PREFERRED SPECIALTY MANAGEMENT POLICY

POLICY: Metabolic Disorders – Phenylbutyrate Products Preferred Specialty Management Policy

- Buphenyl® (sodium phenylbutyrate tablets and powder for oral solution – Horizon, generic)
- Olpruva® (sodium phenylbutyrate for oral suspension – Acer)
- Pheburane® (sodium phenylbutyrate oral pellets – Medunik)
- Ravicti® (glycerol phenylbutyrate oral liquid – Horizon)

REVIEW DATE: 10/18/2023; selected revision: 02/21/2024

OVERVIEW

Phenylbutyrate products are indicated in combination with dietary management for treatment of **urea cycle disorders (UCDs)**.

- **Sodium phenylbutyrate** products are indicated as adjunctive therapy in the chronic management of adult and pediatric patients with UCDs involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS).[1-3]
  - **Buphenyl** and **Pheburane** can be administered orally in pediatric patients weighing less than 20 kg.
  - Buphenyl powder is compatible with feeding tube administration.
  - **Olpruva** is indicated for use in patients weighing $\geq$ 20 kg and with a body surface area of $\geq$ 1.2 m^2.

  Limitation of use: Sodium phenylbutyrate products are not indicated for the treatment of acute hyperammonemia, which can be a life-threatening medical emergency that requires rapid acting interventions to reduce plasma ammonia levels.
- **Ravicti** is indicated for the chronic management of patients with UCDs that cannot be managed by dietary protein restriction and/or amino acid supplementation alone.[4]

  Limitation of use: Ravicti is not indicated for treatment of acute hyperammonemia in patients with UCDs. Safety and efficacy for treatment of N-acetylglutamate synthetase deficiency has not been established.

POLICY STATEMENT

This Preferred Specialty Management program has been developed to encourage the use of Preferred Products. For all medications (Preferred and Non-Preferred), the patient is required to meet the respective standard *Prior Authorization Policy* criteria. The program also directs the patient to try one Preferred Product prior to the approval of a Non-Preferred Product. Requests for Non-Preferred Products will also be reviewed using the exception criteria (below). All approvals are provided for the duration noted below. If the patient meets the standard *Metabolic Disorders – Phenylbutyrate Products Prior Authorization Policy* criteria, but has not tried a Preferred Product, a review will be offered for the Preferred Product using the respective standard *Prior Authorization Policy* criteria.

Documentation: Documentation will be required where noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes, prescription claims records, and prescription receipts.

Automation: None.

Preferred Products:	generic sodium phenylbutyrate (tablets or powder), Pheburane
Non-Preferred Products:	Brand Buphenyl (tablets or powder), Olpruva, Ravicti

RECOMMENDED EXCEPTION CRITERIA

REFERENCES

1. Buphenyl® tablets and powder for oral solution [prescribing information]. Lake Forest, IL: Horizon; July 2022.
2. Olpruva® oral powder for suspension [prescribing information]. Newton, MA: Acer; December 2022.
3. Pheburane® oral pellets [prescribing information]. Bryn Mawr, PA: Medunik; June 2022.
4. Ravicti® oral liquid [prescribing information]. Lake Forest, IL: Horizon; September 2021.