

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

Original Effective Date: May 2010

Review Date: April 2026

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DISCLAIMER/INSTRUCTIONS FOR USE

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 updated as scientific and medical literature becomes available; therefore, policies are subject to change without notice.

Related Policies:

- [01.02.01 Enteral Nutrition Therapy and Oral Formula](#)

Summary

Description

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. Inborne errors of metabolism (IEM) are considered present at birth and permanent.

Objective

The objective of this medical policy is to support the proper determination of whether the service would be considered medically necessary or non-covered as not a contract benefit.

PRIOR APPROVAL

Not applicable.

POLICY

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

Medical Foods

Medical foods are **considered a non-covered benefit and not 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 (**S9433, S9434, S9435**) may be considered **medically necessary** when the following criteria are met:

1. The product must be a medical food for oral feeding (see [Regulatory Status](#)) that is prescribed by or authorized by a *licensed healthcare practitioner;
2. The product must be labeled and used for the dietary management of a specific medical disorder, disease, or condition;
3. The individual's condition is associated with a permanent 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**
4. The product is the primary source of nutrition because adequate nutrition is not possible by dietary adjustment.

Individuals not meeting the above criteria or 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 ADHD or autism
- 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 PoDiaPn for neuropathy
- Theramine for chronic pain
- Vayacog and Axona for Alzheimer's
- 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**.

POLICY GUIDELINES

*Covered Provider

Physician or licensed qualified non-physician practitioner (a licensed qualified non-physician may include the following: Nurse Practitioners, Physician Assistants, or Clinical Nurse Specialist Practitioners).

Coding

See the [Codes table](#) for details.

BACKGROUND

The U.S. Food and Drug Administration (FDA) define 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.

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 Enteral Nutrition Therapy and Oral 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)(3) of the Orphan Drug Act (21 U.S.C. 360ee (b) (3)) a medical food is defined as the following:

"A food which is formulated to be consumed or administered enterally 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."

The following criteria clarifies the definition of a medical food and can be found in the FDA's regulations at 21 CFR 101.9(j)(8): "A medical food is exempt from the nutritional labeling requirements of 21 CFR 101.9 only if:

- i. It is specially formulated and processed product (as opposed to a naturally occurring foodstuff used in its natural state) for the partial or exclusive feeding of a patient by means of oral intake or enteral feeding by tube, meaning a tube or catheter that delivers nutrients beyond the oral cavity directly into the stomach or small intestine;
- ii. It is intended for the dietary management of a patient who, because of therapeutic or chronic medical needs, has limited or impaired capacity to ingest, digest, absorb, or metabolize ordinary foodstuffs or certain nutrients, or who has other special medically determined nutrient requirements, the dietary management of which cannot be achieved by the modification of the normal diet alone;
- iii. It provides nutritional support specifically modified for the management of the unique nutrient needs that result from the specific disease or condition, as determined by medical evaluation;
- iv. It is intended to be used under medical supervision; and

- v. It is intended only for a patient receiving active and ongoing medical supervision wherein the patient requires medical care on a recurring basis for, among other things, instructions on the use of the medical food.

Medical Foods can be classified into the following categories:

- a) Nutritionally complete formulas.
- 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).
- c) Formulas for metabolic (genetic) disorders in patients over 12 months or age; or
- d) Oral rehydration products.

U.S. Food and Drug Administration. Regulatory Information. Section 5(b)(3) of the Orphan Drug Act. Available

at: <http://www.fda.gov/Food/GuidanceRegulation/GuidanceDocumentsRegulatoryInformation/MedicalFoods/default.htm>.

SUPPLEMENTAL INFORMATION

The purpose of the following information is to provide reference material. Inclusion does not imply endorsement.

Practice Guidelines and Position Statements

Guidelines or position statements will be considered for inclusion in ‘Supplemental Information’ if they were issued by, or jointly by, a US professional society, an international society with US representation, or National Institute for Health and Care Excellence (NICE). Priority will be given to guidelines that are informed by a systematic review, include strength of evidence ratings, and include a description of management of conflict of interest.

Ongoing and Unpublished Clinical Trials

Some currently ongoing and unpublished trials that might influence this review can be located at clinicaltrials.gov.

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CODES

To report provider services, use appropriate CPT codes, HCPCS codes, Revenue codes, and/or ICD diagnosis codes.

Codes	Number	Description
CPT		
	None	
HCPCS		
	B4100	Food Thickener, administration 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 of inborn errors of metabolism
	S9435	Medical foods for inborn errors of metabolism
Type of Service	Administrative	
Place of Service	Home/Outpatient	

POLICY HISTORY

Date	Action	Action
April 2026	Annual Review	Policy Revised
April 2025	Annual Review	Policy Renewed
April 2024	Annual Review	Policy Renewed
April 2023	Annual Review	Policy Revised
April 2022	Annual Review	Policy Revised
April 2021	Annual Review	Policy Revised
April 2019	Annual Review	Policy Revised
June 2018	Annual 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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