunciation. Secondly, continuing to use a “total communication” approach, teaching simple signs and gestures, using pictures, as well as building verbal vocabulary all help to allow the child to communicate what’s on his or her mind. A helpful on-line resource for therapeutic speech interventions is www.apraxia-kids.org.

Low saliva production is a common symptom of PWS. Low saliva production looks like stringy, thick saliva that produces “crusties” around the corners of the mouth and can interfere with clear sounding speech and articulation and can cause a host of oral hygiene problems. Products that are made to reduce dry mouth symptoms, such as Biotene products made by Smith Kline Beecham, can be quite helpful. Biotene toothpaste can be purchased in the toothpaste aisle of most grocery stores, or in the local pharmacy.

The Principles of Food Security should begin to be implemented during this time. Access to food should be restricted which may include locking the refrigerator and food pantries. Schedules and routines for meals and snacks should be created. More information about specific strategies may be found in the DVD The Principles of Food Security: Practical Management for the Child and Adult with Prader-Willi Syndrome available from the Prader-Willi Syndrome Association | USA.

Regarding other activities for parents to do with their toddlers, all of the regular family and parent-child activities are highly appropriate and beneficial for toddlers with PWS. While having PWS impacts children’s motor and speech development and may make behavioral challenges more difficult, in general, toddlers with PWS are more like than different from toddlers without PWS. They are sociable, playful, mischievous, communicative, impatient and loving little people just as are all toddlers are. Cuddling in bed, going to the playground or the beach, family outings, shopping, playing with siblings, grandparents, cousins and neighbors, reading books together, watching videos and television for appropriate amounts of time, doing arts and crafts together, playing with dolls and trucks, are all highly appropriate and help toddlers develop in every way.



Chapter 4

Brothers and Sisters and Family Life

Parents often wonder how having a child with Prader-Willi syndrome will impact other children in the family. Research studies have shown that siblings of children with developmental disabilities do not necessarily or automatically have greater difficulties than other children. Further, several studies have found that siblings, especially older sisters, of children with disabilities are more likely to go into the “helping professions” such as education or medicine. Siblings learn very early a different and invaluable set of values around what is important - that children are loved and lovable not for their competitive accomplishments but for who they are. Siblings also learn to celebrate even small achievements in others, and to value differences.

Parents should be aware of some of the more common negative experiences of siblings. Siblings can feel envious, jealous, angry or resentful of the amount of time parents spend attending to and assisting their sibling with PWS. Siblings can feel left out, fear the disability is contagious or inherited, feel embarrassed or lonely, feel the parents love the disabled sibling more, and may desire to be “sick” themselves to get more attention. It is important for parents to help the sibling understand that these feelings are normal and understandable. To the extent possible, spend some individual time each week playing or “hanging out” with the sibling, and create a special activity or routine devoted solely to the sibling without PWS.

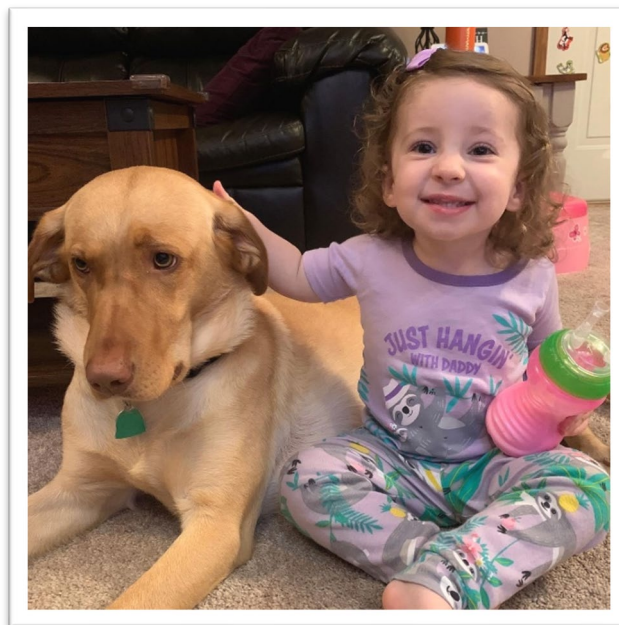
While older siblings can be great helpers and even love to help take care of their new baby sister or brother, care should be taken not to overly depend upon the help of your other child(ren) or place demands upon older siblings that are not age appropriate.

Siblings should be given matter-of-fact explanations about aspects of their brother or sister with Prader-Willi syndrome that may be confusing to them. For instance, they may see neighborhood children of the same age who are walking or talking more. The young sibling (three to five years) can be told something like “He needs help to make his leg muscles/ mouth muscles stronger.” At the same time things that the sibling can do can

be discussed. It is often helpful to use the term “PWS” casually in conversation so it is a word or phrase the sibling and the child with the syndrome is comfortable with even before they have a full understanding of what it is.

Managing meals and snacks to accommodate the differing dietary needs of each individual family member can be a challenge, especially as the child with PWS grows older. Families often report, however, that having a child with PWS has “forced” them into a healthier overall lifestyle that places greater emphasis on exercise, family time, and eating healthier snacks and meals. Many families have found they can share the same type of meal, but make modifications to the meal to be served to the child with PWS. For example, everyone can enjoy a spaghetti dinner but fewer noodles, less sauce and more meat or cottage cheese (for protein) would be served to the child with PWS.

Young siblings may not understand the insatiable appetite that their brother or sister with Prader-Willi syndrome experiences. Siblings may try to “sneak” food to their sibling with PWS, thinking that they are being underfed, and may be angry at parents for denying food for the child with PWS. A matter-of-fact explanation that “Mikey has a special diet” and “Junk food could make him sick” or other such explanation may be helpful. If the child with Prader-Willi syndrome is showing clear signs of a strong appetite, it may also be helpful to explain that “Her brain makes her feel hungry but eating more wouldn’t be healthy for her body” and then maybe to add something like “Like how you feel with your Halloween candy, like you want to eat more candy even though it could make you feel sick.” Of course, these explanations can be helpful to the young child with Prader-Willi syndrome as well as the siblings.





Chapter 5

Parental Adaptation

The initial characteristics of infants with Prader-Willi syndrome have significant consequences for parents. The infant's difficulty achieving an aroused and alert state significantly interferes with their ability to interact socially with their parents. For mothers, traditionally the “nurturers”, this difficulty connecting to their child is often compounded by the mother's inability to nourish her newborn through either the breast or bottle. Mothers and fathers therefore need to develop other strategies to foster solid emotional attachment. Parents should remain alert for signs of the child's own personality and work to understand the child's inherent rhythms. Fortunately, infants with PWS welcome holding, communicating through songs and glances, and respond to the loving intervention of their parents. In spite of the challenges, infants and young children with PWS have many delightful qualities that enable parents to become strongly bonded and feel effective and competent as their sleepy infant develops into a sturdy, playful and communicative toddler.

Learning that a baby has PWS often feels overwhelming to parents. A combination of disbelief, guilt, anger, fear, and grief are common and typical feelings, and will likely recur at various stages throughout the child's development. For most parents, however, feelings of well being will emerge as they bond with their baby and create typical, day-to-day schedules and routines.

Some parents will want to learn as much as possible about Prader-Willi syndrome as soon as the diagnosis is confirmed. For these parents, contacting other parents of young children with PWS either through Early Intervention, PWSA | USA and their local PWS organization will be helpful. Other parents may at first decide not to investigate much about PWS, or to talk much about it. Parents should remember that there is no one right way to cope or feel, and that different people use different coping strategies to manage their feelings of grief and stress in order to proceed with the job of parenting their child.

Couples who have different styles of coping may find it helpful to discuss their differences openly with each other. Taking care of the marital relationship is extremely important – as important as taking care of the child. Of particular challenge to parents with a child who has PWS is the fact that spouses may very well be at a different emotional places; while one parent may be closer to the realm of “acceptance” and ready to make an action plan, their spouse may be fully involved in denial or feeling more anger or depression. While one spouse may be ready to connect with a PWS support group or other parents, their spouse may not be ready to deal with the syndrome. It is important to remind ourselves that we must allow our own process to proceed at its own pace and we must tolerate and support our partner’s process as it proceeds at its own pace. Understanding that the grief we feel is chronic may provide us greater motivation to seek out ongoing support systems and help us develop patience and a deeper understanding of ourselves and our spouse who is also experiencing chronic grief. It is imperative, however, that parents do become knowledgeable about Prader-Willi syndrome as quickly as possible so that they can provide their child with the appropriate and helpful therapies, medications, and home and school environments that will enable their child to experience a happy and healthy childhood.

While these are broad generalizations, it can be helpful to keep in mind that generally men are able to talk about their thoughts about a given situation, whereas women will talk about their feelings. Asking one’s husband to share his thoughts on a subject may yield better results than asking him to share his feelings. Men are generally “doers” or “fixers” and are often quick to try to “fix” a situation someone may wish to simply share with him. It can be helpful to preface a conversation with a “fixer” by stating clearly at the beginning of the conversation, “I’m not looking for you to do anything about what I’m about to share with you. It will be helpful to me for you to just listen.”

Fathers who are overwhelmed by feelings of grief often look for solace or relief by working longer hours at their workplace. Mothers who are overwhelmed by feelings of grief may be more emotional (feeling a roller coaster of sadness, anger, anxiety, fear, hopelessness, etc), withdrawn, or overly focused upon the welfare of her child to the exclusion of everything else. Extreme care should be taken to attend to, understand, and comfort your spouse as you comfort and care for yourself and your family as a whole.

One unnecessary burden some parents experience is blaming or being blamed by the other parent, or being blamed by extended family members for “causing” the child to have Prader-Willi syndrome. In almost all cases, there is no known cause for Prader-Willi syndrome. Remember, nothing either parent did or didn’t do at any time in their lives, including during pregnancy, caused their child to have PWS. There are some rare instances where a specific type of Prader-Willi syndrome may be passed from parent to child, and therefore it is recommended that parents seek out the

guidance of a genetic counselor if they intend to have more children and are concerned about the possibility of having another child with PWS.

More helpful information about how to improve the marital relationship and family dynamics can be found in the article, *Laughter Through Tears: Creating a Strong, Supportive and Healthy Marriage and Family* available from PWSA | USA.

It may be reassuring to know that over time, parents can absolutely once again experience happiness and well being both within themselves and with their child and family. Parents may find it helpful to talk with their physician, their child's doctor, or a mental health professional for some extra support while coping with the initial adjustment. Most parents report that while the unexpected and initially devastating shift in their lives has brought them sadness and challenges they did not anticipate, having a child with PWS has also eventually brought them to experiences, people, and feelings that they deeply appreciate and would not otherwise have known.

Following is a partial list of some valuable resources and support services available to parents, extended family members and care providers. Please contact the national Prader-Willi Syndrome Association | USA at 941-312-0400 or visit www.pwsausa.org to learn more about any of these resources, or to receive the most current list of resources, books, DVDs and other materials.





Support Services

Parent Mentors and Grandparent Mentors are available through the Prader-Willi Syndrome Association | USA to provide in-person and/or telephone support to new parents and grandparents.

Online Support Groups are available for parents and care providers through the Prader-Willi Syndrome Association | USA. These “eSupport Groups” are separated by age of the child with PWS, i.e., Birth to Five Years; Six to Twelve Years, Teens, Adults. Additional eSupport Groups are also available for Military families, Siblings, dually diagnosed PWS and Autism, Spanish Speaking families, and more. Visit www.pwsausa.org and search for the eSupport Group(s) that interests you.

Additional Articles, Books and DVD Resources

The Early Years - A collection of articles regarding young children with Prader-Willi syndrome. Prader-Willi Syndrome Association | USA.

My Grandchild has Prader-Willi Syndrome: Now What? By Barbara McManus and Michele Leightman. Booklet written by grandparents for grandparents. Prader-Willi Syndrome Association | USA. Updated 2010.

The Genetics of Prader-Willi Syndrome: An Explanation for the Rest of Us. Prader-Willi Syndrome Association | USA. Updated 2004.

Management of Prader-Willi Syndrome, 3rd Edition. Butler, M.G., Lee, P.D.K. & Whitman, B.Y. (editors)., M.G. Butler, P.D.K. Lee & B.Y. Whitman (eds.), Springer Verlag Publishers, New York, NY. pp. 1-550, 2006.

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Physical Therapy for the Child with Prader-Willi Syndrome, by Janice Agarwal, PT. Prader-Willi Syndrome Association | USA. Updated 2010

Nutrition Care for Children with PWS, Infants and Toddlers. Prader-Willi Syndrome Association | USA. Updated 2004

Nutritional phases in Prader-Willi syndrome. Miller, J.L., Lynn, C.H., Driscoll, D.C., Goldstone, A.P., Kimonis, V., Dykens, E., Butler, M.G., Shuster, J.J. & Driscoll, D.J. American Journal of Medical Genetics Part A. 155(5):1040-1049. May 2011

Red Yellow Green: System for Weight Management, by Karen Balko, RD. Available for purchase from Prader-Willi Syndrome Association | USA. Updated 2005.

Cookbook for the PWS Diet, by Donna Unterberger. Prader-Willi Syndrome Association | USA. Cookbook is filled with low-fat, low-sugar recipes designed to be used by the whole family. Updated 2003.

See Me, Hear Me, I'm Here, Too. Prader-Willi Syndrome Association | USA. A book about siblings, for siblings, and by siblings. Updated 2006.

Sometimes I'm Mad, Sometimes I'm Glad – A Sibling Booklet, by Sarah Heinemann as told to her mother, Janalee Heinemann. Prader-Willi Syndrome Association | USA. Updated 2005.

Prader-Willi Syndrome Medical Alerts Booklet, Prader-Willi Syndrome Association | USA. Important pocket-size resource for parents to give to their child's doctor, ER staff, caregiver, etc. Updated 2022.

Growing Up with Prader-Willi Syndrome: Personal Reflections of a Mother, by Janalee Heinemann. A collection of articles and tips for managing family life on a practical level. Prader-Willi Syndrome Association | USA. Updated 2003.

Nobody's Perfect, Nancy B. Miller, Ph. D.

The Seven Principles for Making Marriage Work, John M. Gottman, Ph.D. and Nan Silver

Laughter Through Tears: Creating a Strong, Supportive and Healthy Marriage and Family, by Janalee Heinemann, M.S. and Lisa Graziano, M.A., MFT. Prader-Willi Syndrome Association | USA.

Food, Behavior & Beyond: Practical Management for the Child and Adult with Prader-Willi Syndrome. DVD featuring Linda M. Gourash MD, developmental Pediatrician and Janice L. Forster, MD, child and adolescent Neuropsychiatrist. Prader-Willi Syndrome Association | USA. Available with Spanish subtitles. Updated 2005.



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