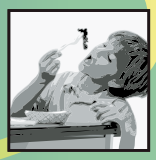
Nutrition Focus

for children with special health care needs





Volume 33, Number 6

Prader-Willi Syndrome – Nutrition Update

INTRODUCTION

- **Max**, a 6-month-old infant, has recently had a gastrostomy tube placed after receiving breast milk via NG tube due to significant hypotonia. His length is <5th percentile, weight is at 5th percentile, and weight-for-length is 50th percentile.

- **Maddie** is 4 years old and participates in Head Start. Her parents report that she asks for food between meals and regular snacks, and will often have tantrums if denied. The Head Start meals are served family style, but Maddie's parents would like to have more control over her intake. Her BMI has gone from the 75th to the 95th percentile in the past 6 months.

- **Drew** is a 12-year-old middle school student who moves from classroom to classroom during the day. Although he has a modified low-calorie school lunch (total of 350 calories), he frequently begs more food from classmates or sneaks food from their sack lunches. Teachers feel it is not fair to deny Drew classroom food treats. He prefers sedentary activities, so doesn't mind being kept in at recess for getting into trouble. His BMI in the past two years has fluctuated from the 90th percentile to >95th percentile.

- **Cody** is 17 years old and in high school special education. His single mother has worked hard to keep his weight and diet in good control over the years, with ups and downs. Although her other two children haven't always helped, now that they are older they understand and are supportive. In the past few years Cody has developed symptoms of obsessive-compulsive disorder (OCD) and receives counseling and medication. His mother is worried about his future – what he will do after high school graduation, how will his diet be managed, and who will take care of him when she can't? His BMI for the past 3 years has been 75th to >95th percentile.

EDITOR'S NOTE

This issue is an update of an article written by Betty Lucas, MPH, RD, CD and Sharon Feucht, MA, RD, CD in 2009.

Thank you to Anne Leavitt, MD for contributions to this issue.

All of these children have Prader-Willi syndrome (PWS). Their nutrient needs will depend on their age, stage of development, medical complications, and motor/activity level. Each individual will manifest the characteristics of the syndrome differently, so the "typical" does not always apply to the individual. Each family is unique in their strengths, management style, and the education and support needed. The role of the Registered Dietitian Nutritionist (RDN) is to work with the family to ensure optimal nutrition and weight management, provide anticipatory guidance, and communicate with the health care team and educators.

At the end of this issue, we'll revisit each of these children to see how nutrition interventions may play a role.

INCIDENCE, DEFINITION, AND DIAGNOSIS

When Prader-Willi syndrome (PWS) was first described in the 1950s, diagnosis was based on specific clinical diagnostic criteria. Molecular technology now provides genetic testing, and the age of diagnosis has fallen in the past decade, so that many are diagnosed in early infancy. This has made a significant difference in providing early intervention, specialty medical services, and anticipatory guidance for families. It is estimated that the incidence of PWS is between 1 in 10,000 and 1 in 30,000 live births in all racial groups.[1-3]

The definition of PWS is the lack of expression of genes on the paternally derived chromosome 15q11-q13, which can be confirmed by a DNA methylation test. These genes may not be expressed because they are missing, defective, or silenced. Another test, fluorescence in situ hybridization (FISH), is used to distinguish uniparental disomy (UPD)

or imprinting error subtypes. Since imprinted genes show DNA methylation based on parental origin, children with PWS will have a maternal-only imprint because they are lacking the paternal contribution. About 70% of individuals with PWS have a paternal deletion of chromosome 15q11-q13. About 20% of the time both of the individual's chromosome 15 come from the egg – maternal UPD. Imprinting center defects occur in <5% of cases.[4]

CHARACTERISTICS OF PWS

Clinical and developmental characteristics of Prader-Willi syndrome include:

- Hypotonia, poor suck, and failure to thrive in infancy
- Global developmental delay; cognitive impairment
- Specific physical features, i.e., almond-shaped eyes, small hands and feet
- Short stature, slow growth
- Hyperphagia, obsession with food after infancy
- Hypogonadism, delayed (or no) puberty

Persons with PWS are also at increased risk for scoliosis, sleep disorders, hypothyroidism, eye problems, skin picking, temper outbursts, repetitive and ritualistic behaviors, affective disorders and psychiatric illness.[1]

There appear to be differences in some of the characteristics depending on the genetic presentation, e.g., those with UPD have higher verbal intelligence scores but an increased risk of psychosis.

Each individual with PWS is unique in his or her expression of the syndrome and related risk factors. There is need for a coordinated interdisciplinary approach for each person, in order to provide early intervention, ensure medical and nutrition management, prevent complications and increase the quality of life.[2]

MANAGEMENT OF PWS

The American Academy of Pediatrics (AAP) published clinical guidelines management of PWS. This report outlines clinical diagnostic criteria that indicate the need for genetic testing. (See Table 1 on page 3 for a list of clinical findings.) Recommendations for management of PWS include nutrition management, growth hormone therapy and other endocrine issues, behavioral food controls, behavior management, and considerations for transition to adult care. Age-specific evaluation concerns and anticipatory guidance are also described.[4]

Because of the complexity of PWS management, Duis, et al propose a multidisciplinary "centers of excellence" approach that includes involvement of professionals from the following disciplines: endocrine, genetics, ophthalmology, pulmonology, nutrition, speech/occupational/physical therapy, behavior, psychology, neurology, and dentistry.[2] One example of an interdisciplinary approach is that of Seattle Children's Hospital's PWS Clinic. At each visit, families meet with providers from nutrition, endocrinology, sleep medicine, and developmental behavioral pediatrics. Appointments with providers from general psychiatry can be scheduled as needed.

Care coordination is necessary, and telemedicine and/or community and home-based health services may improve access to care.

Growth and Growth Hormone

Birth length is short in infants with PWS, and this continues as they grow. Lack of a pubertal growth spurt also results in adults with shorter than average height. Their body composition is also different, with increased body fat and reduced muscle, and a pattern of central fat deposition. [3-7]

Growth hormone (GH) treatment is recommended for individuals with PWS. GH therapy has positive effects on linear growth, lean body mass, and motor development,[8] and, when combined with diet management, can help prevent obesity. It is also thought that GH therapy can improve bone density and stabilize behavioral decline and may have positive effects on cognitive, communication, and adaptive skills.[1,4] For many patients, the GH therapy may start soon after diagnosis, once cleared by sleep provider. Unlike other diagnoses, growth hormone insufficiency is not required before starting the treatment. A discussion of GH therapy and macronutrient composition of the diet was presented in a previous issue of Nutrition Focus. (See Nutrition Focus 29(4).)

NUTRITION-RELATED ISSUES

Initial problems with slow weight gain and growth are common, and virtually all infants with PWS require tube feeding because they have a weak suck and/or low muscle tone or are easily fatigued. Some also do not provide clear hunger and satiety cues, so parents need support establishing a feeding schedule. Centers may routinely recommend video fluoroscopic swallowing studies (VFSS) before oral feedings are started because of increased risk for aspiration.

Between 1 and 6 years of age children enter a stage of hyperphagia, with an increased rate of weight gain and increased interest in food. In the later preschool or school years, food-seeking behaviors begin. Children eat large amounts if allowed free access to food and have apparent lack of satiety. However, each child will vary in the degree he/she demonstrates these stages. Miller, et al described seven distinct nutritional phases associated with PWS. (See Table 2 on page 4.)

Table 1. Selected Clinical Findings in Prader-Willi syndrome

Fetal
- Breech position
- Reduced fetal activity
- Polyhydramnios

Growth
- Short stature
- Failure to thrive in infancy
- Central obesity

Head and neck
- Long, narrow head
- Almond-shaped eyes
- Strabismus
- Up-slanting palpebral fissures
- Myopia
- Hyperopia
- Thin upper lip
- Small-appearing mouth
- Down-turned corners of mouth
- Thick, viscous (reduced) saliva
- Enamel hypoplasia
- Early dental caries
- Dental crowding and malocclusion

Respiratory
- Hypoventilation
- Obstructive sleep apnea
- Central sleep apnea

Gastrointestinal
- Feeding problems in infancy
- Gastroesophageal reflux
- Decreased vomiting

Genitourinary
- Hypogonadism

Skeletal
- Osteoporosis
- Osteopenia
- Scoliosis
- Kyphosis
- Small hands and feet
- Narrow hands with straight ulnar border

Skin, nails, hair
- Hypopigmentation
- Blonde to light-brown hair
- Frontal hair upsweep

Neurologic
- Severe neonatal hypotonia that improves with age
- Poor neonatal suck and swallow reflexes
- Poor gross motor coordination
- Poor fine motor coordination
- Mild-to-moderate intellectual disability
- Learning disabilities
- Increased risk of seizures
- Global developmental delay
- Speech-articulation problems
- Hyperphagia

Sleep
- Snoring/obstructive sleep apnea
- Central apnea during sleep
- Excessive daytime sleepiness
- Early-morning waking
- Night-awakening for food-seeking

Endocrine
- Hyperinsulinemia
- Growth hormone deficiency
- Hypogonadotropic hypogonadism
- Type 2 diabetes

Behavior/mental health
- Skin picking
- Rectal picking

Food-related behavioral problems
- Temper tantrums
- Difficulty with transitions
- Stubbornness
- Obsessive behaviors
- Perseverant speech
- Obsessive-compulsive disorder
- Psychosis

Miscellaneous
- Temperature instability
- High pain threshold
- Unusual skill with jigsaw puzzles

Adapted from McCandless SE. Clinical report—health supervision for children with Prader-Willi syndrome. Pediatrics. 2011;127(1):195-204.

Table 2. Nutritional Phases Associated with Prader-Willi syndrome		
Phase	**Age**	**Clinical Characteristics**
Phase 0 Decreased fetal movements and lower birthweight	In utero/at birth	• Full-term birth weight and BMI ~15-20% less than siblings • Normal gestational age (typically) • ~85% have decreased fetal movements
Phase 1a Hypotonia with difficulty feeding	0 to 9 months	• Weak, uncoordinated suck • Needs assistance with feeding (e.g., tube feeding or special, widened nipples)
Phase 1b No difficulty feeding and growing appropriately	9 to 25 months	• No longer needs assisted feeding • Growing steadily along growth curve • Normal appetite
Phase 2a Weight increasing without an increase in appetite or excessive calories	2.1 to 4.5 years	• Starts crossing growth centile lines • No increase in appetite; appetite appropriate for age • Will become obese if given RDA for calories or a "typical" toddler diet • Typically needs to be restricted to 60-80% RDA to prevent obesity
Phase 2b Weight increasing with an increase in appetite	4.5 to 8 years	• Increased interest in food; frequently asking "food-related" questions • Preoccupied with food; very concerned about the next meal/snack (e.g., "Did you remember to pack my lunch?") • Increased appetite • Will eat more food than a typical child if allowed • Will eat food within their line of sight if allowed • Will become obese if allowed to eat what they want • Can be fairly easily redirected about food • Can feel full; may stop eating voluntarily
Phase 3 Hyperphagic, rarely feels full	8 years through adulthood	• Constantly thinking about food with an insatiable drive to eat • While eating one meal, already thinking about the next meal • Will awaken from sleep early thinking about food • Will continue eating if portion size is not limited • Rarely (truly) feels full • Will steal food or money to pay for food • Can eat food from garbage and other unsavory/inedible sources (e.g., dog food, frozen food, crayons, etc.) • Typically not truthful about what they have eaten • Will gain significant weight over a short period of time if not supervised • Food typically needs to be locked up; frequently will ask parent to lock food if the parent has forgotten • Temper tantrums and "meltdowns" frequently related to food • Needs a diet that is ~50-70% of the RDA to maintain a healthy weight
Phase 4 Appetite is no longer insatiable	Adulthood	• Most adults have not gotten into this phase and many never will • Appetite may be increased or may be normal or less than normal • Previously in Phase 3, now noticeable improvement in appetite control • Can feel full • Appetite can fluctuate in this phase; the key component is noticeable improvement in control of appetite compared to when they were younger • Not as preoccupied with food • Absence of major temper tantrums related to food

Adapted from Miller JL, Lynn CH, Driscoll DC, et al. Nutritional phases in Prader-Willi syndrome. *Am J Med Genet*. 2011;155A(5):1040-49.

The etiology of the hyperphagia and lack of satiety remain unknown. Several approaches used to address obesity in other populations have been explored for PWS without success. For example, individuals with PWS have very high levels of the gastric hormone ghrelin (which stimulates appetite), both during fasting and after a meal, however pharmacologic suppression of ghrelin does not appear to decrease appetite in people with PWS. Generally, medications such as anorexigenic agents have not been found to be effective in controlling hyperphagia. Similarly, bariatric surgery does not control the weight or appetite, and the morbidity with this surgery is quite high for persons with PWS.

NUTRITION ASSESSMENT AND MANAGEMENT

The role of the RDN is key in the nutrition and weight management for persons with PWS. The nutrition assessment process should be similar to any pediatric patient, including diet, growth, activity, feeding development, and behavior. (See Nutrition Focus 22(3), Nutrition Assessment Guidelines for Children with Special Health Care Needs).[9] However, re-assessment is indicated more frequently, especially in the younger ages.

Growth parameters should be plotted on the WHO or CDC growth charts. Growth charts have been developed based on data from individuals with PWS both with and without GH therapy.[10,11] If PWS-specific charts are used, they should be used in conjunction with WHO or CDC growth charts. Since many children with PWS have short stature, it is critical to monitor weight-for-length, BMI, and/or rate of weight and length gain.

Additional information to include in the assessment:

- Use of growth hormone therapy
- Changes in feeding or food intake
- Current motor or physical activity patterns
- Food intake in other environments, e.g., school, day care, relatives, neighbors
- Food or activity objectives in the Individualized Family Service Plan (IFSP) or the Individualized Education Program (IEP)
- Participation in modified school lunch or breakfast
- Food behaviors – requesting/whining for food, seeking/taking food, temper tantrums around food, eating fast, taking food from others' plates, etc.
- Vitamin-minerals supplements, herbals, complementary medicine
- Access to and availability of food in the child's environment

Nutrient Intake

Energy needs of individuals with PWS are significantly lower than age- and sex-matched peers without PWS. Differences are likely multifactorial, but are thought to be due to altered body composition (higher fat mass and lower fat-free mass, even in infants), hormone dysfunction (e.g., hypothyroidism, decreased leptin responsiveness, and lower fat oxidation), short stature, and decreased physical activity.[5] In addition, hyperphagia and lack of satiety create a challenging long-term situation for persons with PWS and their families.

Clinical experience suggests that during growth an intake of 8-11 kcal/cm permits growth and appropriate weight gain for children with PWS.[12] This is significantly less than the dietary reference intake (DRI) for young children. For the overweight child or adolescent, a diet providing 6-8 kcal/cm should result in a slow rate of weight loss and not interfere with linear growth.[12] These are general guidelines, and will vary depending on the child's activity level, use of growth hormone (once a dose is established and rate of height increases, energy needs are usually increased accordingly), and unique metabolism. But overall, individuals with PWS will always have significantly reduced energy needs.[5]

Children with PWS should eat a varied diet focusing on nutrient-rich foods that meet their needs for growth and development. They need protein, fat, carbohydrate, vitamins, minerals, and fiber, but within a calorie-controlled diet and a consistent meal pattern. Each child's meal plan is developed individually by the RDN in consultation with the family and specialty team.

Since individuals with PWS consume a diet limited in energy and total food, they often need a vitamin-mineral supplement, or in some cases single nutrient supplements, depending on the total diet and their needs.[13] Diet reassessment will determine the need for supplements. Some families also offer other non-nutrient supplements and herbal products, and these should be evaluated for safety and efficacy.

Nutrition intervention and weight management

With appropriate dietary management, a physical activity program, and close supervision, appropriate growth and weight management can be achieved for individuals with PWS. This includes a diet that is designed specifically for the individual, with participation of everyone in the individual's food environment; home, day care, school, vocational programs, work and other settings. Compared to other weight management interventions that stress healthy eating and no restriction, persons with PWS need a diet that is calorie-controlled. Table 3 on page 6 provides general guidance for individual age ranges.

Table 3. General Guidelines for Children with Prader-Willi syndrome	
Stage	**Guidelines**
Infancy	• The goal is to support optimal growth and nutrition with breast milk or formula, either orally or by tube. In some situations, breast milk might need to be supplemented to meet nutrient needs. • Gains in both length and weight may be in the lower range of normal. Therefore, it is critical to monitor length as well as weight regularly during infancy. • Solid foods are usually introduced later due to hypotonia and other motor delays. • The RDN should assure adequate intake of fluid, macro- and micronutrients.
Toddler	• Transition to solids and self-feeding, but many still rely on formula past 12 months of age. • Usually not much increased interest in food yet.
Preschool	• More likely to see preoccupation with food, increased intake and weight gain. • Child may be receiving food in other settings, so the family needs to provide information and guidance about food.
School-age	• Important to help family determine diet management at school, day care, camps, parties, and family events. • Food-seeking behaviors and tantrums may escalate. Behavioral counseling for the child and family may be helpful.
Adolescent	• Some individuals will demonstrate increased behavior challenges and psychiatric problems (especially those with UPD type). • Families need to plan for the future, especially for living and work environments, so that food accessibility and intake is limited.

Nutrition education and anticipatory guidance should be initiated when the diagnosis is confirmed. As the child grows older, everyone with whom the child is in contact needs to be included in the plan for control of the food environment and energy intake. The diet management approach should be tailored to each family, whether counting calories, using meal/snack plans with portion control, or using other methods. The choice should be workable for the family and provide an adequate nutrient intake that restricts energy intake and excess weight gain. Preventing weight gain is far superior to needing to implement the very restrictive diets needed for weight reduction. A proactive approach is key, care providers can help families put routines in place before they are actually needed to promote consistency.

Weight-for-length or BMI of individuals with PWS needs to be monitored frequently. One can expect periods when food will be obtained regardless of the controls put into place. Finding the source of the extra calories, and redefining the energy intake levels and goals for growth and weight management will be required. It is clear, however, that with an appropriate energy intake and controlled environment the medical complications of obesity can be avoided.

Food-seeking behaviors are present in most but not all individuals with PWS. These behaviors require rigorous supervision of the food environment by the family and those involved in the child's care. It is necessary for families to lock cupboards and refrigerators.

Food availability away from home is always a problem; adults in those settings need guidance on how to provide food but also how to limit access to it by the child with PWS. Some parents, whose child's weight is in good control, report that well-meaning relatives and school staff want to give the child a snack or treat "just this one time." They do not appreciate the diligence the parents have put into making the child's program successful, and that this additional food presents a problem to be solved. Table 4 on page 7 lists strategies that have been effective and ineffective in efforts to prevent obesity in individuals with the PWS. Table 5 lists a variety of situations for children and adolescents where food may be present.

School Meals

Children with PWS can participate in the school breakfast and lunch program with a written prescription for modified meals at the appropriate calorie level. In addition, diet and activity-related objectives can be included in the Individualized Family Service Plan (IFSP) or Individualized Education Plan (IEP), e.g., specific policy for preschool snacks, how to handle classroom parties, monitoring food access between classes in middle and high school, accommodation for physical education class, or a walking program at recess.

Table 4. Strategies to Prevent of Obesity in Children with Prader-Willi syndrome	
What Works	**What Doesn't Work**
Diets with energy intake individualized for the child	Using the DRI to establish appropriate energy intake
Controlled food environment	Access to food in many settings
Total family support for the weight management program	One or more family members not participating in the program
School support, including goals in IEP or IFSP; modified school meals	Food given by classmates; child with PWS giving money to friends to buy food; school parties/treats not planned
Child participation in menu planning but not portion size	Family-style meal service
Planning diet so that treats and parties are accommodated	Adding parties and snacks to the planned diet
Adjusting menus and energy intake when food is taken	Ignoring extra food taken
Exercise planned as part of the daily routine, including incorporated into the IEP (e.g., daily PE or walk with para-educator)	Lack of regular physical activity
Structured, scheduled meal and snack times	Access to food throughout the day
Visual schedules and supports to define meal and snack times	Unpredictable meal and snack timing

Table 5. Food Access Opportunities to Monitor for the Child with Prader-Willi syndrome

- sack lunches on a school bus
- sack lunches/vending machines in an unsupervised setting at school, work, lounge, or hall
- garbage containers available on the way to the restroom, in a park, etc.
- food left for pets
- visits and/or day care at a home where cupboards/refrigerators are not monitored
- store displays or community events offering free samples
- school or family parties where children can take as much food as they want
- work or study situations, i.e., classes that prepare food and/or work in a cafeteria/restaurant
- community/church activities where food is present

Physical Activity

Activity planned as an integral part of the daily routine is an important part of the weight control program. Individuals with PWS typically prefer activities requiring minimal movement. The child with PWS can be included in all the usual family activities and outings. At school children should be supported to be active during recess as well as participate in physical education. Non-competitive activities such as swimming and walking are good; an aerobic exercise program with simple routines can be incorporated into the child's activities.

FAMILY STORY

Families and their consistent efforts, day in and day out, are what make the diet/weight outcomes of children with PWS successful. But it is not easy, especially when dealing with challenging behaviors. Following is one mother's story of their family's experience thus far, with a daughter who has PWS.

Natalie

Our daughter is a beautiful, happy, loving and determined 3½ year old, who can walk, ask and answer questions, name that tune in three notes, tell you her birthday, tell you the names of everybody in her class and remind us that we are out of grape-flavored fluoride and need to have the prescription filled. She enjoys reading books, dancing, listening to music, singing, painting, swinging, riding her trike and has a strong sense of fashion. Our daughter, Natalie, has a medical diagnosis of Prader-Willi syndrome (PWS).

Our medical team consists of our local pediatrician and the following specialty clinics: ophthalmology, pulmonology, genetics, dentistry, endocrinology, sleep specialist, orthopedics, orthotics, otolaryngology, and audiology. We also continue follow-up with an RDN in the local health department for frequent height and weight checks and the RDN who is part of a team at a university center. It would be our desire to have even more of a team approach, with all care providers meeting and discussing our case but limited resources prevent this. We use the word team as all of these people are specialists in their field and are able to apply their knowledge for Natalie and her diagnosis of PWS.

We currently experience: strict caloric restrictions, scoliosis (curve of 46 degrees), obstructive sleep apnea, accommodation esotropia, low muscle tone, and more recently restrictive lung disease. We participate in growth hormone therapy, ½ liter of oxygen when sleeping, occupational therapy bi- monthly, physical therapy weekly, and Rolfing bi-monthly. We would love to do more for Natalie but insurance is limited to 30 total therapies per year (OT, PT, SLP). We have been asked to share our story in relation to nutrition in how we manage PWS.

Figure 1. Natalie

Our story is much the same as any other person that is surprised by the birth of their child with special needs. Our world was turned upside down. I feared that our marriage would not endure the challenges that Natalie would bring. I feared she would not survive. Day by day we learned that we were making it, we saw that Natalie was gaining strength. We learned that we could adapt and do things differently. We learned that our love and respect for one another was deeper than we could have ever imagined, and that our family would stay whole as we navigate the world of PWS and establish our, "New Normal."

We were fortunate to have an early diagnosis at 2 weeks of age, along with answers to why our new beautiful baby girl was so floppy, sleepy, and not strong enough or motivated to consume her nutrition orally. A nasal gastric (NG) tube was placed and remained for 10 months. During those 10 months, she slept greater than 22 hours per day for the first 4 months with oxygen via nasal canula.

We fed Natalie every 3 hours during the day with oral feeds via a binky trainer (10 cc tastes), then transitioned to a newborn nipple (Avent) with 10-30 cc and finally to a bottle with increased volumes. Night-time feeds were given via NG using a pump. Natalie was offered oral feeds of pumped breast milk 5 of the 8 feedings, the unconsumed milk was then provided via NG tube. I was fortunate to be able to pump breast milk for 9 months for Natalie prior to transitioning to formula.

I expect that most physicians would disagree with the 10-month-long NG tube use; however, Natalie had very low tone, with little independent movement, and the NG tube had little impact on her development. Gastroesophageal reflux (GERD) seemed to be managed via medication and environmental modifications. Growth hormone (GH) therapy was initiated at 6 months of age. The NG tube could have been pulled sooner, however, we embraced the safety net of having a tube to ensure that she was meeting her energy needs.

We had monthly length and weight checks with the RD at the county health department. We were encouraged to have fewer checks, but it was our experience that Natalie's weight could get off balance quickly, despite careful management. Monthly length and weight checks has allowed for the affirmation that we were meeting and not exceeding her nutrition and hydration needs.

At 3 ½ years of age, Natalie can feed herself with utensils, drink from an open cup and is willing to try new foods. She has food likes and dislikes. She doesn't really care for water and has difficulty chewing hard veggies and meats, secondary to the low muscle tone. Despite Natalie's progress with consuming all of her foods and liquids orally and maintaining an acceptable BMI, we are planning for the hallmarks associated with PWS; the insatiable appetite, food seeking, tantrums, and the overwhelming drive to consume food. We have read that the drive to obtain food can be so overwhelming that it can get in the way of learning, experiencing life, cause tantrums, and cause obesity leading to secondary medical issues such as obstructive sleep apnea, heart disease, diabetes, etc.

Have we reached these hallmarks? We don't think so, but we will do everything in our control to prevent the opportunity for unmonitored food access, by observing, anticipating, providing structure, clear roles, clear expectations and most of all, prevention. To date we do not lock the pantry door or the refrigerator, and we are able to have a fruit platter in the corner of the kitchen. In the eye of prevention, our home is such that Natalie is never alone in the kitchen, and this seems natural. We are observant and realize that if Natalie demonstrates more interest in the pantry, refrigerator or fruit platter, that accessibility will become part of the past.

Over the last 3 years, we have had different caregivers for our children. It has become evident that what our family has started to do naturally isn't natural at all, but rather an adaptation to how our "new normal" has to be. We have based our new normal on hours of reading, personal experience, and going to PWS conferences. In establishing our "new normal" the single most important element that we have to embrace entirely is: it is our job as parents of individuals with PWS to create a safe environment that is free from the temptation of food. It is NOT the responsibility of the individual with PWS to be "good" and not to take food. We need to under-

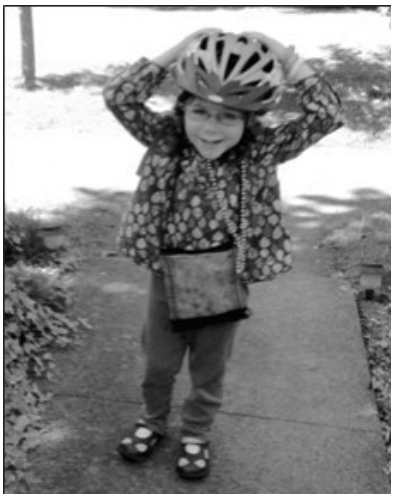


Figure 2. Natalie going trike riding

stand that Natalie has an internal drive to eat, just as we all have an internal drive to breathe.

Mealtime strategies

Our family has three very different caloric needs -- an athletic father that has daily caloric needs of 4000-5000 calories, an early elementary age boy who is active with an irregular eating pattern, a mother that adapts to the situation at hand, and Natalie needing a strictly fixed current caloric intake of 910 calories per day. With these very different needs we are doing our best to adapt. In our family there is no fairness, we do what is right for the individual, in relation to food, toys, activities, media, reading, school, etc. With relation to food, Natalie has an efficient body and only requires 910 calories per day to grow. Strategies for mealtime that have evolved for us (and are a work in progress) are listed in Table 6 on page 10.

Tools for mealtime management

Here are necessary tools for us to calculate calories and amounts:

- Nutrition book with calories for home use; travel nutrient book for outings (or an app for care providers to record and communicate intake)
- Yellow highlighter to mark frequently used foods
- Journal to write down frequently used bulk foods and amounts
- Scale – one that has grams and ounces
- Calculator
- Knowledge of conversions, e.g., 1 cup has 16 tablespoons and there are 8 ounces in a cup
- Permanent marker to write calculated figures on cracker box, pasta, and bulk foods
- Scissors to cut out food labels of bulk items
- Glass measuring bowls (3/4 cup) to serve food in and weigh meats
- Food chopper has been a necessity to chop foods like meats or veggies that Natalie has difficulty chewing
- 6 oz. juice glasses for her milk

We have created a sitter handbook to assist caregivers with the expectations of the household since we found that orienting a sitter seemed a bit overwhelming with the details. The sitter book consists of a three ring binder: house address, lists of sitters, emergency contacts, household expectations regarding food, calories, kitchen, media, diet, behavior, expectations for the park or outings, nap and toileting, current skills with reference to motor skills and safety, suggested activities, and pet information. Our sitters have reported this to be a useful tool as well as my husband when mom isn't available.

How are holidays managed? First of all, any family member of an individual with PWS will tell you that the holidays and celebrations with food are the most stressful times. We have learned that we cannot depend on others, including family, to truly understand the magnitude of the issues. While the family is supportive and has the best intentions, the level of follow through required to prevent unwanted food access is not a realistic expectation to have of others outside the primary family unit. Our goal is to prevent food access and deal with it appropriately, for Natalie and the people around us.

An example: We were invited to birthday party that had a child size swimming pool filled with Cheerios. We walked in and my heart sank with "How will we ever manage this one?" We greeted the guests, and then Natalie joined the other children in the swimming pool. Natalie watched the other children eat the Cheerios. She seemed to know that it wasn't a mealtime. After about 20-25 minutes Natalie started to place the Cheerios in her mouth. I was diligent and positive, "Not for eating" or "Cheerios out." Before it became a power struggle, we got a cup of Cheerios from the box and had Natalie sit on our lap to eat this snack. We focused on her using her pincer grasp to pick up each Cheerio. This seemed to be the best alternative for us and for the family member putting on the party. Additional ideas for management at events with food are listed in Table 7 on page 12.

We plan food-free celebrations such as egg hunts (including plastic ones filled with stickers, etc.), singing songs, games and activity, face painting, giving play-doh or pencils, and costumes. For Halloween/Harvest, county fairs, festivals, etc. – we approach these celebrations as a family, with a focus on the experiences rather than food. We highlight experiences such as music, face painting, costumes, stories, shows, and animals. We often bring our own lunch and purchase a snack that would meet our caloric allotment for that snack. I estimate the caloric value. I find estimation to be one of the most challenging components of calorie counting. I like accuracy and it has been quite a process to allow myself to relax and know that sometimes, estimating has to be accurate enough.

Physical activity management

In addition to diet management, physical activity is very important part of our every day. Each daytime schedule allows time for focused exercise, trike riding, walking to the park, early morning pajama walks/ hikes, jumping, ball time, swimming, walking, kid yoga, dancing, swinging, climbing, and scooter board. Our home is very much a home with built in opportunities for physical activity. We also lead by example. Both my husband and I lead active lives and take our children along whenever possible. Bicycling, kayaking, rowing, hiking, swimming, and wrestling are activities are encouraged and supported using the phrase "we enjoy moving our bodies." We allow Natalie time to complete a task independently and do not rush her. Finding the additional patience and time has been a work in progress. However, we know that providing more op-

Table 6. Mealtime Strategies Used by Natalie's Family

- As a family we attempt to make healthy choices at the market.
- Natalie can help with food preparation as her sibling does (mixing, pouring, etc.).
- We don't add extra fats or creams; we use plain yogurt, applesauce or water when possible.
- We use a small plate in order to give the appearance of more food, use smaller utensils with sweet foods (ice cream) to provide the opportunity for extended tasting time.
- We have 4 meals per day with meals dished up on the plate prior to presenting them to each person; Natalie's meals are divided into three meals: 200 calorie s+ 67 calorie of milk and one snack: 110 calorie snack with water.
- When Natalie asks for more food, I direct her to her milk: "If your tummy is still hungry, you can drink your milk."
- When the food and milk are gone: "Your food is gone, your tummy is full," all stated in a matter-of-fact manner, being aware of not providing any sense of feeling sorry for her, yet compassionate to her feelings.
- If there is a food that I anticipate Natalie will want more of, I only give her ¾ of the caloric allotment, and then provide her with additional food when she asks. The statement would then be, "When the blueberries are gone, your tummy will be full," "This is your last cracker." She does well as long as she knows that the caregiver will follow through.
- Desserts are attempted to be natural (fruit, yogurt), however sometimes we have ice cream. Natalie will get 1 tablespoon in her dish. The calories would then be shifted throughout the day in anticipation for dessert.
- With prior knowledge of a birthday party, high volume/low caloric foods will be offered below the usually caloric expectations at prior meals so that there will be 100 calories for the party. Often Natalie and mom will share the cake and ice cream with mom having control of the spoon/fork; this has been accepted well.
- Food or drinks are never used as a reward, NEVER.
- Food or drinks are never used as a distraction.
- When Natalie asks to eat at times other than mealtime, we confirm that her words were heard, "I hear you, that you are hungry. We will eat after we pick up Jonas from school."
- We do not offer breakfast before 7:00 am and no food is offered after dinner.
- Parents and siblings do not hide or sneak food. If she asks us what is in our mouth, we tell her. On the rare occasion that food is eaten by a family member other than at mealtime, that person can do one of two things: (1) say "This is not for Natalie," or (2) give her one or two bites with the caloric value being subtracted over the next meals. Mom or Dad need to anticipate that Natalie or her brother may ask for a bite and make the decision of sharing prior to eating, before any undesirable behavior is displayed.
- If Natalie doesn't eat all of her food at a given meal, those calories are added to another meal. We want to encourage her to leave food on her plate rather than encouraging her to clean her plate.
- Juice is rarely offered, yet if it is, it is diluted 1 to 4 (juice to water).
- We talk about our food when we are eating it, just as we would do with any child enjoying their meal
- No values are placed on foods; we offer food that she may not like and add/subtract the calories as necessary. We offer dessert last as to know how many calories can be offered.

portunities for Natalie to perform everyday tasks will build strength, coordination, self-confidence and independence.

In the event of a tantrum

We acknowledge her feelings, stay near to keep her safe, hold/hug her if she allows, WAIT until it is over. She may ask me to leave. Consequently I give her some space yet in view but not looking at her. The tantrums are usually short as we are consistent. Caregivers that have difficulty with expressions of displeasure often get more screaming and tears. If she learns that the expectations can be altered to her benefit with tears or screaming she will use it as often as necessary to fulfill her wants.

Case Study Summary

Establishing our "new normal" has been a process. At times the vast profile of PWS has been overwhelming. This process initially was a steep learning curve, then seemed to come more gradually. The profile of PWS continues to evolve and as it changes we return to the strength of our family and read, learn, ask questions and trust that we are the experts on Natalie. It is our responsibility to navigate the medical system and receive the knowledge of providers to best meet her needs.

Natalie is truly loved and adored by all those who have the pleasure of meeting her. Her smile can warm the hearts of an auditorium. She has Prader-Willi syndrome, yet she is an individual first and with prevention and clear expectations, Natalie can eat to be healthy with the expectations that her caregivers keep the environment safe for her to laugh, love, learn and live.

CONCLUSION

Earlier age of diagnosis and increased awareness of PWS has made it possible to provide early medical and therapy/education services. This early provision of services has allowed management of complications before they become severe, and more likely prevention of excessive weight gain. Although research continues on PWS, current efforts still rely on medical and weight management, behavior intervention, education strategies, and long- term planning for adulthood. The RDN is a key team member for individuals with PWS throughout their lives. But it is the family who works within the "new normal" as mentioned in the parent story above, and must coordinate with many disciplines, providers, environments, and issues that arise over time.

Let's revisit the children described at the beginning of this issue:

Max

Max, a 6-month-old infant, has recently had a gastrostomy tube placed after receiving breast milk via NG tube due to significant hypotonia. His length is <5th percentile, weight is at 5th percentile, and weight-for-length is 50th percentile.

Max's presentation is typical. Nutrition support is needed to help him maintain an appropriate weight-for-length and good nutrition status. In addition to addressing immediate medical and nutritional needs, the team working with Max's family discusses potential issues and helps them identify proactive strategies that will work for their individual family.

Maddie

Maddie is 4 years old and participates in Head Start. Her parents report that she asks for food between meals and regular snacks, and will often have tantrums if denied. The Head Start meals are served family style, but Maddie's parents would like to have more control over her intake. Her BMI has gone from the 75th to the 95th percentile in the past 6 months. Her recommended energy intake has been 10 kcal/cm.

Maddie's parents met with the Head Start staff and provided documentation from her medical team that family style meals are not appropriate for Maddie. Maddie's lunch and snacks are served to her pre-portioned, and her teachers monitor to ensure that she does not take food from other children (even if they offer it to her).

Drew

Drew is a 12-year-old middle school student who moves from classroom to classroom during the day. Although he has a modified low-calorie school lunch (total of 350 calories), he frequently begs more food from classmates or sneaks food from their sack lunches. Teachers feel it is not fair to deny Drew classroom food treats. He prefers sedentary activities, so doesn't mind being kept in at recess for getting into trouble. His BMI in the past two years has fluctuated from the 90th percentile to >95th percentile.

Drew's parents and RDN provided a short in-service to Drew's teachers and paraeducators. With a better understanding of PWS and the use of consistent routines and messages (see Table 8 on page 13), they became more comfortable being firm when telling Drew "no." It was helpful for his teachers to hear that Drew is less anxious at home when he knows that food is locked and he cannot access it. A physical activity goal was added to Drew's IEP, and he takes daily walks with a paraeducator and peer mentor. See Figure 3 on page 13 for an example of the letter that is kept in Drew's health folder.

Cody

Cody is 17 years old and in high school special education. His single mother has worked hard to keep his weight and diet in good control over the years, with ups and downs. Although her other two children haven't always helped, now that they are older they understand and are supportive. In the past few years Cody has developed symptoms of obsessive-compulsive disorder (OCD) and receives counseling and medication. His mother is worried about his future – what he will do after high school graduation, how will his diet be managed, and who will take care of him when she can't? His BMI for the past 3 years has been 75th to >95th percentile.

Cody's care had previously been managed by his pediatrician who consulted with some specialists with experience with PWS. Recently, he was referred to a PWS clinic at the children's hospital that is a 4-hour drive from his home. The team has been working with Cody and his mother to plan for the future. He can receive special education services until age 21, the team is identifying state and local resources, and Cody and his family are exploring assisted living opportunities.

Table 7. Management Strategies for Events Away from Home

- An adult (mom/dad) is always with Natalie or giving a watchful eye if food is present. She is not to receive food from anyone but mom at a party.
- She is to ask for food prior to eating. "Thank you for asking," with a bright smile.
- If Natalie were to pick a cracker off the floor, the parent would calmly retrieve the cracker from her hand or mouth along with a "Thank you for giving Mommy that dirty cracker. I will throw it away." In the event that she does eat and swallow the morsel from the floor, I would thank her for telling me and note the caloric content.
- Mom and Natalie are usually a team and Dad and Natalie's brother are a team for food. Dad attends to the brother and Mom attends to Natalie, consequently most of our needs will be met.
- We sit away from food.
- We visually scan the floor for any dropped food that may be a temptation and gracefully pick it up. We are very discreet with picking food up off the floor, both for Natalie and the hostess.
- We carry hand sanitizer.
- Bring age appropriate activities for when we are done eating.
- Calculating calories can be difficult at outings. We do our best to estimate and trust that we meet our 910 calories per day 90% of the time.
- We usually discreetly bring food as a backup and use if needed.
- If Natalie doesn't notice or ask for food/cake we don't offer it. However that usually means that mom doesn't eat any sweets either.

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RESOURCES

Prader-Willi Syndrome Association (USA)

http://www.pwsausa.org/

Provides information, support, and mentoring for families, supports research, and provides information and materials for families and professionals.

Foundation for Prader-Willi Research

http://www.fpwr.org/

Supports research and therapeutic development for Prader-Willi syndrome

International Prader-Willi Syndrome Organisation

https://www.ipwso.org/

Supports individuals with PWS and their families worldwide

Table 8. Attitude is Important	
PWS is a chronic condition that affects every aspect of a family's life. Families note – and health care providers observe – that attitude and approach is important to establishing and maintaining the consistent routines that are critical to good outcomes.	
Caregivers' attitude	**Rationale/explanation**
An exception becomes an expectation	When an exception to the rule is made individuals learn to expect the special, "just this once" treat.
Be matter-of-fact and calm	Although a calm, matter-of-fact approach can be difficult for a worried parent, messages given without the stress of underlying emotion are better received (and understood).
No doubt, no hope, no disappointment[1]	This approach can be helpful for evaluating self-regulation: • No doubt: the person with PWS knows how much, what kinds of foods, and when the food will be served • No hope: there will be no opportunity to obtain additional food • No disappointment: there is not an emotional let-down due to false anticipation or unfulfilled expectations about food
Everyone is unique, and different family members need different amounts of food to stay healthy	It can be helpful to explain and reinforce differences between individuals. Starting to become comfortable with differences early (e.g., young child at home) may make adjusting to differences in other situations (e.g., at school) easier later.
It's okay to be different and to eat differently than classmates and siblings	Reinforcing the concept of "different rules for different people" can help families and care providers consistently adhere to established limits.
References: (1) Gourash LM and Forster JL. Regulation of weight in Prader-willi syndrome: theoretical and practical considerations. Online: https://www.pwsausa.org/wp-content/uploads/2016/01/Self-Regulation-in-PWS-by-LG-final-.pdf. 2009. (2) Leavitt AM. Clinical experience.	

To whom it may concern:

_____ is a __-year-old child with Prader-Willi Syndrome (PWS). Information about his/her PWS diagnosis should be included in the medical section of his/her IEP.

PWS is a rare genetic disorder that causes hypotonia (low muscle tone), cognitive impairment, and obesity secondary to an insatiable appetite and food-seeking behaviors that cannot be controlled. _____'s food environment at school must be closely controlled, with no unsupervised access to food at school, and food cannot be used as a reinforcer. He/she will always need constant adult supervision at school, even as he/she gains independent skills as he/she gets older, including coming and going from the bus, as well as when he/she is at recess or in the hallways. Any food seeking behaviors that he/she may eventually exhibit should not be viewed as "stealing." Consequences and other efforts to shape behavior around food will not be effective. The drive for food is something that children with PWS cannot control. Instead, they need scaffolding. Rather than reacting to episodes of food seeking, they need to be prevented by appropriate supervision.

As they get older, individuals with PWS will forage for food in garbage, and in the cafeteria will eat other students' leftover food. He/she cannot be allowed to go through the cafeteria line at lunchtime and choose his/her own foods without supervision. For specific guidelines about what _____can eat at school, his/her teacher or school nurse can contact _____'s PWS nutritionist, NAME and CONTACT INFORMATION). There needs to be clear daily communication from school to home about exactly what food and drink were consumed at school.

These measures will be most effective if they are implemented now, before they may be necessary. Being proactive and preventing problems is preferable to reacting once problems have emerged.

Sincerely,

Figure 3. Sample IEP Letter Explaining Prader-Willi Syndrome

LETTER FROM THE EDITOR

Dear Nutrition Focus Subscribers,

January 1986 – Volume 1 published Nutrition News – Improvement of Nutrition Services for Children with Handicaps or Chronic Illnesses. This first issue, in a series of nutrition newsletters from the "Nutrition Services for Children with Handicaps" project, was printed with 35 copies sent to 10 states including Alaska, Idaho, Oregon, Colorado, Montana, North and South Dakota, Wyoming, Utah and Washington. My colleague, Betty Lucas, had received funding from the Maternal and Child Health Bureau to provide consultation and continuing education in these states for RDNs who were seeing more children with special needs. The states had requested a newsletter to provide practical and current information and resources. At the end of the project's 3 years, there was encouragement and demand for the six issue per year newsletter to continue. The publication transitioned to subscriptions for anyone across the US and the world.

After 33 years as editor of the newsletter, I am retiring, handing those duties to my colleague Beth Ogata here at the University of Washington.

In the beginning, little information was available about nutrition for this population.Thirty-three years have seen changes in terminology, name, and availability but my goal as editor remained the same – to provide practical and current information and resources primarily for RDNs but also other subscribers including nurses, physicians, therapists, early intervention centers, hospital libraries and others. I wanted to publish an issue that a health care provider - who does not see a child everyday with Down syndrome, feeding issues, cerebral palsy, Autism Spectrum Disorder or any other special health care need - could read and plan for their interaction with the family and this child related to growth, feeding, and nutrition.

Today the internet opens our world to information about many special health care needs. However, *Nutrition Focus* remains a resource concentrating on specific growth, feeding and nutrition concerns, supports the RDN who provides nutrition services, and educates other health care professionals and families about the importance of nutrition in this population.

I want to thank all of you who have subscribed to our newsletter and sent messages of support and encouragement over the three decades. Children with special health care needs, and their families, benefit from nutrition guidance as part of the services to ensure each child's growth and development. We hope the *Nutrition Focus* newsletter continues to help each of you in your daily work with families and their children with special health care needs.

With appreciation,
Sharon Feucht, MA, RDN, CD

THANK YOU, SHARON FEUCHT!

Thank you, Sharon for all of your hard work with *Nutrition Focus*. I am grateful for your vision, dedication, skillful editing, and so much more. It has been a pleasure to watch the articles evolve – from an idea (often based on issues that you or colleauges have seen in practice, or new advances or knowledge in nutrition and children with special health care needs), to a connection with an expert in the field, to a draft, and finally to a polished article. Your work continues to provide valuable resources to RDNs and others.

Congratulations on a well-deserved retirement. (But don't worry, we won't let you get away too easily. Readers, look for an article authored by Sharon in the next few months!)

Best wishes,
Beth Ogata

Continuing Education Opportunity

To participate in this continuing education opportunity login to the Nutrition Focus website http://depts.washington.edu/nutrfoc/webapps/?page_id=1345 and select "Continuing Education" to access the quiz related to this issue. You must correctly answer 80% of the questions to pass. Cost is $20 (subscribers) or $40 (non-subscribers) for 2 CPEU.

1. Individuals are being diagnosed with Prader-Willi syndrome (PWS) at younger ages than in the past. Which of the following explains this change:
   a. The prevalence of PWS syndrome is increasing
   b. Molecular testing techniques makes genetic testing possible
   c. Characteristics of PWS are more widely recognized than in the past
   d. Recognition of the benefit of early intervention led to increased screening

2. Which of the following was NOT listed as a clinical or developmental characteristic of PWS:
   a. Short stature, slow growth
   b. Hyperphagia, obsession with food after infancy
   c. Specific physical features, i.e., long fingers and toes
   d. Hypotonia, poor suck, and failure to thrive in infancy

3. Which of the following providers would be most likely to be on a multidisciplinary team for PWS management:
   a. Psychiatrist
   b. Social worker
   c. Endocrinologist
   d. Exercise physiologist

4. True or false: For individuals with PWS, growth hormone (GH) therapy has positive effects on linear growth, lean body mass, and motor development and can help prevent obesity when combined with diet management.
   a. True
   b. False, GH therapy will not prevent obesity
   c. False, GH therapy will not affect an individual's motor development
   d. False, although it has positive effects on linear growth, GH therapy will not have positive effects on lean body mass

5. Although this can vary from individual-to-individual, an 11-year old with PWS is most likely in which of the nutritional phases that were described by Miller, et al:
   a. Phase 2a – Weight increasing without an increase in appetite or excessive calories
   b. Phase 2b – Weight increasing with an increase in appetite
   c. Phase 3 – Hyperphagic, rarely feels full
   d. Phase 4 – Appetite is no longer insatiable

6. Temper tantrums and "meltdowns" frequently related to food are characteristic of which of the nutritional phases that were described by Miller, et al:
   a. Phase 2a – Weight increasing without an increase in appetite or excessive calories
   b. Phase 2b – Weight increasing with an increase in appetite
   c. Phase 3 – Hyperphagic, rarely feels full
   d. Phase 4 – Appetite is no longer insatiable

7. Which of the following was NOT identified as information that is helpful to include in the nutrition assessment of a child with PWS:
   a. Vitamin-mineral supplements
   b. Use of growth hormone therapy
   c. Food intake in all environments, including school, day care, relatives, neighbors
   d. All of the above are helpful to include

8. Individuals with PWS have different energy needs than age-matched, typically developing peers. Which of the following is true:
   a. GH therapy will normalize energy needs, so the DRI can be used to estimate calorie needs
   b. Although they can vary, energy needs are generally 50-75% of usual energy intake of those without PWS
   c. For the overweight child or adolescent, a diet providing 16-20 kcal/cm should result in a slow rate of weight loss and not interfere with linear growth
   d. Infants with PWS have energy needs that are higher than their typically developing peers, but energy needs of older children with PWS are significantly lower

9. Why do most individuals with PWS need a vitamin and mineral supplement?
   a. The diets of children with PWS are typically low in nutrient-dense foods
   b. Their needs for most vitamins and minerals are higher than individuals without PWS
   c. It is difficult to meet vitamin and mineral needs and adhere to the necessary calorie restriction
   d. None of the above; children with PWS should be able to meet vitamin and mineral needs with food

10. Which of the following was is true about physical activity for the child with PWS:
   a. Individuals with PWS should avoid high-impact activities because of joint problems
   b. Individuals with PWS need more physical activity than typically developing peers
   c. Activity planned as part of the daily routine is an important part of the weight control program
   d. At school, children should be supported to be active during recess but should not participate in physical education

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