



**BlueCross BlueShield  
of Vermont**

*Independent Licensees of the Blue Cross and Blue Shield Association.*



## **Corporate Medical Policy**

### **Medical Nutrition for Inherited Metabolic Disease**

**File name:** Medical Nutrition for Inherited Metabolic Disease

**Origination:** 10/98

**Last Review:** 10/07

**Next Review:** 10/08

**Effective Date:** 10/25/07

#### **Description of Procedure or Service**

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Medical nutrition therapy is an important part of treatment of many inherited metabolic diseases. This policy explains the medical conditions and the nutritional formulae and medical food supplements, which will be considered for reimbursement for medically necessary treatment of an inherited metabolic disease. For the purposes of this policy the following definitions apply:

*“Inherited metabolic disease,”* means a disease caused by an inherited abnormality of body chemistry for which the state screens newborn infants.

*“Low protein modified food product”* means a food product that is specifically formulated to have less than one gram of protein per serving and is intended to be used under the direction of a physician for the dietary treatment of a metabolic disease.

*Medical food* means an amino acid modified preparation that is intended to be used under the direction of a physician for the dietary treatment of an inherited metabolic disease.

#### **Policy**

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Benefits are subject to the terms, limitations and conditions of the subscriber contract.

Prior approval is required for all lines of business.

Coverage for *low protein modified food products* and medical food prescribed for medically necessary treatment of an inherited metabolic disease will be capped at

\$2,500.00 for medical food items for each calendar year for any insured individual.  
Coverage for these formulas will not be subject to any benefit maximums

Benefits are available for members with the following inherited metabolic diseases: Biotinidase Deficiency, Congenital Hypothyroidism, Galactosemia, Homocystinuria, Maple Syrup Urine Disease, Phenylketonuria (PKU), and Sickle Cell Disease.

### **Benefits Application**

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This benefit is not provided under the pharmacy benefit; therefore medical co-insurance and deductibles (if applicable) apply.

### **When service or procedure is covered**

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Benefits for medical nutrition for inherited metabolic disease are eligible for:

- Formula supplements for the diagnosis of inherited metabolic disease
- Low protein modified food product and medical food for the diagnosis of inherited metabolic disease

### **When service or procedure is not covered**

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- When the above conditions are not met.
- When prior approval is not obtained.
- Nutritional formulae or supplements, except for up to \$2,500 per year for “medical foods” prescribed for the Medically Necessary treatment of an inherited metabolic disease or formulae and supplements administered through a feeding tube.

### **Billing Guidelines**

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See Attachment I and II.

The Children with Special Health Care Needs Clinic (CSHNC) will purchase the formula for members with these diagnoses. CSHNC will submit a HCFA 1500 and invoice the Plan for reimbursement quarterly.

Bill HCPCS code B9998, NOC for enteral supplies.

Bill HCPCS code S9434, Modified solid food supplements for inborn errors of metabolism

Bill HCPCS code S9435, Medical foods for inborn errors of metabolism.

Members will purchase their own foods through one of three vendors (Attachment II) and will then submit a subscriber claim form with a copy of the invoice attached for reimbursement to the Plan.

**Policy Implementation/Update information**

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9/07 Annual review, minor wording changes to match current certificate language.

Reviewed by the CAC 11/07.

8/06 modifier removed from HCPCS code

3/05 updated, referral requirement removed for TVHP. 5/03 – removed case management requirement, Nutritional Formulae and Medical Food Supplements 5/1/92, Medical Nutrition Therapy For Inherited Metabolic Diseases, 8/00

**References**

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8 V.S.A § 4089(e)

**Approved by BCBSVT and TVHP Medical Directors**

Stephen E. Perkins, M.D. (BCBSVT) and Frank Provato, M.D. (TVHP)    Date

Approved:

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## Attachment I- Infant Metabolic Disorders and Formulae Covered

Included in the following matrix are the disorders for which formula will be covered.

Disorder	Diagnosis	Substance Tested For	NDC/Product Name
Biotinidase Deficiency		Biotinidase	No Specific Formula Available at this time
Congenital Hypothyroidism	243.0	Thyroxine (T4), thyroid stimulating hormone (TSH)	No Specific Formula Available at this time
Galactosemia	271.1	Galactose, galactose-1-phosphate uridyl transferase	00087-0616-14 Lactofree Powder, 14 oz. 00087-0614-13 Lactofree Concentrated Liquid, 13 oz. 00087-0615-32 Lactofree Ready-to-Use Liquid, 32 oz 00087-0308-01 Prosobee Concentrated Liquid, 13 oz 00087-0309-01 Prosobee Ready-to-Use Liquid, 32 oz. 00087-3101-21 Prosobee Powder, 14 oz. 00087-0498-01 Nutramigen Concentrated Liq, 13 oz. 00087-0499-01 Nutramigen Ready-to-Use Liq 32 oz. 00087-0338-01 Nutramigen Powder, 16 oz. 00087-0367-43 Pregestimil Powder, 16 oz. 00087-0333-43 Pregestimil 20 Liquids, 3 oz. 00087-0379-43 Pregestimil 24 Liquids, 3 oz. 00087-1410-41 Next Step Soy Concentrated Liq, 13oz. 00087-1409-41 Next Step Soy Powder, 12 oz.  70074-4211-00 Isomil 70074-4017-30 Isomil 70074-2010-70 Isomil 70074-6023-00 Isomil 70074-5039-30 Isomil
Disturbances of sulphur-bearing amino acid metabolism	270.4	Methionine, Homocystinuria	00087-4065-41 HOM-1 Powder, 500 grams 00087-4066-41 HOM-2 Powder, 500 grams 70074-5111-60 Hominex
Disturbances of branch-chain amino-acid metabolism	270.3	Maple Syrup Urine Disease, Leucine	00087-0418-42 MSUD Diet Powder, 16 oz 00087-4060-41 MSUD-1 Powder, 500 grams 00087-4061-41 MSUD-2 Powder, 500 grams 70074-5111-20 Ketonex
Phenylketonuria (PKU)	270.1	Hyperphenylalaninemia	00087-0340-42 Lofenalac Powder, 16 oz 00087-0368-42 Phenyl-Free Powder, 16 oz 00087-4055-41 PKU-1 Powder, 500 grams 00087-4056-41 PKU-2 Powder, 500 grams 00087-4057-41 PKU-3 Powder, 500 grams 70074-5112-00 Phenex
Sickle Cell Anemia	282.61 (w/o crisis) 282.62 (w/crisis)	Hemoglobin S (HbS)	70074-4037-30 PediaSure

## **ATTACHMENT II – FOOD VENDORS**

### Metabolic Nutrition Therapy for Inherited Metabolic Diseases

Dietary Specialties, Inc.  
PO Box 227  
Rochester, NY 14601  
Tel: 1-800-544-0099

Med-Diet, Inc.  
3050 Ranchview Lane  
Plymouth, MN 55447  
Tel: 1-800-633-3438

Children With Special Health Needs  
State of Vermont, Department of Health  
P.O. Box 70  
Burlington, VT 05402  
802-863-7338