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**Title:** Glycerol Phenylbutyrate (Ravicti®)

**Policy #:** Rx.01.135

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***Application of pharmacy policy is determined by benefits and contracts. Benefits may vary based on product line, group, or contract. Some medications may be subject to precertification, age, quantity, or formulary restrictions (ie limits on non-preferred drugs). Individual member benefits must be verified.***

***This pharmacy policy document describes the status of pharmaceutical information and/or technology at the time the document was developed. Since that time, new information relating to drug efficacy, interactions, contraindications, dosage, administration routes, safety, or FDA approval may have changed. This Pharmacy Policy will be regularly updated as scientific and medical literature becomes available. This information may include new FDA-approved indications, withdrawals, or other FDA alerts. This type of information is relevant not only when considering whether this policy should be updated, but also when applying it to current requests for coverage.***

***Members are advised to use participating pharmacies in order to receive the highest level of benefits.***

**Intent:**

The intent of this policy is to communicate the medical necessity criteria for **Glycerol phenylbutyrate (Ravicti®)** as provided under the member's prescription drug benefit.

**Description:**

**Urea Cycle disorders** are inherited deficiencies of enzymes or transporters necessary for the synthesis of urea from ammonia (NH<sub>3</sub>, NH<sub>4</sub><sup>+</sup>). Absence of these enzymes or transporters results in the accumulation of toxic levels of ammonia in the blood and brain of affected patients.

**Glycerol phenylbutyrate** binds to nitrogen waste product and allows excretion via the urine. Glycerol phenylbutyrate is a triglyceride containing 3 molecules of phenylbutyrate (PBA). Phenylacetate (PAA), the major metabolite of PBA, is the active moiety of glycerol phenylbutyrate. PAA conjugates with glutamine (which contains 2 molecules of nitrogen) via acetylation in the liver and kidneys to form phenylacetylglutamine (PAGN), which is excreted by the kidneys. On a molar basis, PAGN, like urea, contains 2 moles of nitrogen and provides an alternate vehicle for waste nitrogen excretion

**Glycerol phenylbutyrate (Ravicti®)** is indicated for long-term management of urea cycle disorder in patients who cannot be managed by dietary protein restriction and/or amino acid supplementation alone.

**Policy:**

Glycerol phenylbutyrate (Ravicti®) is approved when ALL of the following inclusion criteria are met:

1. Diagnosis of Urea Cycle Metabolism Disorder; and
2. Member cannot be managed by dietary protein restriction and/or amino acid supplementation alone; and
3. Inadequate response to or inability to tolerate sodium phenylbutyrate

4. Will be used concomitantly with dietary protein restriction and, in some cases, dietary supplements (e.g. essential amino acids, protein-free calorie supplements, arginine, etc.)

**Black Box Warning as shown in the drug Prescribing Information:**

None

**Guidelines:**

Refer to the specific manufacturer's prescribing information for administration and dosage details and any applicable Black Box warnings.

**BENEFIT APPLICATION**

Subject to the terms and conditions of the applicable benefit contract, the applicable drug(s) identified in this policy is (are) covered under the prescription drug benefits of the Company's products when the medical necessity criteria listed in this pharmacy policy are met. Any services that are experimental/investigational or cosmetic are benefit contract exclusions for all products of the Company.

**References:**

Ravicti® (glycerol phenylbuterate) [package insert]. Lake Forest, IL. Horizon Therapeutics, LLC. April 2017. Available at: [https://www.accessdata.fda.gov/drugsatfda\\_docs/label/2017/203284s005lbl.pdf#page=1](https://www.accessdata.fda.gov/drugsatfda_docs/label/2017/203284s005lbl.pdf#page=1). Accessed August 5, 2020.

Urea cycle disorders. Available at: <http://nordphysicianguides.org/urea-cycle-disorders/what-are-urea-cycle-disorders/>. Accessed August 5, 2020.

**Applicable Drugs:**

Inclusion of a drug in this table does not imply coverage. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

Brand name	Generic name
Ravicti®	Glycerol Phenylbutrate

**Cross References:**

Off Label Use Rx.01.33

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<b>Policy Version Number:</b>	8.00
<b>P&amp;T Approval Date:</b>	July 09, 2020
<b>Policy Effective Date:</b>	October 01, 2020
<b>Next Required Review Date:</b>	July 09, 2021

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The Policy Bulletins on this web site were developed to assist the Company in administering the provisions of the respective benefit programs, and do not constitute a contract. If you have coverage through the Company, please refer to your specific benefit program for the terms, conditions, limitations and exclusions of your coverage. Company does not provide health care services, medical advice or treatment, or guarantee the outcome or results of any medical services/treatments. The facility and professional providers are responsible for providing medical advice and treatment. Facility and professional providers are independent contractors and are not employees or agents of the

Company. If you have a specific medical condition, please consult with your doctor. The Company reserves the right at any time to change or update its Policy Bulletins.

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