

Effective Date: 7/1/2020
Last Reviewed: 4/2020, 2/2021, 2/2022, 7/2023, 4/2024, 4/2025, 1/2026
Scope: Medicaid

Glycerol phenylbutyrate oral liquid (generic Ravicti)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met, and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication

Glycerol phenylbutyrate oral liquid is indicated for the chronic management of patients with urea cycle disorders (UCDs) who cannot be managed by dietary protein restriction and/or amino acid supplementation alone. Glycerol phenylbutyrate oral liquid must be used with dietary protein restriction and, in some cases, dietary supplements.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR APPROVAL

Urea Cycle Disorder (UCD) Chronic Management

An authorization of 6 months may be granted for chronic management of urea cycle disorder (UCD) when the following criteria are met:

1. The diagnosis is confirmed and documented by enzymatic, biochemical, or genetic testing.
2. Documentation that the member has elevated plasma ammonia levels at baseline, evidenced by a lab value of a fasting plasma ammonia level > 0.5 upper limit of normal OR a glutamine level that is $> 1,000$ $\mu\text{mol/L}$.
3. Documentation that the member must have had an inadequate response to and continue to be on a protein restricted diet or amino acid supplementation.
4. Member has documentation of an inadequate response to or inability to tolerate one of the following drugs:
 - a. Sodium phenylbutyrate tablets OR powder
 - b. Pheburane
 - c. Olpruva (if member weighs 20kg or greater and has a body surface area (BSA) of 1.2 m^2 or greater)
5. Glycerol phenylbutyrate is not being used for the management of acute hyperammonemia.
6. The prescribed dose does not exceed 19 grams per day (17.5 ml per day).

III. CONTINUATION OF THERAPY

If member has not been approved for this drug by Neighborhood in the past, clinician must submit documentation that initial criteria is met. Authorization of 6 months may be granted for continued

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treatment in members requesting reauthorization for chronic management of a urea cycle disorder (UCD), who are experiencing benefit from therapy as evidenced by a reduction in plasma ammonia levels from baseline (documentation provided).

IV. QUANTITY LIMIT

- Glycerol phenylbutyrate 1.1gm/ml: 17.5 ml per day (525ml per 30 days)

V. REFERENCES

1. Glycerol phenylbutyrate oral liquid. Lupin Pharmaceuticals, Inc. Naples, FL. FDA Package Insert. May 2025.
2. Ravicti (glycerol phenylbutyrate). Horizon Therapeutics. Lake Forest, IL. FDA Package Insert. September 2021.