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## DRUG POLICY

# Ravicti (glycerol phenylbutyrate)

### 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 APPLICATION

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 policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

### DESCRIPTION

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

#### FDA-Approved Indication

Ravicti is indicated for the chronic management of patients with urea cycle disorders (UCDs) who cannot be managed by dietary protein restriction and/or amino acid supplementation alone. Ravicti must be used with dietary protein restriction and, in some cases, dietary supplements.

#### Limitations of Use

- Ravicti is not indicated for treatment of acute hyperammonemia in patients with UCDs.
- Safety and efficacy for treatment of N-acetylglutamate synthase (NAGS) deficiency has not been established.

### POLICY

#### Documentation

- A. Initial Requests:
  1. Enzyme assay, biochemical, or genetic testing results supporting diagnosis; and
  2. Lab results documenting baseline plasma ammonia levels
- B. Continuation of therapy requests: lab results documenting a reduction in plasma ammonia levels from baseline

#### Criteria for Initial Approval

#### A. Urea Cycle Disorder (UCD)

Authorization of 12 months may be granted for chronic management of urea cycle disorder (UCD) when all of the following criteria are met:

1. Diagnosis was confirmed by enzymatic, biochemical, or genetic testing.
2. The medication is prescribed by, or in consultation with, a specialist in metabolic disorders
3. The member has elevated plasma ammonia levels at baseline
4. Member is unable to manage the disorder by dietary protein restriction and/or amino acid supplementation alone
5. The medication will be used along with a protein restricted diet and in some cases, dietary supplements (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements)
6. Member meets ANY of the following criteria:
  - a. Member has tried Buphenyl (sodium phenylbutyrate) and has experienced intolerance to Buphenyl (sodium phenylbutyrate) therapy.
  - b. Member has a comorbid condition that prohibits a trial of Buphenyl (sodium phenylbutyrate) due to its sodium content.
7. The medication is not being used for the treatment of acute hyperammonemia

#### Continuation of Therapy

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for chronic management of a urea cycle disorder (UCD), who are experiencing benefit from therapy as evidenced by a reduction in plasma ammonia levels from baseline.

#### Quantity Limit

19 grams (17.5 mL) per day

#### Dosing and Administration

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

### PROCEDURES AND BILLING CODES

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

- Code(s), if applicable

### REFERENCES

- Ravicti [package insert]. Lake Forest, IL: Horizon Pharma USA, Inc; November 2019.
- Mew NA, Lanpher BC. Urea Cycle Disorders Overview. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2017 [updated April 9, 2015]. Available at: <https://www.ncbi.nlm.nih.gov/books/NBK1217/?report=printable>.
- Häberle J, Boddaert N, Burlina A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders. Orphanet J Rare Dis. 2012;7:32.
- Diaz GA, Krivitzky LS, Mokhtarani M, et al. Ammonia control and neurocognitive outcome among urea cycle disorder patients treated with glycerol phenylbutyrate. Hepatology. 2013;57(6):2171-2179.
- Smith W, Diaz GA, Lichter-Konecki U, et al. Ammonia control in children ages 2 months through 5 years with urea cycle disorders: comparison of sodium phenylbutyrate and glycerol phenylbutyrate. J Pediatr. 2013;162(6):1228-1234.

\*Some content reprinted from CVSHealth

## POLICY HISTORY

**Policy #:** 05.02.04

**Policy Creation:** January 2017

**Reviewed:** February 2022

**Revised:** May 2021

**Current Effective Date:** August 8, 2021