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Medical Food for Inherited Metabolic Disease
Corporate Medical Policy

File Name: Medical Food for Inherited Metabolic Disease
File Code: 5.01.VT203
Origination: 10/1998
Last Review: 09/2023
Next Review: 10/2024
Effective Date: 10/01/2023

Description/Summary

Inherited metabolic disease is defined as a disease caused by an inherited abnormality of body chemistry for which the state screens newborn infants.

Over 700 inborn errors of metabolism (IEM) are known; they are hereditary defects that interfere with one or more biochemical functions that are essential for life. Disorders in the intermediary metabolism of protein, carbohydrate or lipids cause serious problems in infants and are most amenable to treatment with medical foods. In these disorders, either excess of one or more metabolites, or conversely, lack thereof from failure of endogenous synthesis, becomes critical and without correction of the metabolic abnormalities, severe systemic disease ensues. Although some IEM disorders require medical foods for treatment, many do not.

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 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

Coding Information

Click the links below for attachments, coding tables & instructions:
Attachment I- CPT®/HCPCS Code Table & Instruction
Attachment II- ICD-10-CM Code Table

See Attachment III for Medical Food & Medical Disorders See
Attachment IV for various definitions

Policy Guidelines

When a service may be considered medically necessary

Medical foods are considered medically necessary for the dietary treatment of inherited metabolic disease (IMD) and inborn errors of metabolism (IEM), when prescribed by a physician and administered under the direction of a physician and meet **ALL** of the following criteria:

- The product is a medical food, as defined above.
- The product is being used to treat IMD/IEM.
- Use of product is essential for the treatment of IMD/IEM and failure to use medical foods will predictably result in adverse medical outcomes.
- The product is labeled for the dietary management of IMD/IEM.
- The product is labeled to be used under medical supervision.

The Plan covers medical foods for enteral nutrition including low protein formula in accordance with the Vermont State Mandate for the medically necessary treatment of an inherited metabolic disease and inborn errors of metabolism, when prescribed by a physician. See BCBSVT Enteral Nutrition Corporate Medical Policy for additional information.

As of the original effective date of this medical policy, the State of Vermont requires newborn screenings for the following metabolic disorders. Therefore, medical foods and formulas that meet criteria above, prescribed for these disorders are covered, subject to the benefit limitations described in this medical policy. This list is subject to change and is not all inclusive,and as such, medical food or formulas may be medically indicated for other conditions.

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

- Argininosuccinic acidemia (ASA)
- Beta-ketothiolase deficiency (BKT)
- *Biotinidase Deficiency
- *Carnitine uptake defect (CUD)
- Citrullinemia (CIT)
- *Congenital adrenal hyperplasia (CAH)
- *Congenital Hypothyroidism (CH)
- *Cystic fibrosis (CF)
- Galactosemia (GALT)
- 3-OH 3-CH3 glutaric aciduria (HMG)
- Glutaric acidemia type I (GA I)

- *Hb S/Beta-thalassemia (Hb S/BTh)
- *Hb S/C disease (Hb S/C)
- Homocystinuria (HCY)
- Isovaleric acidemia (IVA)
- *Long-chain L-3-OH acyl-CoA dehydrogenase deficiency (LCHAD)
- Maple Syrup Urine Disease (MSUD)
- *Medium-chain acyl-CoA dehydrogenase deficiency (MCAD)
- 3-Methylcrotonyl-CoA carboxylase deficiency (3MCC)
- Methylmalonic acidemia (Cbl A, B)
- Methylmalonic acidemia (mutase deficiency) (MUT)
- Multiple carboxylase deficiency (MCD)
- Phenylketonuria (PKU)
- Propionic acidemia (PROP)
- *Sickle cell anemia (Hb SS disease) (SS)
- *Trifunctional protein deficiency (TFP)
- Tyrosinemia type I (TYR I)
- *Very long-chain acyl-CoA dehydrogenase deficiency (VLCAD)

Condition Name and Abbreviation — curated by the National Library of Medicine (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.

Medical foods are not generally available in grocery stores, supermarkets, health food stores or over the counter at a retail pharmacy.

When a service is considered a benefit exclusion and therefore non-covered

- Non-medical charges, such as:
    - taxes
    - postage, shipping and handling charges
    - a penalty for failure to keep a scheduled visit
      fees for completion of a claim form.

- Nutritional Formula that does not require a prescription. This includes:
    - Over-the-counter formulas or nutritional supplements and formulas that can be purchased in a supermarket, health food store, over the counter at a retail pharmacy (Ensure®, Similac®, Enfamil®, Sustacal® [not an exhaustive list]) regardless of the route of administration and regardless of whether or not prescribed by a physician.
    - In the category of personal service, comfort or convenience items are formulas and supplements described by HCPCS Codes B4102-B4104, B4149-B4152, B4158-B4160.

- Food and nutritional formula or supplements except for “medical foods” prescribed for the medically necessary treatment of an inherited metabolic

disease or prescription formula and supplements administered through a feeding tube.

Legislative Guidelines

This policy complies with Vermont Act 128 V.S.A. § 4089e. Added 1997, effective April 27, 1998.

Reference Resources

1. Rezvani I. Defects in metabolism of amino acids. Behrman RE, Kleigman the RM, Jenson HB, editors. In: Nelson textbook of pediatrics. 17 Philadelphia, PA: W.B. Saunders Company; 2004.p.398-433
2. Rezvani I. An approach to inborn errors of metabolism. Kleigman RM editor. In: Nelson textbook of pediatrics. 18 ed. Philadelphia, PA. Saunders 2007
3. The following website is usefulto determine which products are assigned to a specific HCPCS code: https://www.dmepdac.com/
4. The following website was accessed 06/27/13: http://www.fda.gov/downloads/Food/ComplianceEnforcement/UCM073339.pdf
5. Vermont Statute on Treatment of inherited metabolic diseases. 8 V.S.A. §4089e. https://legislature.vermont.gov/statutes/section/08/107/04089e Accessed 8/2023.

Document Precedence

Blue Cross and Blue Shield of Vermont (BCBSVT) Medical Policies are developed to provide clinical guidance and are based on research of current medic al literature and review of common medical practices in the treatment and diagnosis of disease. The applicable group/individual contract and member certificate language, or employer's benefit plan if an ASO group, determines benefits that are in effect at the time of service. Since medical practices and knowledge are constantly evolving, BCBSVT reserves the right to review and revise its medical policies periodically. To the extent that there may be any conflict between medical policy and contract/employer benefit plan language, the member's contract/employer benefit plan language takes precedence.

Audit Information

BCBSVT reserves the right to conduct audits on any provider and/or facility to ensure compliance with the guidelines stated in the medical policy. If an audit identifies instances of non-compliance with this medical policy, BCBSVT reserves the right to recoup all non-compliant payments.

Administrative and Contractual Guidance

Benefit Determination Guidance

Prior approval is required and benefits are subject to all terms, limitations and conditions of the subscriber contract.

NEHP/ABNE members may have different benefits for services listed in this policy. To

confirm benefits, please contact the customer service department at the member's health plan.

Federal Employee Program (FEP): Members may have different benefits that apply. For further information please contact FEP customer service or refer to the FEP Service Benefit Plan Brochure. It is important to verify the member's benefits prior to providing the service to determine if benefits are available or if there is a specific exclusion in the member's benefit.

Coverage varies according to the member's group or individual contract. Not all groups are required to follow the Vermont legislative mandates. Member Contract language takes precedence over medical policy when there is a conflict.

If the member receives benefits through an Administrative Services (ASO) only group, benefits may vary or not apply. To verify benefit information, please refer to the member's employer benefit plan documents or contact the customer service department. Language in the employer benefit plan documents takes precedence over medical policy when there is a conflict.

Related Policies

BCBSVT Payment Policy (including enteral nutrition and total parenteral nutrition)

Enteral Nutrition

Home Infusion
Total Parenteral Nutrition (TPN) Nutritional Counseling
Metabolic Food for Inherited Metabolic Disorders

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.
10/2011	Medical/Clinical Coder reviewed
11/2013	(for 1/1/14 effective date) – Dollar limitation removed per VT State mandate requirement. Medical necessity criteria revised. ICD-10 remediated. Added diagnosis codes.
02/2016	Benefit exclusion criteria revised.
02/2017	Vermont Mandate reviewed with no current changes noted. Minor grammar changes. Removed HCPCS S9434 & S9435 from PA list. Policy Statement Unchanged.

01/2019	Vermont State Mandate reviewed -policy statement remain unchanged. References updated. Reviewed ICD-10-CM table updated table.
02/2020	Policy statement unchanged.
09/2020	Policy reviewed policy statements remain unchanged. Updated related policy statements.
03/2022	Policy statement language clarified, specific to the definition of medical foods to align with State of Vermont statute (8 V.S.A. § 4089e Treatment of inherited metabolic diseases).
10/2022	Annual policy review. Minor formatting changes. No change to policy statement. Deleted ICD-10-CM code in coding table E70.8 and replaced with code E70.81.
09/2023	Policy reviewed. Minor formatting changes. Removed policy language where medical foods can be purchased with manufacturer examples to avoid confusion around eligible manufacturer/distributors. No change to policy statement.

Eligible providers

Qualified healthcare professionals practicing within the scope of their license(s).

Approved by BCBSVT Medical Directors

Tom Weigel, MD, MBA
Vice President and Chief Medical Officer

Tammaji P. Kulkarni, MD
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Attachment I
CPT®/ HCPCS Code list & Instructions

Code Type	Number	Description	Policy Instructions
The following codes will be considered as medically necessary when applicable criteria have been met.			
HCPCS	B9998	Not Otherwise Classified for enteral supplies	Prior Approval required

Attachment II
ICD-10-CM Code List

ICD-10 Code	Description
The following diagnoses are considered medically necessary when applicable criteria is met.	

E70.0	Classical phenylketonuria
E70.1	Other hyperphenylalaninemias
E70.20	Disorder of tyrosine metabolism, unspecified
E70.21	Tyrosinemia
E70.29	Other disorders of tyrosine metabolism
E70.40	Disorders of histidine metabolism, unspecified
E70.41	Histidinemia
E70.49	Other disorders of histidine metabolism
E70.5	Disorders of tryptophan metabolism
E70.81	Aromatic L-amino acid decarboxylase deficiency
E70.9	Disorder of aromatic amino-acid metabolism, unspecified
E71.0	Maple-syrup-urine disease
E71.110	Isovaleric acidemia
E71.111	3-methylglutaconic aciduria
E71.118	Other branched-chain organic acidurias
E71.120	Methylmalonic acidemia
E71.121	Propionic acidemia
E71.128	Other disorders of propionate metabolism
E71.19	Other disorders of branched-chain amino-acid metabolism
E71.2	Disorder of branched-chain amino-acid metabolism, unspecified
E72.10	Disorders of sulfur-bearing amino-acid metabolism, unspecified
E72.11	Homocystinuria
E72.12	Methylenetetrahydrofolate reductase deficiency
E72.19	Other disorders of sulfur-bearing amino-acid metabolism
E72.20	Disorder of urea cycle metabolism, unspecified
E72.21	Argininemia
E72.22	Arginosuccinic aciduria
E72.23	Citrullinemia
E72.29	Other disorders of urea cycle metabolism
E72.3	Disorders of lysine and hydroxylysine metabolism
E72.4	Disorders of ornithine metabolism
E72.50	Disorder of glycine metabolism, unspecified

E72.51	Non-ketotic hyperglycinemia
E72.59	Other disorders of glycine metabolism
E72.89	Other specified disorders of amino-acid metabolism

Attachment III
Medical Disorders & Medical Food

Metabolic Disorder	Medical Food
Glutaric acidemia (GLA)	XLys, XTrp Analog XLys, XTrp Maxamaid XLys, XTrp Maxamum Glutarex-1 Glutarex-2 GA
Homocystinuria (HCU)	HCY1 HCY2 Hom2 Methionaid
	XMet Analog XMet Maxamaid XMet Maxamum Hominex-1 Hominex-2 HCU Gel HCU Express
Isovaleric acidemia (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 PKU Express PKU Cooler Camino Pro
Sulfite Oxidase Deficiency	XMet, XCys Analog XMet, XCys Maxamaid
Tyrosinemia	TYROS 1 TYROS 2 Tyr2 XPhe, XTyr Analog XPhe, XTyr Maxamaid XPTM Analog Tyrex-1 Tyrex-2 TYR Gel TYR Express TYR Cooler
Urea Cycle Disorders (UCD) (Citrullinemia, Arginosuccinic Acidura)	WND 1 WND 2 UCD2 Essential Amino Acid Mix Cyclinex-1 Cyclinex-2
All Disorders (Protein Free Energy Modules)	PFD1 PFD2 Super Soluble Duocal Pro-Phree

i Vermont law Added 1997, No. 128 (Adj. Sess.), § 1, eff. April 27, 1998: 8 V.S.A § 4089e. Treatment of inherited metabolic diseases.

Attachment IV
Definitions

Low protein modified food products are 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 foods are different from the broader category of foods in that they are an amino acid modified preparation, specially formulated and processed. They are intended for use under medical supervision to meet distinctive nutritional requirements of an inherited metabolic disease or condition.

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 (ketoaciduria), 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. Some types of medical foods may 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 in born 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.