

Applicable Drugs: Buphenyl (sodium phenylbutyrate), Olpruva (sodium phenylbutyrate), Pheburane (sodium phenylbutyrate), Ravicti (glycerol phenylbutyrate)

Preferred: Glycerol phenylbutyrate (generic), Sodium phenylbutyrate (generic)

Non-preferred: Buphenyl (sodium phenylbutyrate), Olpruva (sodium phenylbutyrate), Pheburane (sodium phenylbutyrate), Ravicti (glycerol phenylbutyrate)

Date of Origin: 5/7/2015

Date Last Reviewed / Revised: 3/13/2026

PRIOR AUTHORIZATION CRITERIA

(May be considered medically necessary when criteria I through IV are met)

- I. Documented diagnosis of urea cycle disorders (UCDs) and meets all the following criteria (A through C):
 - A. Biochemical, enzymatic, or molecular genetic analysis confirming diagnosis of UCD.
 - B. Documentation that the patient's condition is not adequately managed with dietary protein restriction and/or amino acid supplementation alone.
 - C. Documented attestation that phenylbutyrate will be used concomitantly with dietary protein restriction.
- II. Treatment must be prescribed by or in consultation with a physician experienced in the management of UCDs.
- III. Request is for a medication with the appropriate FDA labeling, or its use is supported by current clinical practice guidelines.
- IV. Refer to the plan document for the list of preferred products. If the requested agent is not listed as a preferred product, must have documented treatment failure or contraindication to the preferred product(s).

EXCLUSION CRITERIA

- Treatment of acute hyperammonemia.
- Treatment of N-acetylglutamate synthase (NAGS) deficiency.

OTHER CRITERIA

- Documentation of failure to sodium phenylbutyrate due to reasons such as "bad taste" or "taste aversion" will only be allowed for patients who are toddlers (2 to 3 years old), preschoolers (3 to 5 years old), and middle childhood (6 to 11 years old). Patients who are young teens (12 to 14 years old), teenagers (15 to 17 years old), and adults (18 years and older), must use sodium phenylbutyrate powder, suspension (compounded with Ora-Plus

and Ora-Sweet or Ora-Sweet SF), or tablets. Extemporaneously compounded suspensions of sodium phenylbutyrate, 200 mg/mL, in a 1:1 mixture of Ora-Plus and Ora-Sweet or Ora-Sweet SF are stable for at least 90 days when stored in 2-oz amber plastic bottles at room temperature.

QUANTITY / DAYS SUPPLY RESTRICTIONS

The quantity is limited to a 30-day supply per fill. Refer to FDA labeling or appropriate clinical guidelines for dosing regimen.

APPROVAL LENGTH

- **Authorization:** 1 year
- **Re-Authorization:** 1 year. An updated letter of medical necessity or progress notes showing current medical necessity criteria are met, the medication is effective, and the patient is actively on dietary protein restriction.

APPENDIX

- Urea cycle disorders (UCDs) include deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), and argininosuccinic acid synthetase (AS).

REFERENCES

1. Buphenyl. Prescribing Information. Horizon Therapeutics USA; 2023. Accessed March 13, 2026. https://www.pi.amgen.com/-/media/Project/Amgen/Repository/pi-amgen-com/buphenyl/buphenyl_fpi_english.pdf
2. Olpruva. Prescribing Information. Acer Therapeutics; 2025. Accessed March 13, 2026. <https://olpruva.com/documents/OLPRUVA-PI-10-2025.pdf>
3. Pheburane. Prescribing Information. Lucane Pharma; 2023. Accessed March 13, 2026. <https://files.medunik.com/usa/pheburane/prescribing-information.pdf>
4. Ravicti. Prescribing Information. Horizon Therapeutics USA; 2025. Accessed March 13, 2026. https://www.pi.amgen.com/-/media/Project/Amgen/Repository/pi-amgen-com/ravicti/ravicti_fpi_english.pdf
5. Häberle J, Burlina A, Chakrapani A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders: First revision. *Journal of Inherited Metabolic Disease*. 2019;42(6):1192-1230. doi:10.1002/jimd.12100
6. Caruthers RL, Johnson CE. Stability of extemporaneously prepared sodium phenylbutyrate oral suspensions. *American Journal of Health-System Pharmacy*. 2007;64(14):1513-1515. doi:10.2146/ajhp060450

DISCLAIMER: Medication Policies are developed to help ensure safe, effective and appropriate use of selected medications. They offer a guide to coverage and are not intended to dictate to providers how to practice medicine. Refer to Plan for individual adoption of specific Medication Policies. Providers are expected to exercise their medical judgement in providing the most appropriate care for their patients.