Medical Coverage Policy

Effective Date.....3/15/2025
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Coverage Policy Number..... 0136

Nutritional Support

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INSTRUCTIONS FOR USE

The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted for services that are not accompanied by covered code(s) under the applicable Coverage Policy

will be denied as not covered. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

Overview

This Coverage Policy addresses the use of oral and enteral nutritional formula that may be necessary for some individuals to maintain adequate nutrition.

Coverage Policy

Coverage for oral formula for insured and other state-regulated benefit plans is frequently governed by state mandates. Oral nutritional formula benefit plan language differs significantly across plans but coverage is usually limited to formula for infants under 12 months of age with an inborn error of metabolism. Refer to the applicable benefit plan document to determine benefit availability and the terms, conditions and limitations of coverage.

Oral and Enteral Infant Nutritional Formula
Infant (i.e., ≤ 12 months of age) nutritional formula is considered medically necessary when specifically formulated for the treatment of an inborn error of metabolism (e.g., disorder of amino acid or organic acid metabolism).

Not Medically Necessary Items
Each of the following items is considered not medically necessary for ANY indication:

- food thickeners
- lactose-free products; products to aid in lactose digestion

Health Equity Considerations

Health equity is the highest level of health for all people; health inequity is the avoidable difference in health status or distribution of health resources due to the social conditions in which people are born, grow, live, work, and age.

Social determinants of health are the conditions in the environment that affect a wide range of health, functioning, and quality of life outcomes and risks. Examples include safe housing, transportation, and neighborhoods; racism, discrimination and violence; education, job opportunities and income; access to nutritious foods and physical activity opportunities; access to clean air and water; and language and literacy skills.

Acute malnutrition is primarily a problem in resource-limited regions especially South Asia (including Afghanistan, India, Pakistan, Bangladesh, and Nepal) and sub-Saharan Africa. Acute malnutrition is uncommon in North America, Australia, and other resource-rich regions (Goday, 2024). The prevalence of chronic malnutrition has gradually declined in most regions during the past three decades (from 39.3% in 1990 to 22% in 2020). The decline is associated with improvements in education, socioeconomic status, sanitation, access to maternal health services, and family planning. However, the prevalence of stunting remains unacceptably high in many

regions, particularly South Asia and sub-Saharan Africa, where it affects more than 30% of children (Goday, 2024).

Some malabsorption syndromes can be estimated by discussing the epidemiology of subgroups. Gluten-sensitive enteropathy (GSE) is present at its highest rates in Europeans and North Americans. GSE can be found in parts of India and is rarest in those of Asian, Caribbean, and African descent. Tropical sprue is known for affecting residents and visitors to Puerto Rico, the Caribbean, West Africa, northern South America, south-east Asia, and India (Zuvarox and Belletieri, 2023).

In the United States today, access to medical foods is not ensured for many individuals who are affected despite their proven efficacy in the treatment of IEMs, their universal use as the mainstay of IEM management, the endorsement of their use by professional medical organizations, and the obvious desire of families for effective care.

General Background

Specialized nutritional support is often required for patients who have chronic disease or for those undergoing long-term rehabilitation who are at risk for malnutrition. Nutritional support can be provided orally, enterally (through a tube into the stomach or small intestine) and intravenously.

Malnutrition is commonly defined in the medical literature as a non-edematous or post-dialysis weight loss of at least 10% of ideal body weight over a three-month period or a serum albumin of less than 3.4 grams/dL. Malnutrition can occur in otherwise healthy individuals when they are deprived of adequate nutrients for an extended period of time. Compared to younger adults, malnutrition in older individuals is more common and can have a greater impact on health outcomes. The prevalence of malnutrition in older adults is dependent upon the population studied, and varies by geography, age and living situation (Ritchie and Yukawa, 2024).

Oral and Enteral Nutrition
Nutritional support provided via the gastrointestinal tract can be taken by mouth or provided enterally. Oral nutrition refers to nutrition taken through the mouth. Enteral nutrition is commonly defined as the provision of nutritional requirements through a tube in the stomach or small intestine. The major types of enteral nutrition formulas include the following (Pham and McClave, 2021; Makola, 2005):

- Elemental/amino acid-based: contains protein in the form of free amino acids and are nearly fat-free (i.e., less than 2% to 3% of the caloric content is fat)
- Semi-elemental: contains protein in the form of small chain peptides, simple sugars, glucose polymers or starch and fat, primarily as medium chain triglycerides (MCT)
- Standard or polymeric: contains 15% to 20% calories from proteins, 45% to 60% calories from carbohydrates, and 30% to 40% calories from fats
- Specialized/disease-specific and immune-enhancing: contains biologically active substances or nutrients such as glutamine, arginine, nucleotides or essential fatty acids

For certain metabolic and malabsorption disorders, an infant or child may require an elemental form because of the inability to digest whole proteins found in standard formula. Individuals may require enteral nutritional therapy to provide sufficient nutrients to maintain weight and strength commensurate with their overall health status if their nutritional needs cannot be met through dietary adjustments and/or oral supplements. In general, patients may require enteral nutritional therapy when they have one of the following:

- a functional impairment or disease of the structures that normally permit food to reach the small bowel
- a disease of the small bowel that impairs digestion and/or absorption of an oral diet

For most facility inpatient stays, room and board, including food, are covered. Room and board charges are generally inclusive of the bed, related room charges, and food, including special diets such as cardiac, diabetic, clear liquids, etc., along with general services and activities needed for the care of an inpatient. Some services such as enteral and intravenous feedings may be reimbursed separately.

Inborn Errors of Metabolism
Oral or enteral nutritional formula may be required for infants, defined by the Centers for Disease Control and Prevention (CDC) as zero to one year of age (CDC, 2024), with inborn errors of metabolism (IEM). The broad category of "metabolic diseases" includes inborn errors of amino acid metabolism, such as phenylketonuria, maternal phenylketonuria, maple syrup urine disease, homocystinuria, methylmalonicacidemia, propionicacidemia, isovalericacidemia, and other disorders of leucine metabolism; glutaricaciduria type I and tyrosinemia types I and II; and urea cycle disorders. These are all disorders treatable by dietary modifications, which can prevent complications like severe intellectual disability and death (Greer, American Academy of Pediatrics Committee on Nutrition, 2003). In these disorders, the metabolic pathway is disrupted and excessive accumulation of an amino acid or other product results. If the appropriate dietary restriction of one or more applicable amino acids is introduced early in life, complications can be prevented or limited. Specialized medical food, including low protein and amino acid modified food, may be required as the infant outgrows the need for formula. Globally, different IEM classifications and IEM screening methods are used which causes the prevalence of IEMs to be different between countries. Because of their heterogeneity, different disorders have different distinct epidemiologies, presentations, and inheritability. There are certain populations that have increased carrier rates for IEM and testing of asymptomatic future parents decreases disease prevalence. Preconception screening started in the Ashkenazi Jewish population which screened for carriers of Tay-Sachs disease. Due to the carrier screening, the prevalence of Tay-Sachs disease decreased by 90% between 1970 and 1993 in the Jewish populations of North America (Jeanmonod, et al., 2022; Ismail, et al., 2020; Kruszka and Regier, 2019).

While conditions such as mitochondrial disorders are considered metabolic diseases, there is typically no specific treatment for these disorders. As such, specific nutritional replacement therapy does not treat the condition or prevent neurologic injury and subsequent developmental issues in infants and children.

Malabsorption Syndromes
Malabsorption is impaired nutrient absorption at any point where nutrients are absorbed. Malabsorption disorders can be caused by mucosal abnormalities usually resulting in malabsorption of multiple nutrients or malabsorption of specific nutrients (carbohydrate, fat, protein, vitamins, minerals, and trace elements). Malabsorption affects millions of people worldwide and has multiple etiologies that obscures the prevalence and incidence. However, some malabsorption syndromes can be estimated by discussing the epidemiology of subgroups. Gluten-sensitive enteropathy (GSE) is present at its highest rates in Europeans and North Americans. GSE can be found in parts of India and is rarest in those of Asian, Caribbean, and African descent. Tropical sprue is known for affecting residents and visitors to Puerto Rico, the Caribbean, West Africa, northern South America, south-east Asia, and India (Zuvarox and Belletieri, 2023).

Individuals with malabsorption syndromes may benefit from enteral nutritional support. Enteral nutritional support may be indicated when the formula comprises the primary source of nutrition (i.e., 60% or more of caloric nutritional intake).

Malabsorption syndromes may be associated with or due to a number of diseases, including but not limited to (Zuvarox and Belletieri, 2023; Mason, 2023; Shamir, 2020):

- pancreatic amylase deficiency
- lactase deficiency (hypolactasia)
- lactose malabsorption
    - tropical sprue
    - celiac disease
    - Crohn's disease
    - ulcerative colitis (when there are documented objective signs and symptoms of malabsorption such as serum albumin levels)
    - small bowel resection
- small bowel disease
- small intestinal bacterial overgrowth
    - cystic fibrosis
    - chronic pancreatitis
    - pancreatic cancer
    - Whipple disease
- liver disease
- cholestasis
- inflammatory bowel disease (IBS)
    - bariatric surgery
    - fat malabsorption
    - vitamin B12 deficiency

Malabsorption can also be due to the use of specific drugs that cause inadequate digestion or bind or precipitate bile salts such as neomycin, cholestyramine and orlistat.

Not Medically Necessary Items
U.S. Food and Drug Administration (FDA) – Medical Foods: Per the FDA, medical foods "are not those simply recommended by a physician as part of an overall diet to manage the symptoms or reduce the risk of a disease or condition. Not all foods fed to patients with a disease, including diseases that require dietary management, are medical foods". The FDA defines medical foods as "foods that are specially formulated and processed (as opposed to a naturally occurring foodstuff used in a natural state) for a patient who requires use of the product as a major component of a disease or condition's specific dietary management. Medical foods are intended solely to meet the dietary needs of individuals who have specific metabolic or physiological limitations that restrict their ability to digest regular food" (FDA, 2023).

Regular food products are not considered medical items and are not considered medically necessary. These products can include the following:

- standardized or specialized infant formula for conditions other than inborn errors of metabolism, including, but not limited to: food allergies; multiple protein intolerances; lactose intolerances; gluten-free formula for gluten-sensitive enteropathy/celiac disease; milk allergies; sensitivities to intact protein; protein or fat maldigestion; intolerances to soy formulas or protein hydrolysates; prematurity; or low birthweight
- baby food
- banked breast milk provided to a non-hospitalized infant
- dietary additives and food supplements
- food thickeners
- gluten-free food products

- grocery items that can be blenderized and used with an enteral feeding system
- high protein powders and mixes
- lactose-free products; products to aid in lactose digestion
- low carbohydrate diets
- normal grocery items
- nutritional supplement puddings
- oral/enteral formula used to replace fluids and electrolytes

Medicare Coverage Determinations

	Contractor	Determination Name/Number	Revision Effective Date
NCD	National	No Determination found	
LCD	CGS Administrators, LLC	Enteral Nutrition L38955	1/1/2024
LCD	CGS Administrators, LLC	Parenteral Nutrition L38953	1/1/2024

Note: Please review the current Medicare Policy for the most up-to-date information.
(NCD = National Coverage Determination; LCD = Local Coverage Determination)

Coding Information

Notes:

1. This list of codes may not be all-inclusive since the American Medical Association (AMA) and Centers for Medicare and Medicaid Services (CMS) code updates may occur more frequently than policy updates.
2. Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Oral and Enteral Infant Nutritional Formula

Considered Medically Necessary when infant (i.e., ≤ 12 months of age) nutritional formula is specifically formulated for the treatment of an inborn error of metabolism (e.g., disorder of amino acid or organic acid metabolism):

HCPCS Codes	Description
B4154†	Enteral formula, nutritionally complete, for special metabolic needs, excludes inherited disease of metabolism, includes altered composition of proteins, fats, carbohydrates, vitamins and/or minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit
B4155†	Enteral formula, nutritionally incomplete/modular nutrients, includes specific nutrients, carbohydrates (e.g. glucose polymers), proteins/amino acids (e.g. glutamine, arginine), fat (e.g. medium chain triglycerides) or combination, administered through an enteral feeding tube, 100 calories = 1 unit
B4157	Enteral formula, nutritionally complete, for special metabolic needs for inherited disease of metabolism, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit

HCPCS Codes	Description
B4162	Enteral formula, for pediatrics, special metabolic needs for inherited disease of metabolism, includes proteins, fats, carbohydrates, vitamins and minerals, may include fiber, administered through an enteral feeding tube, 100 calories = 1 unit
S9435[††]	Medical foods for inborn errors of metabolism

[†]Note: Generally excluded from coverage when used to treat conditions other than infant inborn errors of metabolism

[††]Note: Considered Medically Necessary if used to report infant nutritional formula for the treatment of inborn errors of metabolism

ICD-10-CM Diagnosis Codes	Description
D81.810	Biotinidase deficiency
D81.818	Other biotin-dependent carboxylase deficiency
E70.0	Classical phenylketonuria
E70.1	Other hyperphenylalaninemias
E70.20-E70.29	Disorder of tyrosine metabolism
E70.40-E70.49	Disorders of histidine metabolism
E70.5	Disorders of tryptophan metabolism
E70.81	Aromatic L-amino acid decarboxylase deficiency
E70.89	Other disorders of aromatic amino-acid metabolism
E70.9	Disorder of aromatic amino-acid metabolism, unspecified
E71.0	Maple-syrup-urine disease
E71.110-E71.118	Branched-chain organic acidurias
E71.120-E71.128	Disorders of propionate metabolism
E71.19	Other disorders of branched-chain amino-acid metabolism
E71.2	Disorder of branched-chain amino-acid metabolism, unspecified
E71.30	Disorder of fatty-acid metabolism, unspecified
E71.310-E71.318	Disorders of fatty-acid oxidation
E71.32	Disorders of ketone metabolism
E71.39	Other disorders of fatty-acid metabolism
E71.40	Disorders of carnitine metabolism, unspecified
E71.41	Primary carnitine deficiency
E71.42	Carnitine deficiency due to inborn errors of metabolism
E71.448	Other secondary carnitine deficiency
E71.50	Peroxisomal disorder, unspecified
E71.520	Childhood cerebral X-linked adrenoleukodystrophy
E71.53	Other group 2 peroxisomal disorders
E71.541	Zellweger-like syndrome
E71.542	Other group 3 peroxisomal disorders
E71.548	Other peroxisomal disorders

ICD-10-CM Diagnosis Codes	Description
E72.00-E72.09	Disorders of amino-acid transport
E72.10-E72.19	Disorders of sulfur-bearing amino-acid metabolism
E72.20-E72.29	Disorder of urea cycle metabolism
E72.3	Disorders of lysine and hydroxylysine metabolism
E72.4	Disorders of ornithine metabolism
E72.50-E72.59	Disorder of glycine metabolism
E72.89	Other specified disorders of amino-acid metabolism
E72.9	Disorder of amino-acid metabolism, unspecified
E74.00-E74.09	Glycogen storage disease
E74.21	Galactosemia
E74.29	Other disorders of galactose metabolism
E74.4	Disorders of pyruvate metabolism and gluconeogenesis
E75.21	Fabry (-Anderson) disease
E75.22	Gaucher disease
E75.23	Krabbe disease
E75.26	Sulfatase deficiency
E78.72	Smith-Lemli-Opitz syndrome
E88.41	MELAS syndrome
E88.42	MERRF syndrome

Additional Items

Considered Not Medically Necessary:

HCPCS Codes	Description
B4100	Food thickener, administered orally, per ounce

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Revision Details

Type of Revision	Summary of Changes	Date
Focused Review	No clinical policy statement changes	3/15/2025
Focused Review	No clinical policy statement changes	11/15/2024
Annual Review	Removed policy statements for: • Home enteral infusion pumps • Home parenteral nutrition • Home parenteral infusion pumps • Intradialytic Parenteral Nutrition • standardized or specialized infant formula for conditions other than inborn errors of metabolism, including, but not limited to: food allergies; multiple protein intolerances; lactose intolerances; gluten-free formula for gluten-sensitive enteropathy/celiac disease; milk allergies; sensitivities to intact protein; protein or fat maldigestion; intolerances to soy formulas or protein hydrolysates; prematurity; or low birth-weight • baby food • banked breast milk provided to a non-hospitalized infant • dietary additives and food supplements • gluten-free food products • grocery items that can be blenderized and used with an enteral feeding system • high protein powders and mixes • low carbohydrate diets • normal grocery items • nutritional supplement puddings • oral/enteral formula used to replace fluids and electrolytes • oral vitamins and minerals • weight-loss foods and formula; products to aid weight loss	9/15/2024