

AMERICAN ACADEMY OF PEDIATRICS

POLICY STATEMENT

Organizational Principles to Guide and Define the Child Health Care System and/or Improve the Health of All Children

Committee on Nutrition

Reimbursement for Foods for Special Dietary Use

ABSTRACT. Foods for special dietary use are recommended by physicians for chronic diseases or conditions of childhood, including inherited metabolic diseases. Although many states have created legislation requiring reimbursement for foods for special dietary use, legislation is now needed to mandate consistent coverage and reimbursement for foods for special dietary use and related support services with accepted medical benefit for children with designated medical conditions.

ABBREVIATION. AAP, American Academy of Pediatrics.

BACKGROUND

Special foods are recommended by physicians to foster normal growth and development in some children and to prevent serious disability and even death in others. Many of these special foods are technically specialized formulas for which there may be a relatively small market, which makes them more expensive than standard formula. Since publication of the American Academy of Pediatrics (AAP) policy statement in 1994,¹ many states have created legislation for mandating reimbursement for foods for children with inborn errors of metabolism. Third-party payment for foods for special dietary use is inconsistent, however, and state statutes regarding reimbursement vary widely. Some states require coverage only for inherited metabolic diseases, such as phenylketonuria, and others include a range of metabolic conditions. Legislation is now needed to mandate consistent coverage and reimbursement for all aspects of foods for special dietary use and related supplies and services for children with designated medical conditions.

There is a great need for pediatricians and the AAP to take a leadership role in this area of child health affecting infants, children, and adolescents by building on the legislative advancements that have been made as a result of the 1994 policy.¹ A model bill (Pediatric Medical Nutrition Support Act) for proposed legislation is available on the AAP Web site (<http://www.aap.org/policy/m972.html>).

DEFINITION OF FOODS FOR SPECIAL DIETARY USE

The US Food and Drug Administration, in the Code of Federal Regulations,² defines special dietary use of foods as the following:

- a. Uses for supplying particular dietary needs that exist by reason of a physical, physiologic, pathologic, or other condition, including but not limited to the conditions of diseases, convalescence, pregnancy, lactation, allergic hypersensitivity to food, [and being] underweight and overweight;
- b. Uses for supplying particular dietary needs which exist by reason of age, including but not limited to the ages of infancy and childhood;
- c. Uses for supplementing or fortifying the ordinary or usual diet with any vitamin, mineral, or other dietary property. Any such particular use of a food is a special dietary use, regardless of whether such food also purports to be or is represented for general use.

MEDICAL CONDITIONS COVERED

Diseases covered would include all chronic diseases or conditions of childhood requiring special dietary intervention, including inherited metabolic diseases (Table 1).

Many childhood chronic diseases (eg, inflammatory bowel disease, cystic fibrosis, celiac disease, cancer, congenital heart disease, renal failure, hepatic diseases) are associated with increased nutritional requirements and metabolic demands or with decreased nutrient intakes, limitations of digestion and absorption, and/or increased nutrient losses. An optimal state of nutrition is especially important in these children, and poor nutrition is associated with increased risk of infections, inadequate growth, and poor response to treatment modalities, including surgery.^{3,4} The goals of nutritional support for children with these chronic illnesses include normal growth and development, promotion of catch-up growth, and improved clinical outcome. Specialized nutritional support is often required, including enteral tube feedings or parenteral nutrition.

Special nutritional supplements and feeding approaches often are required for infants and children with devastating neurologic diseases or impairments, such as severe cerebral palsy, and progressive neurodevelopmental diseases.⁵

TABLE 1. Medical Conditions for Which Foods for Special Dietary Use May Be Reimbursed*

Conditions requiring specific dietary components
Chronic pulmonary insufficiency
Congenital or acquired chronic cardiac insufficiency
Movement disorder (neuromuscular)
Catch-up growth in children attributable to undernutrition
Rett syndrome
Inflammatory bowel disease
Conditions requiring the alteration of specific dietary components
Short bowel syndrome (long-segment Hirschsprung disease, small intestinal atresia, necrotizing enterocolitis, volvulus, gastroschisis)
Intestinal pseudo-obstruction syndromes
Nonspecific malabsorption syndromes (eg, postviral gastroenteropathy, small intestinal bacterial overgrowth)
Intestinal lymphangiectasia
Abetalipoproteinemia
Microvillus inclusion disease
Eosinophilic gastroenteropathy
Partial villus atrophy attributable to food protein sensitivity
Intestinal transplantation
Exocrine pancreatic insufficiency (eg, cystic fibrosis, Schwachman syndrome, Johanson-Blizzard syndrome, Pearson syndrome, pancreatic hypoplasia, congenital trypsinogen deficiency)
Chronic liver disease with cholestasis
Impaired bile acid synthesis
Bile duct atresia
Interrupted enterohepatic circulation (eg, ileal resection, congenital malabsorption of bile acids, blind loop syndrome)
Enterokinase deficiency
Immunodeficiency states (eg, villus atrophy associated with immunodeficiency conditions, autoimmune enteropathy, or enterocolitis associated with immune deficiency)
Chylolothorax
Conditions impairing adequate oral intake
Cystic fibrosis
Acquired immunodeficiency syndrome
Transplant patients: bone marrow, liver, kidney, heart, lung, or multiple organs
Cerebral palsy
Aspiration syndrome
Oral dysfunction
Pharyngeal dysfunction
Esophageal dysfunction
Neurologic disorders
Oral feeding aversion (resulting from chronic use of enteral tube feedings or parenteral nutrition)
Pancreatitis
Treatment of childhood cancer
Chronic renal failure, hemodialysis, and peritoneal dialysis
Anatomic, congenital, or postsurgical defects precluding oral nutritional intake (eg, craniofacial defects)

Metabolic diseases include inborn errors of amino acid metabolism, such as phenylketonuria, maternal phenylketonuria, maple syrup urine disease, homocystinuria, methylmalonicacidemia, propionicacidemia, isovalericacidemia, and other disorders of leucine metabolism; glutaricaciduria type I and tyrosinemia types I and II; and urea cycle disorders. These are all disorders treatable by dietary modifications, which can prevent complications like severe mental retardation and death.⁶ Manipulations of precursors and limitation of substrates in the diet form a major portion of available therapies. In the case of phenylketonuria, a special National Institutes of Health consensus panel recently recommended uniform policies to remove individual and family finan-

TABLE 1. Continued.

Metabolic disorders
Disorders of carbohydrate metabolism
Glycogen storage disease
Glucose transport protein deficiency
Galactosemia
Hereditary fructose intolerance
Pyruvate dehydrogenase complex deficiency
Phosphoenolpyruvate carboxykinase deficiency
Disorders of lipid metabolism
Fasting chylomicronemia
Mitochondrial fatty acid oxidation defects (eg, α -ketoadipicaciduria, methylmalonicaciduria, long-chain acyl-CoA dehydrogenase deficiency, and medium-chain acyl-CoA dehydrogenase deficiency)
β -ketothiolase deficiency
Succinyl-CoA ketoacid transferase
Other organic acidemias
Disorders of vitamin metabolism
Biotinidase deficiency
Holocarboxylase synthetase deficiency
Methylmalonicacidemia
Methylcrotonyl-CoA carboxylase deficiency
Thiamine-responsive maple syrup urine disease
Pyridoxine responsive seizures
Disorders of mineral metabolism
Hypercalcemia
Williams syndrome
Disorders of amino acid or nitrogen metabolism
Phenylketonuria
Cystinosis
Homocystinuria
Glutaric acidemia (types I and II)
Disorders of branched-chain amino acid metabolism (eg, disorders of leucine metabolism, isovaleric acidemia, maple syrup urine disease, 3-hydroxy-3-methylglutaricaciduria, 3-methylcrotonylglycinemia, 3-methylglutaconicaciduria)
Tyrosinemia (types I and II)
Lysinuric protein intolerance
Urea cycle defects (eg, ornithine transcarbamylase deficiency, hyperornithine-hyperammonemia-homocitrullinuria syndrome, argininemia, argininosuccinicaciduria, carbamyl phosphate synthetase deficiency, citrullinemia)
Methylmalonicacidemia
Propionicacidemia
Gyrate atrophy of the choroid and retina
Adrenoleukodystrophy
Miscellaneous mitochondrial disorders and mitochondrial electron transport defects

* List is not intended to be all inclusive.
CoA indicates coenzyme A.

cial barriers to acquiring modified low-protein foods and outlined support services required to maintain appropriate phenylalanine concentrations.⁷ Children with other metabolic disorders, including those of carbohydrate metabolism, lipid metabolism, vitamin or cofactor metabolism, or mineral metabolism, may also benefit from dietary interventions.

DEFINITION OF REIMBURSABLE EXPENSES

Any health insurance policy that is delivered, issued for delivery, renewed, extended, or modified in a particular state by any health care insurer and that provides coverage for a child should optimally provide coverage of foods for special dietary use of accepted medical benefit. This is meant to cover nutritional support costs over and above usual foods. In addition to the cost of the foods, all medical equipment and medical supplies necessary for the delivery of foods for special dietary use should be covered.

This includes but is not limited to administration tubing, bags, and pumps. Costs of management by health care professionals as necessary to administer or monitor the safe administration of foods for special dietary use should also be covered.

RECOMMENDATIONS

1. All foods for special dietary use with accepted benefit for treatment of a medical condition should be reimbursed as a medical expense, provided the costs are over and above usual foods. Individual and family financial barriers to obtaining these foods should be removed.
2. All states should enact legislation that would require health insurance policy providers to reimburse all foods for special dietary use with accepted medical benefit recommended by a physician to prevent death and serious disability or to foster normal growth and development.
3. All expenses for medical equipment and medical supplies necessary for the delivery of foods for special dietary use should be reimbursed.
4. Reimbursement for foods for special dietary use should be mandatory for the following:
 - a. Any medical condition for which specific dietary components or the restriction of specific dietary components is necessary to treat a physical, physiologic, or pathologic condition resulting in inadequate nutrition.
 - b. An inherited metabolic disorder, including but not limited to disorders of carbohydrate metabolism, lipid metabolism, vitamin metabolism, mineral metabolism, or amino acid and nitrogen metabolism.
 - c. A condition resulting in impairment of oral intake that affects normal development and growth.

COMMITTEE ON NUTRITION, 2001–2002
Nancy F. Krebs, MD, Chairperson
Robert D. Baker, Jr, MD, PhD
*Frank R. Greer, MD
Melvin B. Heyman, MD
Tom Jaksic, MD, PhD
Fima Lifshitz, MD

LIAISONS

Sue A. Anderson, PhD, RD
US Food and Drug Administration
Donna Blum-Kemelor, MS, RD
US Department of Agriculture
Margaret P. Boland, MD
Canadian Paediatric Society
William Dietz, MD, PhD
Centers for Disease Control and
Prevention
Van S. Hubbard, MD, PhD
National Institute of Diabetes and
Digestive and Kidney Diseases
Elizabeth Yetley, PhD
US Food and Drug Administration

STAFF

Pamela Kanda, MPH

*Lead author

REFERENCES

1. American Academy of Pediatrics, Committee on Nutrition. Reimbursement for medical foods for inborn errors of metabolism. *Pediatrics*. 1994;93:860
2. Code of Federal Regulations, Title 21. Food and Drugs, Vol 2, Part 105. Foods for Special Dietary Use. Sec 105.3. Definitions and interpretations. Revised April 1, 1999. Washington, DC: US Government Printing Office
3. Heird WC, Cooper A. Nutritional management of infants and children with specific diseases and/or conditions. In: Shils ME, Olson JA, Shike M, Ross AC, eds. *Modern Nutrition in Health and Disease*. 9th ed. Baltimore, MD: Williams & Wilkins; 1999:1081–1090
4. American Academy of Pediatrics, Committee on Nutrition. Nutritional management of children with a chronic illness. In: Kleinman RE, ed. *Pediatric Nutrition Handbook*. 4th ed. Elk Grove Village, IL: American Academy of Pediatrics; 1998:485–494
5. Cloud HH. Developmental disabilities. In: Queen PM, Land CE, eds. *Handbook of Pediatric Nutrition*. Gaithersburg, MD: ASPEN Publishers Inc; 1993:400–421
6. Elsas LJ, Acosta PB. Nutritional support of inherited metabolic disease. In: Shils ME, Olson JA, Shike M, Ross AC, eds. *Modern Nutrition in Health and Disease*. 9th ed. Baltimore, MD: Williams & Wilkins; 1999: 1003–1056
7. National Institutes of Health, Consensus Development Panel. Phenylketonuria: screening and management. *Pediatrics*. 2001;108: 972–982

All policy statements from the American Academy of Pediatrics automatically expire 5 years after publication unless reaffirmed, revised, or retired at or before that time.