Preventing Protein Catabolism

by Debra Geary Hook, MPH, RD

Pediatric Dietician, Loma Linda University Children's Hospital

All children need protein for growth and health. This protein can come from two different sources; endogenous, from the body, or exogenous, from the diet. Proteins, regardless of their source, are made up of individual amino acids which are linked together in a specific order. Our bodies are constantly taking amino acids from one protein, breaking the links between the amino acids, rearranging them, and forming another protein. To illustrate, visualize lego blocks which can be snapped together to form an object and then unsnapped and rearranged to form another object.

The main purpose of dietary protein is to supply essential amino acids for protein synthesis and nitrogen to form components of cells. Essential amino acids can only be obtained from the diet, since the human body lacks a way of manufacturing them. However, only a small part of the amino acids used in protein metabolism is supplied through the diet. The majority of the amino acids used in protein synthesis comes from the breakdown of protein within the body. Since human beings cannot store the excess nitrogen, the unneeded amino acids containing nitrogen are converted into substances which can be used for energy. For this to take place, the nitrogen is removed and ammonia is formed. Ammonia is toxic, especially to the brain. To get rid of this toxic waste product, the body normally converts ammonia into urea, which is non-toxic. In urea cycle disorders, this process is impaired and ammonia is allowed to accumulate in the body, resulting in high ammonia levels. To prevent these high ammonia levels, a combination of medication and diet manipulation are prescribed. Most children will follow a low protein diet.

It is true that limiting protein intake reduces the production of waste nitrogen and serves to keep ammonia levels within normal range; however, if the amount of protein is too restrictive, the body will break down its own protein stores and in doing so will create nitrogen waste. Because the body uses protein from two different sources, endogenous and exogenous, the dietary management of of urea cycle disorders becomes complicated. One cannot merely reduce the amount of protein in the diet and thus prevent high ammonia levels. The body will respond to this low amount of dietary amino acids by breaking down its own lean muscle mass. The result will be high levels of ammonia in the blood resulting in brain damage.

To prevent this from happening, the amount of protein in the child's diet must be considered in relationship to his level of activity, periods of growth and development, and individual tolerance to dietary protein. What is needed is the minimum amount of protein to promote growth, while preventing catabolism of body protein. The problem is that the minimum protein requirements of infants and children are not precisely known. Guidelines from the World Health Organization provide safe levels of protein intake for children. Based on these guidelines, safe protein intake levels have been suggested. It must be stressed that these are *estimated levels* and should be used by health care professionals in determining a dietary program for each individual patient.

Age	Protein	Energy(calories/kg)
<3 months	1.2-1.8	130-145
3-6 months	1.0-1.4	125-145
6-9 months	1.0-1.3	120-125
9-12 months	0.9-1.2	115-135
1-4 years	0.7-1.0	110-120
4-7 years	0.6-0.8	110-120
7-11 years	0.5-0.7	80-90
11-18 years	0.4-0.6	55-65

Source: Elsas, L.J., Acosta, P.B. Nutrition Support of Inherited Metabolic Disorders. In: Shils, ME, Young, VR, eds. Modern Nutrition in Health and Disease 7 ed. Philadelphia: Les & Febiger, 1988.

Many children are supplemented with essential amino acids to further decease the requirement of dietary protein. These essential amino acids are provided in the form of medical foods and roughly half of the daily protein intake comes from this medical food.

To further spare protein from being broken down and used as energy, it is necessary to provide generous calories. For this reason, the recommended daily calories are *higher* than for individuals without urea cycle disorders. Besides energy, the higher carbohydrate, sugar intake can be used to supply parts to build nonessential amino acids. Given an adequate calorie intake from carbohydrate and fat, dietary amino acids can be spared to provide the nitrogen necessary for protein synthesis.

Dietary management of a urea cycle is difficult and time consuming although not impossible. Through constant monitoring, it is possible to find the delicate balance that exists while supplying enough protein for growth, while not over-restricting. The best way to determine if you are on the right track is to look at your child's growth, ammonia levels, serum amino acid levels, and protein status.

(Reprinted from NUCDF Newsletter Spring 1997)