

Generic Name: Sodium Phenylbutyrate, glycerol phenylbutyrate

Therapeutic Class or Brand Name:

Phenylbutyrates

Applicable Drugs (if Therapeutic Class):

Sodium Phenylbutyrate (generic) powder and Buphenyl® (sodium phenylbutyrate) tablets, Ravicti® (glycerol phenylbutyrate)

Preferred: Sodium Phenylbutyrate (generic) powder and Buphenyl® (sodium phenylbutyrate) tablets.

Non-preferred: Ravicti® (glycerol phenylbutyrate).

Date of Origin: 5/7/2015

Date Last Reviewed / Revised: 11/18/2024

PRIOR AUTHORIZATION CRITERIA

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

- I. Documentation to support the diagnosis of urea cycle disorders (UCDs).
- II. Documentation that the patient's condition is not adequately managed with dietary protein restriction and/or amino acid supplementation alone.
- III. Documentation showing phenylbutyrate will be used concomitantly with dietary protein restriction.
- IV. Minimum age requirement:
 - A. Ravicti®: 2 months old.
- V. Treatment must be prescribed by or in consultation with a physician experienced in the management of UCDs.
- VI. Request is for a medication with the appropriate FDA labeling, or its use is supported by current clinical practice guidelines.
- VII. 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

- Documented diagnosis of acute hyperammonemia.
- Ravicti®:
 - Documented diagnosis 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.³

QUANTITY / DAYS SUPPLY RESTRICTIONS

- Sodium Phenylbutyrate (generic), Buphenyl®: Quantities of up to 600 grams (1200 tablets) per 30 days.
- Ravicti®: Quantities of up to 525 mLs per 30 days.

APPROVAL LENGTH

- **Authorization:** 1 year
- **Re-Authorization:** 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

Conversion Information		
Total daily dosage of sodium phenylbutyrate tabs(g)	Equals	$\frac{\text{Total daily dosage of Ravicti® (mL)}}{0.86}$
Total daily dosage of Ravicti® (mL)	Equals	total daily dosage of sodium phenylbutyrate tabs (g) x 0.86
Total daily dosage of sodium phenylbutyrate powder (g)	Equals	$\frac{\text{total daily dosage of Ravicti® (mL)}}{0.81}$
Total daily dosage of Ravicti® (mL)	Equals	total daily dosage of sodium phenylbutyrate powder(g) x 0.81

REFERENCES

1. Buphenyl. Prescribing Information. Horizon Therapeutics USA, Inc.; 2023. Accessed September 10, 2024. <https://www.hzndocs.com/BUPHENYL-Prescribing-Information.pdf>.
2. Ravicti. Prescribing Information. Horizon Therapeutics USA Inc.; 2021. Accessed September 10, 2024. <https://www.hzndocs.com/RAVICTI-Prescribing-Information.PDF>.
3. Caruthers RL, Johnson CE 2007. Stability of extemporaneously prepared sodium phenylbutyrate oral suspensions. Am J Health Syst Pharm 64(14):1513-1515.

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.