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Section:	Prescription Drugs	**Effective Date:**	April 1, 2025
Subsection:	Endocrine and Metabolic Drugs	**Original Policy Date:**	November 8, 2013
Subject:	Ravicti	**Page:**	1 of 4

Last Review Date: March 7, 2025

Ravicti

Description

Ravicti (glycerol phenylbutyrate)

Background
Urea cycle disorders (UCDs) are genetic disorders that involve deficiencies of specific enzymes involved in the urea cycle, a series of biochemical steps normally required to remove ammonia from the blood. When protein is absorbed and broken down by the body, it produces nitrogen as a waste product. The urea cycle removes nitrogen from the blood and converts it to urea, which is removed from the body through urine. In people with UCDs, nitrogen accumulates and remains in the body as ammonia, which can travel to the brain and cause brain damage, or coma (1).

Ravicti, a liquid taken three times a day with meals, helps dispose of ammonia in the body. It is intended for patients whose UCD cannot be managed by a protein-restricted diet or amino acid supplements alone. Ravicti must be used with a protein-restricted diet and, in some cases, dietary supplements (1).

Regulatory Status
FDA-approved indication: Ravicti is indicated for use as a nitrogen-binding agent for chronic management of patients with urea cycle disorders (UCDs) that 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) (1).

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 (1).

Related policies
Buphenyl

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Ravicti may be considered **medically necessary** if the conditions indicated below are met.

Ravicti may be considered **investigational** for all other indications.

Prior-Approval Requirements

Diagnosis

Patient must have the following:

Urea cycle disorders (UCDs)

AND ALL of the following:

1. Failure to control ammonia level with dietary restrictions and / or amino acid supplementation
2. Prescribing physician should be experienced in the management of UCDs
3. Must be used with dietary protein restrictions

Prior – Approval *Renewal* Requirements

Diagnosis

Patient must have the following:

Urea cycle disorders (UCDs)

AND the following:

1. Must be used with dietary protein restrictions

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 2 years

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary

Ravicti is indicated for use as a nitrogen-binding agent for chronic management of adult and pediatric patients with urea cycle disorders (UCDs) that 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. Safety and efficacy for treatment of *N*- acetylglutamate synthase (NAGS) deficiency has not been established (1).

Prior approval is required to ensure the safe, clinically appropriate, and cost-effective use of Ravicti while maintaining optimal therapeutic outcomes.

References

1. Ravicti [package insert]. Lake Forest, IL: Horizon Therapeutics, LLC.; September 2021.

Policy History

Date	Action
November 2013	Addition to PA
December 2013	Annual editorial review.
December 2014	Annual editorial review and reference update
June 2015	Annual editorial review and reference update
September 2016	Annual editorial review and reference update; Policy code changed from 5.08.29 to 5.30.29

June 2017	Change of the age requirement from 2 years to 2 months
September 2017	Annual review
November 2018	Annual review and reference update
January 2019	Removal of age requirement
March 2019	Annual review
February 2020	Changed approval duration from lifetime to 2 years
March 2020	Annual review
September 2021	Annual review
September 2022	Annual review and reference update
December 2022	Annual review
September 2023	Annual review
September 2024	Annual review
March 2025	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 7, 2025 and is effective on April 1, 2025.