MEDICAL NUTRITION FOR INHERITED METABOLIC DISEASE
Corporate Medical Policy

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Document Precedence

BCBSVT Medical Policies are developed to provide guidance for members and providers regarding coverage in accordance with all terms, conditions and limitations of the subscriber contract. Benefit determinations are based in all cases on the applicable contract language. To the extent that there may be any conflict between Medical Policy and contract language, the contract language takes precedence.

Medical Policy

Description

Definitions:

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 1 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.

An inherited metabolic disease is caused by a genetic defect which leads to life threatening abnormalities in body chemistry. Certain metabolic enzymes may be absent in individuals, and as a result, products of digestion which are normally further metabolized may accumulate in toxic amounts. Examples of these disorders include, but are not limited to phenylketonuria, hyperphenylalaninemia, maple syrup urine disease (ketoadiduria), histidinemia, homocystinuria, organic acidemias, tyrosinemia, and urea cycle disorders (citrullinemia, arginosuccinic aciduria). The rationale behind the use of "medical" foods is that the elements normally found in food which lead to the accumulation of these products of digestion are eliminated, so the toxic accumulation of by-products does not occur.

Medical foods are intended for the dietary treatment of a disease or condition for which nutritional requirements are established as medically necessary, and are formulated to be consumed or administered under the direction of a physician. Medical foods include:

A. Specially manufactured infant feeding formulas that have been developed to treat inborn errors of metabolism. Certain formulas eliminate the amino acid that cannot be
metabolized. Oral nutrition formula for supplementation or dietary replacement is considered medically necessary for the treatment of inborn errors of metabolism, when the formula is required to prevent significant illness resulting from a byproduct in the metabolic pathway or amino acid accumulation, or to restore an essential nutrient that is lacking as a result of an inborn error of metabolism. See BCBSVT Medical Policy on Enteral Nutrition for more information.

B. Other medical foods for use by older children and adults are products that resemble natural food, but which have been either chemically synthesized or processed from natural food. These products are generally not available in supermarkets or grocery stores, but are obtained directly by mail order from the manufacturer. *Low protein modified food products* include special breads, pastas, baked goods, flour mixes, cheeses and broths which have been formulated to have less than one gram of protein per serving and do not include natural foods that are naturally low in protein.

Policy

Medical foods are considered **medically necessary** for the dietary treatment of inherited metabolic disease when prescribed by a physician and **administered under the direction of a physician**.

Administrative and Contractual Guidance

**Benefit Determination Guidance**

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

Prior approval is required for all lines of business.

For New England Health Plan (NEHP) members an approved referral authorization is required.

FEP members may have benefits that differ from those described in this policy. Please consult the FEP Service Plan brochure.

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 plan year for any insured individual. This benefit limit does not apply to medical foods and formulae administered through a feeding tube. Please see BCBSVT medical policy on Enteral Nutrition for additional information.

As of the effective date of this medical policy, the State of Vermont requires newborn screenings for the following metabolic disorders. Therefore medical foods and formulas prescribed for these disorders must be covered, subject to the benefit limitations described in this medical policy. This list is subject to change.

**NOTE:** *Not all listed conditions require medical foods or formulas.*

- Argininosuccinic acidemia (ASA)\(^1\) (270.6, E72.21)\(^2\),
- Beta-ketothiolase deficiency (BKT) (270.3; E71.19),
- *Biotinidase Deficiency (277.6, D81.810),*
- *Carnitine uptake defect (CUD) (277.81, E71.41),*
**Condition Name and Abbreviation** — curated by the NLM and selected from among the names used by the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC), National Newborn Screening Information System (NNSIS), the American College of Medical Genetics (ACMG), the HHS Office of the National Coordinator for Health Information Technology (ONC)/American Health Information Community (AHIC) Personalized Health Care Work Group, and input from the newborn screening community.

1. Citrullinemia (CIT) (270.6, E72.23),
2. *Congenital adrenal hyperplasia (CAH) (255.2,E25.0),
3. *Congenital Hypothyroidism (CH) (243,E03.1),
4. *Cystic fibrosis (CF) (277.00,E84.9),
5. Galactosemia (GALT) (271.1,E74.21),
6. 3-OH 3-CH3 glutaric aciduria (HMG) (270.7,E71.118),
7. Glutaric acidemia type I (GA I) (270.7,E72.3),
8. *Hb S/Beta-thalassemia (Hb S/BTh) (282.41,D57.40),
9. *Hb S/C disease (Hb S/C) (282.63,D57.20),
10. Homocystinuria (HCY) (270.4,E72.11),
11. Isovaleric acidemia (IVA) (270.3, E71.110),
12. *Long-chain L-3-OH acyl-CoA dehydrogenase deficiency (LCHAD) (277.85,E71.318),
13. Maple Syrup Urine Disease (MSUD) (270.3,E71.0),
14. *Medium-chain acyl-CoA dehydrogenase deficiency (MCAD) (277.85,E71.311),
15. 3-Methylcrotonyl-CoA carboxylase deficiency (3MCC) (270.3,E71.19),
16. Methylmalonic acidemia (Cbl A, B) (270.3,E71.120),
17. Methylmalonic acidemia (mutase deficiency) (MUT) (270.3,E71.120),
18. Multiple carboxylase deficiency (MCD) (270.3,D81.818),
19. Phenylketonuria (PKU)(270.1,E70.0),
20. Propionic acidemia (PROP)(270.3,E71.121),
21. *Sickle cell anemia (Hb SS disease) (SS) (282.60,D57.1),
22. *Trifunctional protein deficiency (TFP) (277.85,E71.318),
23. Tyrosinemia type I (TYR I) (270.2,E70.21),
24. *Very long-chain acyl-CoA dehydrogenase deficiency (VLCAD) (277.85,E71.310)

Medical foods generally are not available in grocery stores, supermarkets, pharmacies, or health food stores. They generally must be purchased directly from the manufacturer and shipped directly to the patient. Examples of some manufacturers/distributors of medical foods include:

1. Mead Johnson Nutrition
2. Abbott Nutrition
3. Vitaflo (Nestle Nutrition)
4. Applied Nutrition
5. Nutricia, N.A.
6. CamBrooke Foods, LLC.
7. Med-Diet, Inc.
9. Dietary Specialties, Inc.
Benefits are not provided for:

- Conventional food items that are naturally low in protein, even if consumed by patients with an inherited metabolic disease. These are considered grocery items.
- Food supplements (e.g., Ensure, Sustacal) are not considered medical foods as they are generally not intended for the treatment of inherited metabolic disorders.
- Convenience items such as recipe books.
- Shipping and handling fees or costs.

This benefit is not provided under the pharmacy benefit; therefore medical benefit cost sharing (e.g., deductible, co-insurance, or other co-payments) may apply.

Eligible Providers

DME vendors

Related Policies

Enteral Nutrition
Total Parenteral Nutrition

Policy Implementation/Update information

08/2000 new policy supersedes all prior policies concerning this benefit
05/2003 – removed case management requirement,
03/2005 updated, referral requirement removed for TVHP.
08/2006 modifier removed from HCPCS code
09/2007 annual review; minor wording changes to match current certificate language.
09/2008 annual review; benefit clarification made.
05/2009 annual review; no substantive change; approved by CAC
10/2011 Updated and transferred to new format. Vermont neonatal screening list updated. Official abbreviations and ICD-9 and ICD-10 codes for each condition added. Obsolete language referring to sources of formula removed. Added scientific references.
Medical/Clinical Coder reviewed 10/24/11 SAF

Scientific Background and Reference Resources

8 V.S.A § 4089(e)

3. The following website is useful to determine which products are assigned to a specific HCPCS code: https://www.dmepdac.com/

Approved by BCBSVT Medical Directors         Date Approved

Antonietta Sculimbene MD
Chair, Medical Policy Committee
## Attachment I

<table>
<thead>
<tr>
<th>Codes</th>
<th>Number</th>
<th>Description</th>
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<tr>
<td>HCPCS</td>
<td>S9434</td>
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<td>S9435</td>
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<td>B9998</td>
<td>NOC Enteral supplies</td>
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<td>ICD-9 Diagnosis</td>
<td>270.1</td>
<td>Phenylketonuria (PKU)</td>
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<tr>
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<td>270.2</td>
<td>Other disturbances of aromatic amino-acid metabolism (Tyrosinemia type I)</td>
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<tr>
<td></td>
<td>270.3</td>
<td>Disturbances of branched-chain amino-acid metabolism (Maple syrup urine disease, beta-ketothiolase deficiency, isovaleric acid deficiency, methylmalonic acidemia, multiple carboxylase deficiency, propionic acidemia, 3-Methylcrotonyl-CoA carboxylase deficiency)</td>
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<td>270.4</td>
<td>Disturbances of sulphur-bearing amino-acid metabolism (Homocystinuria)</td>
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<td>270.5</td>
<td>Disturbances of histidine metabolism</td>
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<td>270.6</td>
<td>Disorders of urea cycle metabolism (Argininosuccinic acidemia, citrullinemia)</td>
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<td>Other disturbances of straight chain amino-acid metabolism (3-OH 3-CH3 glutaric aciduria, Glutaric acidemia type I)</td>
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<td>Type of Service</td>
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## Attachment II

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<thead>
<tr>
<th>Metabolic Disorder</th>
<th>Medical Food</th>
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<tr>
<td>Glutaric acidemia (GLA)</td>
<td>XLys, XTrp Analog</td>
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<td>XLys, XTrp Maxamaid</td>
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<td>XLys, XTrp Maxamum</td>
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<td>Glutarex-1</td>
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<td>Glutarex-2</td>
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<td>GA</td>
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<td>Condition/Syndrome</td>
<td>Products</td>
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| Methionemia (MTH)  | Methionaid  
|                    | XMet Analog  
|                    | XMet Maxamaid  
|                    | XMet Maxamum  
|                    | Hominex-1  
|                    | Hominex-2  
|                    | HCU Gel  
|                    | HCU Express  |
| Isovalericacidemia (IVA) | XLeu Analog  
|                     | XLeu Maxamaid  
|                     | XLeu Maxamum  
|                     | I-Valex-1  
|                     | I-Valex-2  
|                     | LMD  |
| Maple Syrup Urine Disease (MSUD) (Ketoaciduria) | BCAD1  
|                     | BCAD 2  
|                     | MSUD2  
|                     | MSUD Analog  
|                     | MSUD Maxamaid  
|                     | MSUD Maxamum  
|                     | Acerflex  
|                     | Ketonex-1  
|                     | Ketonex-2  
|                     | Complex MSUD Complex Essential MSD  
|                     | MSUD Gel  
|                     | MSUD Express  |
| Propionic acidemia (PPA)  
| Methylmalonic acidemia (MMA) | OA1  
|                     | OA2  
|                     | OS2  
|                     | XMTVI Analog  
|                     | XMTVI Maxamaid  
|                     | XMTVI Maxamum  
|                     | Propimex-1  
|                     | Propimex-2  
|                     | MMA /PA Gel  
|                     | MMA/PA Express  |
| Phenylketonuria (PKU), Hyperphenylalaninemia | PhenylFree 1  
|                     | PhenylFree 2  
|                     | Phenyl-Free 2HP  
|                     | PKU1  
|                     | PKU2  
|                     | PKU3  
|                     | XPhe Analog  
|                     | XPhe Maxamaid  
|                     | XPhe Maxamum  
|                     | Lophlex  
|                     | Periflex  
|                     | Phlexy-10  
|                     | Phlexy-Vits Add-Ins  
|                     | Phenex-1  
|                     | Phenex-2  
|                     | PhenylAde  
|                     | PKU Gel  
<p>|                     | PKU Express PKU Cooler Camino Pro  |</p>
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<td>XMet, XCys Maxamaid</td>
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<td>XPhe, XTyr Analog</td>
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<td>XPTM Analog</td>
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<td>Tyrex-1</td>
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<td>TYR Gel</td>
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<td>TYR Express TYR Cooler</td>
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<td>Urea Cycle Disorders (UCD)</td>
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<td>(Citrullinemia, Arginosuccinic Acidura)</td>
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<td>Essential Amino Acid Mix</td>
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<td>Cyclinex-1</td>
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<td>Cyclinex-2</td>
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<td>All Disorders (Protein Free Energy Modules)</td>
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