Department:	Medical Management	**Original Approval:**	05/31/2016
Policy #:	MM147	**Last Approval:**	05/08/2018
Title:	Enteral Therapy Products for Enrollees with Inherited Metabolic Disorders and for nutritional support		
Approved By:	UM Committee		

BACKGROUND

This Policy is written and disseminated to ensure compliance with a directive from the WA Healthcare Authority regarding coverage of oral and tube-delivered enteral products for metabolic disorders in children and adults. CHPW reserves right to do medical necessity reviews regarding enteral and oral formulas and may do post payment reviews. This includes medical necessity determinations for the particular formula that has been used.

DEFINITIONS

Enteral therapy is liquid nutrition formulated for tube feeding for:

- People with inherited metabolic disorders who are dependent on special formulas (supplied by tube or orally) due to a diagnosed metabolic disorder that otherwise presents a life threatening inability to consume normal food. The formulas can supply a particular vital nutrient that the member is unable to absorb (that cannot be corrected with supplements to a normal diet) or eliminate a component of normal food which would be toxic to the member (and cannot be corrected by modifications to a normal diet). Some examples are amino acid, fatty acid, and carbohydrate metabolic disorders including PKU. Additional rare disorders are listed in the appendix at the end of this policy.

- People with a functioning GI system who, because of a neurologic or anatomic problem (congenital or acquired), are unable to take in sufficient nutrition by mouth to maintain their weight, (i.e. are dependent on tube feeding for over 50% of their caloric intake).

ENTERAL THERAPY FOR A MEMBER WITH AN INHERITED METABOLIC DISORDER:

WA APPLE HEALTH ENROLLEES:

INDICATIONS/CRITERIA

The criteria for enteral therapy for members with inherited metabolic disorders include all of the following:

- The member has a documented inherited metabolic disorder.
- The member requires the formula to supply at least 50 % of their daily caloric intake.
- The member has documented need for the type of formula used,

- The rationale for the particular formula that is requested and if the member has failed other formulas.

COVERAGE (ENTERAL THERAPY FOR A MEMBER WITH AN INHERITED METABOLIC DISORDER):

- For Children under 21 years of age**:** No prior authorization is required for either oral or tube feeding preparations of nutritional replacement products for enrollees with an inherited metabolic disorder. Prescriptions are required at least once per year, or sooner as per physician recommendations. These products and prescriptions are subject to post-payment medical necessity review.
- For Adults 21 years of age and older: No prior authorization is required for tube feeding preparations of nutritional replacement products for enrollees with an inherited metabolic disorder who are receiving 100% of their nutritional needs via feeding tube. Prescriptions are required at least once per year, or sooner as per physician recommendations. These products and prescriptions are subject to post-payment medical necessity review.
- For Adults 21 years of age and older with inherited metabolic disorders who can take their nutritional replacement products orally the following process must occur:
  - Oral nutritional supplements for adults are a "non-covered benefit".
  - When ETR request is received and information is complete with diagnosis and prescription of nutritional replacement product ("formula"), treat it as an Exception to Rule and send for physician review.
  - ETR requests are not required more often than once per year.
  - The Exception to Rule is granted by this policy ONLY to nutritional replacement formulas. These products and prescriptions are subject to post-payment medical necessity review.

NOTE: There is no Exception to Rule granted for "special" foods. These remain non-covered . If there are other food products available, such that avoiding a certain food would allow the member to have a fairly normal diet, there should not be an expectation of exception to the rule.

MEDICARE ADVANTAGE ENROLLEES (INHERITED METABOLIC DISORDERS):

CHPW uses the CMS guidelines for enteral nutrition for decisions related to enrollees with inherited metabolic disorders. CMS Enteral Therapy guidelines:

NCD 180.2:
http://www.cms.hhs.gov/manuals/downloads/ncd103c1_Part3.pdf
See section 180.2.

ENTERAL THERAPY FOR NUTRITIONAL SUPPORT IN A MEMBER WITHOUT AN INHERITED METABOLIC DISORDER:

INDICATIONS/CRITERIA

For Medicare and Apple Health:
Enteral nutrition is considered reasonable and necessary for a patient with a functioning gastrointestinal tract who, due to pathology to, or non-function of, the structures that normally permit food to reach the digestive tract, cannot maintain weight and strength commensurate with his or her general condition. Typical examples of conditions that qualify for coverage are head and neck cancer with reconstructive surgery and central nervous system disease leading to interference with the neuromuscular mechanisms of ingestion of such severity that the beneficiary cannot be maintained with oral feeding. Enteral therapy may be given by nasogastric, jejunostomy, or gastrostomy tubes.

Enteral nutrition is considered medically necessary for either of the following:

- The diet consists of more than 50% enteral nutrition, OR
- If the diet is less than 50 percent enteral nutrition and more than 50 percent standard diet for age, AND:
  - The enteral product is used as part of a defined and limited plan of care in transition from a diet of more than 50 percent enteral products to standard diet for age; OR
  - Medical records document a medical basis for the inability to maintain appropriate body weight and nutritional status prior to initiating or after discontinuing use of an enteral supplement as well as ongoing evidence of response to the enteral nutrition.

Preferred Formulas:
The preferred formulas are: B-4150 basic enteral formula, B-4152 Caloric dense formula, B-4158 Pediatric nutritionally complete, B-4159 Pediatric nutritionally complete Soy based, B-4160 Pediatric nutritionally complex caloric dense.
The need for a specific enteral formula (other than the preferred formulas)), being prescribed must be documented including

- Failed trials of the preferred formulas, or
- Known intolerance to the preferred formulas

WA APPLE HEALTH ENROLLEES (WITHOUT AN INHERITED METABOLIC DISORDER):

CHPW uses the Medicaid Provider Guide Enteral Nutrition (Chapter 182-554 WAC) available at:
https://www.hca.wa.gov/assets/billers-and-providers/enteral-nutrition-bi-20180101.pdf
See most recent guide.

MEDICARE ADVANTAGE ENROLLEES (WITHOUT AN INHERITED METABOLIC DISORDER):

CHPW uses the CMS guidelines for this service. Visit the following web site for Enteral Therapy guidelines:
NCD 180.2:
http://www.cms.hhs.gov/manuals/downloads/ncd103c1_Part3.pdf

See section 180.2.

Oral Nutritional Supplementation

Some patients require supplementation of their daily protein and caloric intake. Nutritional supplements are often given as a medicine between meals to boost protein-caloric intake or the mainstay of a daily nutritional plan. **Oral Nutritional Supplementation is not covered under Medicare.**

SPECIAL CONSIDERATIONS

None.

LIMITATIONS/EXCLUSIONS

WIC: Women, Infants, and Children and tube-delivered enteral nutrition

If a WIC-eligible tube-fed client can use a standard formula available from WIC, the client must receive the product from WIC. All clients under age 5, including tube-fed clients, must receive products and formulas directly from WIC unless:

- The client is not eligible for the WIC program.
- The client is eligible for the WIC program, but the need for the enteral nutrition product or formula exceeds WIC's allowed amount.
- The requested product or formula, or the equivalent, is not available through the WIC program.

Please refer to a product line's certificate of coverage for benefit limitations and exclusions for these services:

PRODUCT LINE	LINK TO CERTIFICATE OF COVERAGE
MEDICARE ADVANTAGE	http://healthfirst.chpw.org/for-members/resource-library/handbooks-and-guides
WASHINGTON HEALTH PROGRAM	http://chpw.org/our-plans/apple-health/

Citations & References

CFR		
WAC	WAC Chapter 182-554 WAC	
RCW		
Contract Citation	☒ WAH	http://chpw.org/our-plans/apple-health/
	☒ IMC	
	☒ MA	http://healthfirst.chpw.org/for-members/resource-

		library/handbooks-and-guides
Other Requirements		
NCQA Elements		

Revision History

Revision Date	Revision Description	Revision Made By
05/31/2016	New policy	Kate Brostoff, MD
06/02/2016	Approval	MMLT
06/13/2017	Minor formatting changes	Cyndi Stilson, RN
06/15/2017	Verified no changes to tables for 2017	LuAnn Chen, MD
06/16/2017	Approval	MMLT
03/27/2018	Changed from UM157 to MM147	Cindy Bush
04/05/2018	Transferred to new template	Cindy Bush
05/02/2018	Checked links, incorporated MM137-Enteral therapy into this policy. Clarified Medicare coverage of enteral therapy. Explained role of post-payment review. Preferred enteral formulas listed.	LuAnn Chen, MD
05/08/2018	Approval	UM Medical Subcommittee

APPENDIX A:
NON-COMPREHENSIVE LIST OF INHERITED METABOLIC DISORDERS FROM UP-TO-DATE, 2016.
Inclusion on this list does not mean that enteral therapy is definitely indicated for a condition.

UpToDate®

Types of inborn errors of metabolism

Disorders of intermediary metabolism
Amino acid metabolism and transport
Fatty acid oxidation and ketogenesis
Carbohydrate metabolism and transport
Vitamin-related (cobalamin, folate)
Peptide metabolism
Mineral metabolism
Mitochondrial energy metabolism
Disorders of biosynthesis and breakdown of complex molecules
Purine and pyrimidine metabolism
Lysosomal storage
Peroxisomes
Isoprenoid and sterol metabolism
Bile acid and heme metabolism
Glycosylation
Lipoprotein metabolism
Disorders of neurotransmitter metabolism
Glycine and serine metabolism
Pterin and biogenic amine metabolism
Gamma-aminobutyrate metabolism
Other (eg, pyridoxine-dependent or folinic acid-dependent seizures, sulfite oxidase deficiency)

Source: Hoffman GF, Nyhan WL, Zschocke J, et al. Inherited metabolic diseases, Lippincott Williams & Wilkins, Philadelphia 2002.

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Selected disorders of carbohydrate metabolism

Carbohydrate intolerance disorders
Galactosemia
Galactokinase deficiency
UDP galactose epimerase deficiency
Hereditary fructose intolerance
Disorders of carbohydrate production or utilization
Disorders of glycogenolysis (glycogen storage diseases, GSD)
Liver glycogen synthase deficiency (GSD 0)
Glucose-6-phosphatase deficiency (GSD I; von Gierke disease)
Lysosomal acid maltase deficiency (GSD II; Pompe disease)
Glycogen debrancher deficiency (GSD III; Cori/Forbes disease)
Glycogen branching enzyme deficiency (GSD IV; Andersen disease)
Muscle phosphorylase deficiency (GSD V; McArdle disease)
Liver phosphorylase deficiency (GSD VI; Hers disease)
Muscle phosphofructokinase deficiency (GSD VII; Tarui disease)
Phosphoglycerate kinase and phosphoglycerate mutase deficiency
Phosphorylase b kinase deficiency
Disorders of gluconeogenesis
Fructose 1,6-biphosphatase deficiency
Pyruvate carboxylase deficiency
Phosphoenolpyruvate carboxykinase (PEPCK) deficiency
Pyruvate dehydrogenase deficiency

UDP: uridine diphosphate.

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Selected mitochondrial disorders

Pyruvate carboxylase deficiency
Phosphoenopyruvate carboxylase deficiency
Pyruvate dehydrogenase complex deficiency
Kearns-Sayre syndrome
Mitochondrial encephalopathy lactic acidosis and stroke-like episodes (MELAS)
Myoclonic epilepsy, ragged red fiber disease (MERRF)
Freidrich ataxia
Pearson syndrome
Succinate dehydrogenase deficiency
Cytochrome C oxidase (COX) deficiency
Mitochondrial neurogastrointestinal encephalopathy syndrome (MNGIE)
Mitochondrial DNA depletion syndromes

Graphic 69937 Version 4.0

Peroxisomal disorders

Disorders of peroxisome biogenesis
Zellweger syndrome
Neonatal adrenoleukodystrophy (NALD)
Infantile Refsum disease
Rhizomelic chondrodysplasia punctata type 1
Disorders with deficiency of a single peroxisomal enzyme
X-linked adrenoleukodystrophy, including adrenomyeloneuropathy (AMN) due to deficiency of adrenoleukodystrophy protein (ALDP)
Refsum disease (phytanoyl CoA hydroxylase deficiency)
Pseudo-NALD (acyl CoA oxidase deficiency)
D-bifunctional enzyme deficiency
Dihydroxy-acetone phosphate acyltransferase (DHAP-AT) deficiency (RCDP type 2)
Alkyl-DHAP synthase deficiency (RCDP type 3)
2-methylacyl-CoA racemase deficiency
Acatalasemia (Catalase deficiency)
Hyperoxaluria type 1 (alanine glyoxylate aminotransferase deficiency)

RCDP: rhizomelic chondrodysplasia punctata; DHAP: dihydroxy-acetone phosphate.

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Selected lysosomal storage disorders

Mucopolysaccharidoses (MPS)
MPS I (Hurler, Hurler-Scheie, Scheie)
MPS II (Hunter)
MPS III (Sanfillippo)
MPS IV (Morquio)
MPS VI (Maroteaux-Lamy)
MPS VII (Sly)
MPS IX (Natowicz)
Sphingolipidoses
GM1 gangliosidosis
GM2 gangliosidosis type 1 (Tay-Sachs)
GM2 gangliosidosis type 2 (Sandhoff)
Fabry disease
Farber disease
Gaucher disease
Niemann-Pick disease
Krabbe disease
Metachromatic leukodystrophy
Oligosaccharidoses (glycoproteinoses)
Galactosialidosis
Fucosidosis types
Mannosidosis
Aspartylglucosaminuria
Mucolipidosis
Mucolipidosis type I (Sialidosis)
Mucolipidosis type II (I-cell)*
Mucolipidosis type III (pseudo-Hurler)*
Mucolopidosis type IV (Sialolipidosis)

* These disorders are caused by the same enzyme deficiency (N-acetylglucosamyl phosphotransferase).

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Disorders of fatty acid oxidation

Disorders
Carnitine uptake defect
Short chain acyl-CoA dehydrogenase (SCAD) deficiency
Short chain hydroxyacyl-CoA dehydrogenase (SCHAD) deficiency
Medium chain acyl-CoA dehydrogenase (MCAD) deficiency
Very long chain acyl-CoA dehydrogenase (VLCAD) deficiency
Long chain hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency
Trifunctional protein (TFP) deficiency
Carnitine palmitoyltransferase II (CPTII) deficiency
Carnitine-acylcarnitine translocase (CACT) deficiency
Carnitine palmitoyltransferase I (CPTI) deficiency
Multiple acyl-CoA dehydrogenase deficiency (glutaric acidemia type II)

List of common fatty acid oxidation disorders and associated acylcarnitine abnormalities. Note that the first newborn
disorders (eg, VLCAD and CPTII), further testing such as DNA mutation analysis may be required to establish a diagı