AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Ravicti (glycerol phenylbutyrate)	Page:	1 of 2
Effective Date:	1/1/2026	Last Review Date:	4/4/2025

Applies to:	☒Illinois	☒Florida Kids	☐New Jersey
	☒Maryland	☒Pennsylvania Kids	☐Virginia

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Ravicti under the patient's prescription drug benefit.

Description:

Ravicti is indicated for use as a nitrogen-binding agent 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.

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.

All other indications are considered experimental/investigational and not medically necessary.

Applicable Drug List:

Non-Formulary: Ravicti

Policy/Guideline:

Documentation:
Submission of the following information is necessary to initiate the prior authorization review:
Initial Requests:

- Enzyme assay, biochemical, or genetic testing results supporting diagnosis; and
- Lab results documenting baseline plasma ammonia levels.

Continuation of therapy requests: lab results documenting a reduction in plasma ammonia levels from baseline.

Prescriber Specialties
This medication must be prescribed by or in consultation with a physician who specializes in the treatment of enzyme or metabolic disorders.

Coverage Criteria:

Urea Cycle Disorders (UCDs)

Authorization of 12 months may be granted for chronic management of a UCD when BOTH the following criteria are met:

- The diagnosis is confirmed by enzymatic, biochemical, or genetic testing.
- The member has elevated plasma ammonia levels at baseline)

Continuation of Therapy:

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in the coverage criteria section who are experiencing benefit from therapy as evidenced by a reduction in plasma ammonia levels from baseline.

Approval Duration and Quantity Restrictions:

Approval: 12 months

Quantity Level Limit: Reference Formulary for drug specific quantity level limits

References:

1. Ravicti [package insert]. Deerfield, IL: Horizon Therapeutics USA, Inc.; September 2021.
2. 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 June 22, 2017]. Available at: https://www.ncbi.nlm.nih.gov/books/NBK1217/?report=printable.
3. Häberle J, Boddaert N, Burlina A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders. J Inherit Metab Dis. 2019;42(6):1192-1230.
4. 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.
5. 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.