BOSTON MEDICAL CENTER HEALTHNet PLAN

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Pharmacy Policy

Urea Cycle Disorder Agents

Policy Number: 9.305
Version Number: 2.0
Version Effective Date: 9/1/2021

Product Applicability	☐ **All Plan+ Products**
Well Sense Health Plan ☐ New Hampshire Medicaid	**Boston Medical Center HealthNet Plan** ☒ MassHealth - MCO ☒ MassHealth - ACO ☐ Qualified Health Plans/ConnectorCare/Employer Choice Direct ☐ Senior Care Options

Note: Disclaimer and audit information is located at the end of this document.

Prior Authorization Policy

Products Affected:

- **Carbaglu (carglumic acid)**
- **Ravicti (glycerol phenylbutyrate)**

The Plan may authorize coverage of the above products for members meeting the following criteria:

Covered Use	All FDA approved indications not otherwise excluded
Exclusion Criteria	Use of Ravicti for diagnosis of N-acetylglutamate synthase (NAGS) deficiency Use of Ravicti for treatment of acute hyperammonemia.
Required Medical Information	**Carbaglu (carglumic acid)**: 1. Member has diagnosis of hepatic enzyme N-acetylglutamate synthase deficiency with hyperammonemia; **AND** • Diagnosis is confirmed by DNA mutation analysis (documentation required); **OR** • Member has hyperammonemia diagnosed with ammonia level above the upper limit of normal (lab records within past 3 months is required); **OR**

+ *Plan* refers to Boston Medical Center Health Plan, Inc. and its affiliates and subsidiaries offering health coverage plans to enrolled members. The Plan operates in Massachusetts under the trade name Boston Medical Center HealthNet Plan and in other states under the trade name Well Sense Health Plan.

	2. Member has diagnosis of hyperammonemia due to propionic acidemia (PA) or methylmalonic acidemia (MMA); **AND** • Plasma ammonia level is ≥ 50 micromol/L(lab records within last 7 days is required); **AND** • Carbaglu is prescribed in conjunction with other ammonia-lowering therapies (e.g intravenous glucose, insulin, L-carnitine, and protein restriction) **Ravicti (glycerol phenylbutyrate)** 1. A diagnosis of a urea cycle disorder (except NAGS) confirmed by enzyme analysis or DNA mutation analysis (documentation required); **AND** 2. Inadequate response, intolerance, or contraindication to sodium phenylbutyrate (Buphenyl®); **AND** 3. Ravicti is being used as an adjunct to at least **one** of the following therapies: a. Dietary protein restriction b. Dietary supplements (e.g., arginine, citrulline, essential amino acids, protein-free calorie supplements)
Prescriber Restriction	Prescribed by or in collaboration with a physician experienced in management of Urea cycle Disorder
Coverage Duration	Carbaglu: 1. NAGS deficiency: a. Diagnosis confirmed by genetic testing: Initial 12 months; reauthorization 12 months b. Diagnosis of hyperammonemia : Initial 3 months ; reauthorization 12 months 2. Propionic acidemia (PA) or methylmalonic acidemia (MMA): 7 days Ravicti: Initial and reauthorization: 12 months
Other criteria	Reauthorization: **Carbaglu (carglumic acid)** 1. For NAGS deficiency, member is responding positively to Carbaglu treatment as shown by normal or improved ammonia level (lab records within the past 6 months must be included) **Ravicti (glycerol phenylbutyrate)** **1.** Member is responding positively to Ravicti treatment as shown by normal or improved ammonia level (lab within the past 6 months must be included); **AND** 2. Member is actively on protein-restricted diet or is taking Ravicti in conjunction with dietary supplements (e.g., arginine, citrulline, essential amino acid, protein-free calorie supplements).

Clinical Background Information and References

1. Lee B. Urea cycle disorders: Management. Accessed Feb 2015 at www.uptodate.com
2. Product Information. Carbaglu (carglumic acid). Accredo Health Group, Inc. Memphis, Tennessee. August, 2013.
3. Product information. Ravicti (glycerol phenylbutyrate). Hyperion Therapeutics, Inc. Brisbane, CA. June 2015.

4. Ah Mew N, Lanpher BC, Gropman A, et al. Urea Cycle Disorders Overview. 2003 Apr 29 [Updated 2014 Sep 11]. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2015. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1217/

Original Approval Date	Original Effective Date	Policy Owner	Approved by
12/1/2020	1/1/2021	Pharmacy Services	Pharmacy & Therapeutics (P&T) Committee

Policy Revisions History			
Review Date	**Summary of Revisions**	**Revision Effective Date**	**Approved by**
12/1/2020	9.038 Urea Cycle Disorder policy retired, new policy created	1/1/2021	P&T Committee
5/13/2021	P&T Annual review: For Carbaglu-Added new FDA indication; Updated initial criteria to require documentation of enzyme/DNA analysis; Updated approval duration; Added prescriber restriction and updated reauthorization criteria. For Ravicti- Updated initial criteria to require DNA analysis; Updated reauth criteria; Updated approval duration and added prescriber restriction	9/1/2021	P&T Committee

Next Review Date

5/2022

Other Applicable Policies

Reference to Applicable Laws and Regulations, If Any

Disclaimer Information

Medical Policies are the Plan's guidelines for determining the medical necessity of certain services or supplies for purposes of determining coverage. These Policies may also describe when a service or supply is considered experimental or investigational, or cosmetic. In making coverage decisions, the Plan uses these guidelines and other Plan Policies, as well as the Member's benefit document, and when appropriate, coordinates with the Member's health care Providers to consider the individual Member's health care needs.

Plan Policies are developed in accordance with applicable state and federal laws and regulations, and accrediting organization standards (including NCQA). Medical Policies are also developed, as appropriate, with consideration of the

medical necessity definitions in various Plan products, review of current literature, consultation with practicing Providers in the Plan's service area who are medical experts in the particular field, and adherence to FDA and other government agency policies. Applicable state or federal mandates, as well as the Member's benefit document, take precedence over these guidelines. Policies are reviewed and updated on an annual basis, or more frequently as needed. Treating providers are solely responsible for the medical advice and treatment of Members.

The use of this Policy is neither a guarantee of payment nor a final prediction of how a specific claim(s) will be adjudicated. Reimbursement is based on many factors, including member eligibility and benefits on the date of service; medical necessity; utilization management guidelines (when applicable); coordination of benefits; adherence with applicable Plan policies and procedures; clinical coding criteria; claim editing logic; and the applicable Plan – Provider agreement.