Feeding Issues and Nutritional Phases in PWS

Prader-Willi syndrome is the most commonly known genetic cause of obesity which can create great concern for many new parents. It is also commonly known that the hallmark symptom hyperphagia, or increased food drive, can cause myriad problems if left unmanaged. It is therefore critical for parents to understand the physiology and behavioral symptoms related to PWS as well stay knowledgeable about appropriate management strategies that will help the individual with PWS live a healthy life.

Through studies, seven distinct nutritional phases and sub-phases have been identified. If diet is not properly managed, obesity in PWS typically begins between 2 and 4 years of age. As infants, there almost an absence of an appetite drive. The appetite gradually increases in early childhood such that by about 8 years of age the individual with PWS is showing signs of an insatiable appetite (hyperphagia). Food security and environmental controls are needed and enforced based on the nutritional phases.

Infancy: Generally, infancy is characterized by "failing to thrive" and is not the period during which you need to be concerned with hyperphagia symptoms. Phase 0 begins in utero and is characterized by decreased fetal movements and lower birth weight. From birth to approximately 15 months the infant is in Phase 1. This is characterized by hypotonia and decreased arousal levels that make it difficult for infants to maintain and sustain an adequate suck. While there have been some babies who have been able to breastfeed, it is more typical that special feeding techniques will be necessary to help your baby receive adequate nourishment so that he or she gains appropriate weight. If your baby has difficulty sucking and swallowing breast milk or formula from a regular bottle nipple, you may want to discuss with your pediatrician the use of special nipples, such as Nuk nipples or the Haberman nipple system. Some occupational therapists also specialize in feeding issues and can be a helpful resource. If your baby's hypotonia and coordination interferes with their ability to take-in adequate calories, your physician may recommend using a nasogastric tube, or NG-tube, for a short period of time. NG-tube feedings can be used alone or in conjunction with bottle feedings. If feeding remains difficult for a prolonged period of time, a gastric feeding tube, or G-tube, inserted directly into the stomach, may be recommended.

Whichever method works best for your baby, it is important that he or she receive the proper calories to sustain healthy growth and brain development. Whatever feeding technique you use, you may want to bear in mind that some mothers have found it especially bonding to allow their baby to nuzzle the breast after feeding. And remember, as your infant grows older and stronger, feeding issues will resolve. By the end of Phase 1 most infants will begin to have a normal appetite, little to no difficulty feeding, and will begin growing appropriately on a growth curve.

While the Federal Drug Administration (FDA) has not yet approved the use of human growth hormone medication in infants, the current standard of care worldwide is to begin growth hormone as soon as the diagnosis of PWS has been made. It has been reported that infants treated with growth hormone generally improve their suck at a faster rate and experience greater success bottle feeding, and sometimes even breast feeding, than infants who are not treated with growth hormone. The supplement Coenzyme Q-10 may also improve feeding issues in infants, increasing energy levels, and muscle function and coordination. Coenzyme Q-10 supplement may help some babies avoid the need for a G-tube, although for some it may not. Refer to the *Supplements* and *Growth Hormone* sections of this Handbook for more information.

Early childhood: The second main phase occurs when the weight starts to increase and crosses growth percentile lines. This generally begins between 18 and 36 months of age. Sub-phase 2a is begins approximately 2 to 4.5 years when the child's weight increases without an increase in appetite or calories. At this time your child has an appetite appropriate to their age, but will become overweight if given the typical recommended daily allowance of calories or if eating a ''typical'' toddler diet. Current research indicates that early childhood obesity reduces IQ points, therefore it is important to help your child maintain a healthy weight during toddlerhood and throughout childhood. Make sure the overall diet includes an appropriate amount of fat, carbohydrates, and fiber each day.

Childhood: Between ages 4 and 8 your child will enter Phase 2b, when their appetite and interest in food increases. They may begin to exhibit the typical symptoms of hyperphagia, or the food drive, such as a desire to eat more food after finishing a meal, increased anxiety around food, and they will gain weight if their food intake is not monitored. Phase 3 begins at approximately 8 years and lasts through adulthood. During this phase the individual's interest in food can be transient or constant.

For most children the symptom of the hyperphagia food drive starts subtly, while for others it can begin intensely. The degree of hyperphagia also varies; some children may not ask for food unless it is snack or meal time or unless it is being eaten in front of them, while others express a desire for something to eat on a more continuous basis, including shortly after finishing a snack or meal. The degree of hyperphagia may also change from day to day; one day your child may express feeling hungry frequently, while the next day there may be an absence of excess hunger. Remember, hunger patterns of children without PWS can also vary from day to day, depending upon the child's activity level, growth spurts, etc. Children taking growth hormone medication or a child going through a growth spurt may be able to take in more calories without weight gain.

For children of all ages with PWS it is critical throughout the lifetime to:

- Establish a routine and schedule for meals and snacks as early in life as possible. It is recommended to schedule small meals and snacks at 2 ½ to 3 hour intervals. For example, breakfast at around 7:00 a.m.; morning snack at around 10:00 a.m.; lunch at around 12:30 p.m.; afternoon snack at around 3:00 p.m.; dinner at around 6:00 p.m.; bedtime 2-3 hours later.
- Consult with a registered dietitian who has up-to-date knowledge about PWS or will pursue it to ensure your child's total daily caloric intake does not exceed what his or her body can expend, which in general is a little more than half of that of an individual without PWS;
- Implement the Principles of Food Security as outlined in the Food Security section of this Handbook;
- Ensure physical activity (play, exercise) every day

Adulthood:

Some researchers suggest there is a Phase 4 in adulthood where the individual appears to have more control over their appetite and food-related behaviors. In this phase it is suggested that the person's ability to feel full may fluctuate and they may not be as preoccupied with food.

It is far easier to maintain a healthy weight than it is to lose excess weight that has been gained. Eating healthy foods is important not only for your child with PWS but for *every* member of the family!

Resources

Nutrition for Infants and Toddlers, published by PWSA | USA

Nutrition for Ages 3-9, published by PWSA | USA