

# Medical Foods and Specialized Formula for the Treatment of Inborn Errors of Metabolism



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**Medical Policy #: 10.01.15**  
**Original Effective Date:** May 2010  
**Reviewed:** April 2022  
**Revised:** April 2022

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**NOTICE:** This policy contains information which is clinical in nature. The policy is not medical advice. The information in this policy is used by Wellmark to make determinations whether medical treatment is covered under the terms of a Wellmark member's health benefit plan. Physicians and other health care providers are responsible for medical advice and treatment. If you have specific health care needs, you should consult an appropriate health care professional. If you would like to request an accessible version of this document, please contact customer service at 800-524-9242.

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations, or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This medical policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

This Medical Policy document describes the status of medical technology at the time the document was developed. Since that time, new technology may have emerged, or new medical literature may have been published. This Medical Policy will be reviewed regularly and be updated as scientific and medical literature becomes available; therefore, policies are subject to change without notice.

## DESCRIPTION

*Note: Refer to the member's benefit document to determine coverage.*

The U.S. Food and Drug Administration (FDA) defines a medical food in the Orphan Drug Amendment as a food which is formulated to be consumed or administered 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.

In 1994, the American Academy of Pediatrics Committee on Nutrition issued its recommendations on reimbursement for medical foods for inborn errors of metabolism.

Inborn errors of metabolism are a group of rare disorders resulting in the excessive accumulation of an amino acid or other product along the metabolic pathway for lack of a natural enzyme required to digest certain foods. Manifestations of these disorders generally include central nervous system dysfunction, developmental delay, seizures, and liver dysfunction.

The clinical manifestations in many of these disorders can be prevented if diagnosis is achieved early and necessary treatment with dietary protein or amino acid restriction is instituted immediately. These disorders are named for the accumulating amino acid and include, but are not limited to, phenylketonuria (PKU), maple syrup urine disease, citrullinemia, cystinosis, homocystinuria, methylmalonic acidemia, propionic acidemia, isovaleric acidemia type I, tyrosinemia types I and II, and urea cycle disorders.

Treatment might include restriction of specific amino acids, restriction of total nitrogen intake, or supplementation of certain substances. For some of the inborn errors of metabolism, special formulas and medical foods have been developed which eliminate the amino acid that cannot be metabolized from the protein context of the food. As adults, they must avoid certain foods as well. Women with classic PKU desiring pregnancy need to alter their diet by using a special maternal dietary supplement low in phenylalanine. The use of this supplement reduces the risk of severe retardation in the infant of a mother with PKU.

Inborn errors of metabolism (IEM) are inherited genetic disorders that interfere with specific metabolic pathways. These interferences may result in either a deficiency or excess of a particular product. There are three major categories of IEMs: protein disorders, fatty acid oxidation disorders, and carbohydrate disorders. Special food products are required to treat inborn errors of metabolism in-order to avoid long-term complications which can result in mental retardation, seizures, coma, or death. Inborn errors of metabolism are considered present at birth and permanent.

Examples of an inherited metabolic disorder (inborn errors of metabolism) include, but are not limited to:

- Phenylketonuria (PKU)
- Maple syrup urine disease (MSUD)
- Homocystinuria
- Urea cycle disorders
- Organic acidemias (e.g., Isovaleric Acidemia, propionic acidemia, methylmalonic acidemia)
- Histidinemia
- Tyrosinemia
- von Gierke's disease
- Pyruvate dehydrogenase deficiency

Individuals with inborn errors of metabolism require "exempt" specialized metabolic infant formulas. The FDA has requirements for the minimum and maximum amount of nutrients in infant formulas; if these requirements are not met, the infant formula may be "exempt" from these nutrient requirements. "An exempt infant formula is any infant formula which is represented and labeled for use by an infant who has an inborn error of metabolism (IEM), or low birth weight, or who otherwise has an unusual medical or dietary problem." Exempt amino acid-based metabolic formulas require a prescription.

Examples of eligible medical foods and low protein modified food products include, but are not limited to, the following:

ANALOG MSUD	ANALOG XMET	ANALOG XP	ANALOG XPHEN
ANALOG XLEU	ANALOG XLYS, TRY	CALCILO XD	CYCLINEX-1
CYCLINEX-2	GLUTAREX-1	GLUTAREX-2	HIST 1
HIST 2	HOM 1	HOM 2	HOMINEX-1
HOMINEX-2	I-VALEX-1	I-VALEX-2	KETONEX-1
KETONEX-2	LOFENALAC	LYS 1	LYS 2
MAXAMAID MSUD	MAXAMAID XLEU	MAXAMAID XP	METHIONAID
MSUD MAXAMUM	MSUD ANALOG	MSUD DIET	OS 1
OS 2	PHENEX-1	PHENEX-2	PHENYL-FREE
PKU 1	PKU 2	PKU 3	PKU AID
PREGESTIMIL	PORTAGEN	PRODUCT 3200 AB	PRODUCT 3200K
PRODUCT 3232A	PRODUCT 80056	PRO-PHREE	PROPIMEX-1
PROPIMEX-2	PROVIMIN	RCF	RCF SOY PROTEIN FORMULA
TYR 1	TYR 2	TYROMEX-1	TYREX-2
UCD 1	UCD 2	XLEU ANALOG	XLYS, TRY ANALOG
XMET ANALOG	XP ANALOG	XP ANALOG POWDER	XPHEN, TYR ANALOG
XPTM ANALOG	XLEU MAXAMAID	XLYS, TRY MAXAMAID	XLYS, TRY MAXAMUM
XMET MAXAMAID	XMET MAXAMUM	XMTVI MAXAMAID	XMTVI MAXAMUM
XP MAXAMAID	XP MAXAMUM	XPHEN, TYR MAXAMAID	

**Note: Food allergies are not considered an inborn error of metabolism.**

Medical Foods can be classified into the following categories:

- a) Nutritionally complete formulas; **or**
- b) Nutritionally incomplete formulas, including individual "modular" type products that may be mixed with other products before use (e.g., protein, carbohydrate, or fat modulars); **or**
- c) Formulas for metabolic (genetic) disorders in patients over 12 months of age; **or**
- d) Oral rehydration products.

The term Total Enteral Nutrition (TEN) infers that the individual is receiving more than 50% of their daily caloric intake via medical food products. If fewer than 50% of daily

calories are supplied by medical food products, they are considered supplemental. *Refer to the member's benefit document to determine coverage.*

Food products are not considered medical food items, regardless of their intended use, which may include the following: food thickeners, baby food, gluten-free food products, high protein powders and mixes, low carbohydrate diet foods, grocery items, nutritional supplement puddings, weight-loss foods and formula, grocery items that are used in specialized diets or have been modified for a special nutritional need, and regular grocery products that can be mixed in blenders regardless of whether these regular food products are taken orally or parenterally, *see medical policy 01.02.01 Home Enteral Therapy and Oral Enteral Formula.*

Foods that are considered low protein modified food products include, but are not limited to, cookies, bread, pasta, cheeses, soups, and desserts that have been modified to reduce the protein content of the food are available without a prescription and are not necessary as other regular food can be used to provide a complete diet. These foods are for convenience.

### **Regulatory Status**

According to the Food and Drug Administration (FDA) (section 5(b) of the Orphan Drug Act (21 U.S.C. 360ee (b) (3)) a product must meet all of the following minimum criteria to be considered a medical food:

1. The product must be a food for oral or tube feeding.
2. The product must be labeled for the dietary management of a specific medical disorder, disease, or condition for which there are distinctive nutritional requirements.
3. The product must be used under the supervision of a physician.

U.S. Food and Drug Administration. Regulatory Information. Section 5 of Orphan Drug Act. Available at: <http://www.fda.gov/Food/GuidanceRegulation/GuidanceDocumentsRegulatoryInformation/MedicalFoods/default.htm>.

## **PRIOR APPROVAL**

Not applicable.

## POLICY

*Note: Refer to the member's benefit document to determine coverage.*

### See Related Medical Policy

- **01.02.01 Enteral Therapy and Oral Formula**

### Medical Foods

Medical foods are **considered a non-covered benefit and not eligible for eligible for coverage** for any diagnosis **except** for inborn errors of metabolism which are present at birth, and permanent in nature, *see below criteria*.

Medical foods (**S9432, S9433, S9434, S9435**) may be considered **medically necessary** when **ALL** the following criteria are met:

1. The product must be a medical food for oral feeding that is prescribed by or authorized by a licensed healthcare practitioner and meets **ALL** of the following:
  - a) Nutritionally complete formulas for metabolic (genetic) disorders in individuals; **or**
  - b) individual "modular" type products that may be mixed with other products before use (e.g., protein, carbohydrate, or fat modulars); **or**
  - c) Oral rehydration products.; **and**
2. The product must be labeled and used for the dietary management of a specific medical disorder, disease, or condition for which there are distinctive nutritional requirements to avert the development of serious physical or mental disabilities or to promote normal development or function. The condition must be present at birth and considered a permanent condition **and**
  - a) Must be a condition associated with an inborn error of metabolism that interferes with metabolism of specific nutrients, including but not limited to the following:
    - Phenylketonuria (PKU)
    - Maple syrup urine disease (MSUD)
    - Homocystinuria
    - Urea cycle disorders
    - Organic acidemias
    - Histidinemia
    - Tyrosinemia; **and**
3. The product must be labeled for use under medical supervision; **and**
4. The product is the primary source of nutrition (that is, constitutes more than 50 percent of the intake for the individual).

**Note:** *If fewer than 50% of daily calories are supplied by medical food(s) for in-born error of metabolism they are considered supplemental and **non-covered benefit and not eligible for coverage.***

Medical foods for conditions other than permanent inborn errors of metabolism, including but not limited to the following products and conditions, are **considered non-covered benefit and not eligible for coverage:**

- Deplin for Depression and Schizophrenia
- Management of Diabetes
- Management of Pregnancy
- Medical foods/Supplements for Mitochondrial Disease (There are currently no medical food requirements for mitochondrial disease that averts the development of serious physical or mental disabilities or promotes normal development or function.)
- Metanx and Posiapn for Neuropathy
- Theramine for Chronic Pain
- Vayacog and Axona for Alzheimer's
- Vayarin for ADHD or Autism
- VSL #3 for Irritable Bowel Syndrome (IBS)

Food supplements, lactose-free foods, specialized infant formulas to include regular store-bought formula, banked human breast milk, fortifier powder, vitamins and/or minerals taken orally are **considered a non-covered benefit and not eligible for coverage**, regardless of whether these are prescribed or authorized by a licensed healthcare practitioner physician.

### **Modified Grocery Item Foods and Formulas**

Modified grocery item foods and formulas, even if categorized as medical foods by the manufacturer, are considered **non-covered benefit and not eligible for coverage.**

The purchase of food and formula that is modified for special diets is not considered a medical food available for coverage. This includes low protein/low phe groceries and over the counter formula. The coverage of over-the-counter foods are considered a **non-covered benefit and not eligible for coverage.** Foods that are considered low protein modified food products include, but are not limited to, cookies, bread, pasta, cheeses, soups, and desserts that have been modified to reduce the protein content of the food are available without a prescription and are not necessary as other regular food(s) can be used to provide a complete diet. These foods are for convenience **and are considered a non-covered benefit and not eligible for coverage.** *For specialized enteral oral formula see medical policy 01.02.01 Enteral Nutrition Therapy and Oral Formula.*

### **Food Thickener and Food Additives (B4100 and B4104)**

Food thickener and food additives are considered non-covered benefit and not eligible for coverage.

## PROCEDURE CODES AND BILLING GUIDELINES

To report provider services, use appropriate CPT\* codes, Alpha Numeric (HCPCS level 2) codes, Revenue codes, and/or ICD diagnosis codes.

- B4100 Food thickener, administered orally, per oz
- B4104 Additive for enteral formula (e.g., fiber)
- S9432 Medical foods for non-inborn errors of metabolism
- S9433 Medical food nutritionally complete, administered orally, providing 100% of nutritional intake
- S9434 Modified solid food supplements for inborn errors of metabolism
- S9435 Medical foods for inborn errors of metabolism

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<b>POLICY HISTORY</b>		
<b>Date</b>	<b>Reason</b>	<b>Action</b>
April 2022	Annual Review	Policy Revised
April 2021	Annual Review	Policy Revised
April 2020	Annual Review	Policy Revised
April 2019	Annual Review	Policy Revised
June 2018	Interim Review	Policy Revised
April 2018	Annual Review	Policy Revised
April 2017	Annual Review	Policy Revised
April 2016	Annual Review	Policy Revised
May 2015	Annual Review	Policy Revised
June 2014	Annual Review	Policy Renewed
June 2014	Interim Review	Policy Revised
August 2013	Annual Review	Policy Renewed
February 2013	Interim Review	Policy Revised
September 2012	Annual Review	Policy Renewed
September 2011	Annual Review	Policy Renewed

New information or technology that would be relevant for Wellmark to consider when this policy is next reviewed may be submitted to:

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