

# 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."<sup>1</sup>

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.

COMMITTEE ON NUTRITION, 1992 to 1993  
Ronald E. Kleinman, MD, Chairperson  
Susan S. Baker, MD  
Edward F. Bell, MD  
Terry F. Hatch, MD  
William J. Klish, MD  
Rudolph L. Leibel, MD  
John N. Udall, MD

#### LIAISON REPRESENTATIVES

Margaret Cheney, PhD, Bureau of  
Nutritional Sciences, Canada  
Joginder Chopra, MD, Food and Drug  
Administration  
Patricia N. Daniels, US Department of  
Agriculture  
Suzanne S. Harris, PhD, International Life  
Sciences Institute  
Van S. Hubbard, MD, National Institute of  
Diabetes and Digestive and Kidney  
Diseases  
Ephraim Levin, MD, National Institute of  
Child Health and Human Development  
Ann Prendergast, RD, MPH, Bureau of  
Health Care Delivery and Assistance  
Micheline Ste-Marie, MD, Canadian  
Paediatric Society  
Alice E. Smith, MS, RD, American Dietetic  
Association  
Ray Yip, MD, MPH, Centers for Disease  
Control and Prevention  
AAP SECTION LIAISON  
Ronald M. Lauer, MD, Section on  
Cardiology

#### 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.

PEDIATRICS (ISSN 0031 4005). Copyright © 1994 by the American Academy of Pediatrics.

**Reimbursement for Medical Foods for Inborn Errors of Metabolism**  
Committee on Nutrition  
*Pediatrics* 1994;93;860

**Updated Information & Services**

including high resolution figures, can be found at:  
<http://pediatrics.aappublications.org/content/93/5/860>

**Permissions & Licensing**

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:  
<http://www.aappublications.org/site/misc/Permissions.xhtml>

**Reprints**

Information about ordering reprints can be found online:  
<http://www.aappublications.org/site/misc/reprints.xhtml>

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™



# PEDIATRICS®

OFFICIAL JOURNAL OF THE AMERICAN ACADEMY OF PEDIATRICS

## **Reimbursement for Medical Foods for Inborn Errors of Metabolism**

Committee on Nutrition

*Pediatrics* 1994;93;860

The online version of this article, along with updated information and services, is located on the World Wide Web at:

<http://pediatrics.aappublications.org/content/93/5/860>

Pediatrics is the official journal of the American Academy of Pediatrics. A monthly publication, it has been published continuously since 1948. Pediatrics is owned, published, and trademarked by the American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, Illinois, 60007. Copyright © 1994 by the American Academy of Pediatrics. All rights reserved. Print ISSN: 1073-0397.

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™

