

Reimbursement for Medical Foods for Inborn Errors of Metabolism

Committee on Nutrition

Inborn errors of amino acid metabolism such as phenylketonuria, maternal phenylketonuria, maple syrup urine disease, homocystinuria, methylmalonic acidemia, propionic acidemia, isovaleric acidemia and other disorders of leucine metabolism, glutaric acidemia type I and tyrosinemia types I and II, and urea cycle disorders are rare diseases that are treatable by diet. Treatment might include the restriction of one or more amino acids, the restriction of total nitrogen, or the supplementation of specific substances. Untreated, these diseases culminate in severe mental retardation or death.

Once diagnosis is confirmed, treatment of amino acid and urea cycle disorders must be carefully monitored by a physician with expertise in metabolic diseases. Special medical foods, commercially available, are indispensable for the active, ongoing treatment of diagnosed amino acid and urea cycle disorders. Special medical foods would, if used as the sole dietary source, represent a hazard to affected and healthy children. US Public Law (Publ L) 100-290 defines the term medical food as "... a food which is formulated to be consumed or administered enterally under the supervision of a physician and which is intended for the specific dietary management of a disease or condition for which distinctive nutritional requirements, based on recognized scientific principles, are established by medical evaluation."¹

After passage of Publ L 100-290, many states provided funding for these products through Medicaid, and most states offered assistance through Crippled Children's and Women, Infant, and Children's programs. Some states now have laws mandating private insurance coverage for special medical foods.

It is the position of the American Academy of Pediatrics that special medical foods that are used in the

treatment of amino acid and urea cycle disorders are medical expenses that should be reimbursed.

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REFERENCE

1. US Congress, 100th Congress. Orphan Drug Amendments of 1988 (Act to Amend the Federal Food, Drug, Cosmetic Act to Revise the Provisions Respecting Orphan Drugs, for Other Purposes). Pub L No. 100-290. Washington, DC: US Government Printing Office.

The recommendations in this policy statement do not indicate an exclusive course of treatment or procedure to be followed. Variations, taking into account individual circumstances, may be appropriate.

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