MEDICATION POLICY:
Phenylbutyrates

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: 10/13/2022

PRIOR AUTHORIZATION CRITERIA

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

I. Documentation to support a 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. Treatment is prescribed by or in consultation with a provider experienced in the management of metabolic disorders.

V. If request is for Ravicti ®, there must be documented treatment failure (see under Other Criteria) or contraindication to sodium phenylbutyrate.

VI. Request is for a medication with the appropriate FDA labeling, or its use is supported by current clinical practice guidelines.

VII. Refer to plan document for the list of preferred products. If requested agent is not listed as a preferred product, must have a documented failure, intolerance, or contraindication to a 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:** 1 year, with an updated letter of medical necessity showing maintenance or improvement on the medication, 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 (sodium phenylbutyrate). Prescribing information. Horizon Therapeutics USA, Inc; July 2022. Accessed October 13, 2022. https://www.hzndocs.com/BUPHENYL-Prescribing-Information.pdf.
2. Ravicti®. Prescribing information. Horizon Therapeutics USA Inc; September 2021. Accessed October 13, 2022. 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. 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.