AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Ravicti (glycerol phenylbutyrate)	Page:	1 of 2
Effective Date:	3/14/2025	Last Review Date:	2/2025

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

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 (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements).

Limitations of Use:

- Ravicti is not indicated for the treatment of acute hyperammonemia in patients with UCDs because more rapidly acting interventions are essential to reduce plasma ammonia levels.
- The safety and efficacy of Ravicti for the 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.

COVERAGE CRITERIA:

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

- The diagnosis is confirmed by enzymatic, biochemical, or genetic testing.
- The member has elevated plasma ammonia levels at baseline.
- The member meets either of the following:
    - Has experienced an intolerance to sodium phenylbutyrate therapy
    - Has not tried sodium phenylbutyrate because of a comorbid condition that prohibits a trial due to its sodium content (e.g., heart failure, hypertension, renal impairment, edema)

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 April 9, 2015]. 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. Sun A, Crombez EA, Wong D. Arginase Deficiency. 2004 Oct 21 [Updated 2020 May 28]. In: Adam MP, Ardinger HH, Pagon RA, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2021. Available from: https://www.ncbi.nlm.nih.gov/books/NBK1159/