Barth Syndrome Foundation

Nutrition Fact Sheet

NUTRITION FACTS IN BARTH SYNDROME

Nutrition can be a complicated issue for the Barth syndrome individual. Heart failure, fatigue, and various complications of neutropenia can limit appetite and lead to nutritional imbalances. In addition, for some Barth individuals, a true aversion to food and an exaggerated gag reflex further complicate nutrition. These problems often start in early infancy. The guidance of a nutritionist and a speech or occupational therapist may be needed to teach the individual how to chew and swallow correctly during infancy, the toddler years, or even later.

MEDICAL BACKGROUND: Because of their often substantially reduced muscle mass and decreased physical activity, individuals with Barth syndrome typically have lower than normal caloric requirements, a difference that must be appreciated when providing nutritional counseling. Furthermore, nutritionists and physicians unfamiliar with Barth syndrome often assume that a Barth boy's growth retardation is caused by inadequate nutrition. This common error often leads to overfeeding, which only further compromises health with vomiting, diarrhea, and, in some individuals, worsening mitochondrial function. Like any person with reduced muscle mass, a person with Barth syndrome has a reduced reserve of potassium and can rapidly become potassium-depleted with the vomiting or diarrhea of a gastrointestinal illness. Similarly, because of inadequate muscle mass to absorb and buffer changes in the blood potassium level, potassium in intravenous fluids can cause dangerously high blood levels of potassium (hyperkalemia), which has been the apparent cause of death by arrhythmia in several individuals with Barth syndrome. Consultation with your physician is recommended before administering IV fluids containing potassium. With rare exception, potassium should never be put in intravenous fluids without first determining the serum potassium level.

BREAST FEEDING: Infants with Barth syndrome may have difficulty nursing because of low muscle tone, frequent fatigue or rapid breathing associated with heart failure. In these situations, different methods can be used to assist with breastfeeding. First, try to breastfeed as soon after birth as possible. If breastfeeding doesn't work within the first few attempts, start pumping as often as the baby would breastfeed, every two to three hours. The baby can then be bottle fed with the breast milk, which does not require as much energy, especially if a "preemie nipple" is used. When bottle feeding after several attempts fails to provide adequate nutrition, a nasogastric tube or other feeding devices can be used to deliver the milk. Another commonly used aid, a Supplemental Nutrition System (SNS), can be requested from a lactation consultant. Another often useful feeding device is a bottle filled with breast milk and fitted with a thin tube that is taped to your breast or finger.

When attempting to breastfeed, be patient. It may take a few weeks for the baby to learn how to feed efficiently. A baby with poor muscle tone or diminished cardiac function may need extra physical support while nursing. The cuddling and skin-to-skin contact involved in breastfeeding is important, because it provides the stimulation the baby needs to better develop his feeding capabilities. Without this contact and stimulation, the baby may never become an avid and efficient breast feeder. Be assured that any amount of mother's milk from the breast, pumped and given by bottle, or provided by some other method (SNS, syringe, nasogastric tube, Habermann Feeder) will benefit the baby's health and development.

FEEDING ISSUES: It has been reported that some Barth individuals have sensory feeding problems, such as finicky eating, gagging, vomiting, and refusing food on sight without tasting it. Sensory cues provided by the mouth in response to food tell the brain what action, e.g., swallowing or chewing, should be taken. If a Barth infant is given bland food that provides inadequate sensation, the mouth may not know what to do with it and will initiate gagging to get rid of the food. The same problem can be caused by small pieces of food. Individuals with this type of gag reflex often prefer salty, crunchy, sour, or chewy tastes and consistencies. Foods that often are chosen to provide increased oral sensation include crackers, ketchup, blue cheese, pretzels, and pickles.

There are several different approaches to increasing the variety of foods eaten. One is to begin with what a child knows and likes and build on it. One should always seek professional help when feeding issues become a significant behavioral problem, take more time to address than is reasonable, or appear to be limiting growth. The earlier these problems are addressed the more effective will be the outcome.

- ***Taste Rejection:*** *(The idea that, as long as the taste is familiar, the food won't be immediately rejected).* For example, if a child likes cheese, try adding cheese to broccoli or beans, etc. If a particular sauce or juice is a favorite, add the sauce or juice to meats or veggies. Some individuals only like foods that have a lot of flavor. If a child refuses once, don't assume that he doesn't like it. Just try again the next meal or next day. Some kids refuse on sight based on the fact that it is a new food, NOT a flavor food.

- ***Sight Rejection:*** *(These approaches may help a child to look at new food as visually familiar and, thereby, encourage him to try something new).* Try the "food of the week" idea; pick one new food and present it with each meal without special comment. Place it within sight but don't make demands. Give the older child the option of spitting out the new food (it may help them to take a chance). Forcing a child to eat the food will usually be unsuccessful, because it reinforces negative associations with the food.

FOOD & EATING PREFERENCES: Many Barth individuals report enjoying foods that have a strong flavor which increase oral sensations, therefore enhancing "communication" between the mouth and brain regarding what motor sequence is needed (i.e. chewing, swallowing). Low muscle tone often means low sensory input (they travel together). Many individuals with low muscle tone don't like sweets because sweet foods produce more saliva, which tends to "water" down the taste of food, thereby interfering with ongoing communication between the mouth and brain.

It is not recommended to limit the Barth individual's food intake because, with few exceptions, the body knows how much is needed or what important nutrient is lacking. Often individuals with Barth syndrome consume small portions throughout the day rather than eating three large meals. In addition, because vitamin needs often are not decreased in proportion to the lower caloric requirement of individuals with Barth syndrome, it is advisable to give a daily multi-vitamin with minerals to prevent vitamin and other minor nutrient deficiencies.

Observation of Food Preferences - 2004 Family Survey - 26 Respondents				
• Dairy	*Cheese 38%*	*Milk 42% (8 - 32 oz daily)*		
• Eggs	*Want more eggs 31%*	*4+ per day 19%*		
• Other Foods	*Fatty Foods 35%*	*Salty Foods 91%*	*Sour Foods 32%*	*Spicy Foods 32%*
NOTE:	*88% of the respondents reported child to clearly crave particular foods.* *42% of the respondents report a consistent craving and 57% report heavy favoring of these foods.*			

Children with cardiomyopathy should avoid foods that increase the heart rate & induce a risk of arrhythmia. *Examples of Foods to Avoid Due to Cardiomyopathy*			
• Caffeine	• Chocolate	• Sodas	• Coffee

Individuals with neutropenia should avoid foods that are uncooked, undercooked, or contain large amounts of bacteria. *Examples of Foods to Avoid Due to Neutropenia*	
• Fresh fruits or vegetables	• Raw or rare cooked meats, fish or eggs
• Foods from street vendors	• Fresh herbs and seasonings (unless added during cooking)
• Dried fruits or nuts	• Unpasteurized dairy products (raw milk, naturally-aged cheese, yogurt)
• Raw Honey	• Beverages that are not pre-packaged (i.e. cans, bottles, cartons)
• Shellfish	• Cold cuts & processed meats(hot dogs, ham, bologna, salami, sausage)

FEEDING TUBES: To maintain weight, some Barth syndrome individuals require a feeding tube to provide appropriate nutrition and caloric intake. For example, some Barth infants have needed tube feedings shortly after birth due to poor weight gain.

- **Short-term tube feedings:** To help a child through a difficult feeding period, a naso-gastric tube or NG tube is inserted gently through the nose into the stomach and can be removed after a feeding or left in place for additional feedings. The NG tube is taped to the side of the face to keep it in the proper position. There are "long term" tubes that can remain in place for up to 30 days. This type of tube is preferred by therapists because it decreases the negative association of inserting and removing the tube. For children who have feeding problems, anything near the mouth that has negative effects or associations should be avoided or minimized to improve progression to oral feedings.

- **Long-term tube feedings:** To provide proper nutrition, a device called a gastrostomy button can be used. This is a short tube with a plastic cap that lies nearly flush against the skin of the stomach and is inserted surgically directly into the stomach. In some cases, a "j tube" placed in the jejunum (the upper small intestine) is recommended.

- **Drip or continuous feeding:** Drip-feeding uses a bag similar to an IV fluid bag with tubing that is attached to the g or j-tube or the NG tube. A measured amount of the prescribed formula is gently given. Often this is done throughout the night, with or without a pump while the individual is sleeping.

- **Bolus Feedings:** A prescribed amount of formula given over a short period of time, several times a day.

- **Amount:** The prescribed amount of formula for all methods of tube feeding is determined by the size of the child and the calories and volume needed. There are many different kinds of formula that can be used for tube feedings, and the physician and nutritionist together determine which one is right for each individual. Once on tube feedings, the child's weight will need to be monitored; frequent weight measurements are especially helpful during the early stages of tube feedings.