

UnitedHealthcare Pharmacy  
Clinical Pharmacy Programs

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| Program Number    | 2025 P 2129-11   |
| Program           | Prior Authorization/Medical Necessity  |
| Medication        | Ravicti® *(glycerol phenylbutyrate oral liquid), glycerol phenylbutyrate oral liquid |
| P&T Approval Date | 7/2017, 7/2018, 2/2019, 2/2020, 2/2021, 2/2022, 2/2023, 2/2024, 2/2025, 12/2025      |
| Effective Date    | 5/1/2026   |

## 1. Background

Ravicti\* (glycerol phenylbutyrate) is a nitrogen-binding agent indicated for 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 (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements).

Ravicti\* is not indicated for treatment of acute hyperammonemia in patients with UCDs. The safety and efficacy for treatment in N-acetylglutamate synthase (NAGS) deficiency has not been established. <sup>1</sup>

Coverage for Ravicti\* will be provided for patients who meet the following criteria:

## 2. Coverage Criteria<sup>a</sup>:

### A. Initial Authorization

1. **Ravicti\*** will be approved based on **all** of the following criteria:

a. All of the following:

(1) Patient has a diagnosis of a urea cycle disorder (UCD)

-AND-

(2) Patient does not have N-acetylglutamate synthase (NAGS) deficiency

-AND-

(3) Patient has history of inadequate response to **one** of the following:

- (a) Dietary protein restriction
- (b) Amino acid supplementation

-AND-

(4) **One** of the following:

- (a) **Both** of the following

- i. History of failure to sodium phenylbutyrate (Buphenyl®)

-AND-

- ii. Submission of medical records (e.g., chart notes, laboratory values) documenting one of the following while on sodium phenylbutyrate

1. Fasting ammonia level > 0.5 ULN

-OR-

2. Any ammonia level [fasting/non-fasting] above the ULN

-OR-

- (b) **Both** of the following:

- i. History of intolerance to sodium phenylbutyrate oral tablets

-AND-

- ii. Submission of medical records (e.g., chart notes, prescription claims history) documenting trial of sodium phenylbutyrate oral tablets

-OR-

- (c) Submission of medical records (e.g. chart notes) documenting a contraindication to sodium phenylbutyrate

-OR-

- (d) **Both** of the following

- i. Patient is currently on Ravicti\* therapy

-AND-

- ii. Patient has not received a manufacturer supplied sample at no cost in prescriber office, or any form of assistance from the Horizon Therapeutics sponsored TranscendRare™ program (e.g., sample card which can be redeemed at a pharmacy for a free supply of medication) as a means to establish as a current user of Ravicti<sup>+</sup>

<sup>+</sup>Patients requesting initial authorization who were established on therapy via the receipt of a manufacturer supplied sample at no cost in the prescriber's office or any form of assistance from the Horizon Therapeutics sponsored TranscendRare™ program shall be required to meet initial authorization criteria as if patient were new to therapy.

-AND-

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

**Authorization will be issued for 12 months.**

**B. Reauthorization**

1. **Ravicti\*** will be approved based on **both** of the following criteria:

a. Documentation of positive clinical response to Ravicti\* therapy

**-AND-**

b. Patient is actively on dietary protein restriction and, in some cases, dietary supplements (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements)

**Authorization will be issued for 12 months.**

<sup>a</sup> State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

\*Brand Ravicti is typically excluded from coverage. Tried/Failed criteria may be in place. Please refer to plan specifics to determine exclusion status.

**3. Additional Clinical Rules:**

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place

**4. References:**

1. Ravicti<sup>®</sup> [package insert], Lake Forest, IL: Horizon Therapeutics, Inc.; September 2021.
2. Lee B. Urea Cycle Disorders: Management. In: UpToDate, Waltham, MA, 2024. Accessed December 23, 2024
3. Lee B., Diaz GA, Rhead W., et al. Blood ammonia and glutamine as predictors of hyperammonemic crises in patients with urea cycle disorder. *Genet Med.* 2015; 17(7):561-8.

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| Program               | Prior Authorization/Medical Necessity - Ravicti (glycerol phenylbutyrate oral liquid)   |
| <b>Change Control</b> |   |
| 7/2017                | New program   |
| 7/2018                | Annual review. Updated criteria regarding sodium phenylbutyrate intolerance specifying that patient experience intolerance to oral tablets prior to coverage for Ravicti. |

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|---------|--|
| 12/2018 | Administrative change to add statement regarding use of automated processes.   |
| 2/2019  | Updated background and criteria to align with updated indication allowing use in patients less than 2 months of age. |
| 2/2020  | Annual review with no change to clinical coverage. Updated reference.  |
| 2/2021  | Annual review with no change to clinical coverage. Updated references.   |
| 2/2022  | Annual review with no change to clinical coverage. Updated references.   |
| 2/2023  | Annual review with no change to clinical coverage. Updated references.   |
| 2/2024  | Annual review with no change to clinical coverage.   |
| 2/2025  | Annual review with no change to clinical coverage. Updated reference.  |
| 12/2025 | Updated program to note that brand Ravicti is typically excluded from coverage.                                      |