MEDICATION POLICY

Generic Name: Phenylbutyrates

Therapeutic Class or Brand Name: Phenylbutyrates

Applicable Drugs (if Therapeutic Class)**:**

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

Non-Preferred: Ravicti® (glycerol phenylbutyrate).

Date of Origin: 5/7/15 **Date Last Reviewed/Revised:** 1/28/17

GPI Code: 3090803000, 3090806000

Prior Authorization Criteria (may be considered medically necessary when criteria I through V are met):

I. Documented diagnosis of the chronic management of patients with urea cycle disorders (UCDs).

II. Documentation that the patient's condition has not been able to be managed by dietary protein restriction and/or amino acid supplementation alone.

III. Documentation that Phenylbutyrate will be combined with dietary protein restriction.

IV. Prescriber is a physician experienced in the management of UCDs.

V. If request is for Ravicti®, must also meet both of criteria A AND B below:

A. Documented failure (see under Other Criteria) or contraindication to sodium phenylbutyrate.

B. Minimum age requirement: 2 years old.

Exclusion Criteria:

- Documented diagnosis of acute hyperammonemia.
- Ravicti®:
  - Documented diagnosis of N-acetylglutamate synthase (NAGS) deficiency.
  - Patients less than 2 months of age.

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 and that the medication is effective.

Appendix:

Conversion Information

Total daily dosage of sodium phenylbutyrate (g) = $\frac{\text{total daily dosage of Ravicti® (mL)}}{0.86}$

Total daily dosage of Ravicti® (mL) = total daily dosage of sodium phenylbutyrate (g) x 0.86

References:

1. http://www.horizonpharma.com/wp-content/uploads/2016/06/BUPHENYL_PI_April-2016.pdf.
2. https://hznp.azureedge.net/public/ravicti_1-1g-ml_vial_pi_effective_us.PDF.
3. Caruthers RL, Johnson CE 2007. Stability of extemporaneously prepared sodium phenylbutyrate oral suspensions. Am J Health Syst Pharm 64(14):1513-1515.
4. http://www.cdc.gov/ncbddd/childdevelopment/positiveparenting/adolescence.html.
5. Medi-Span.
6. http://blue.regence.com/trgmedpol/drugs/dru323.pdf.
7. http://blue.regence.com/trgmedpol/drugs/dru312.pdf.

MEDICATION POLICY

Historical Tracking Of Changes Made To Policy	
1/28/2017	1. **Updated** "http://www.horizonpharma.com/pdf/BUPHENYL_PI-MedGuide_Feb2014.pdf" **to** "http://www.horizonpharma.com/wp-content/uploads/2016/06/BUPHENYL_PI_April-2016.pdf" **and** "http://www.ravicti.com/sites/default/files/RAVICTI_Prescribing_Information_Updated.pdf" **to** "https://hznp.azureedge.net/public/ravicti_1-1g-ml_vial_pi_effective_us.PDF" **under References**.
11/19/2015	1. **Changed** "A. Documented failure, intolerance, or contraindication to Buphenyl® (sodium phenylbutyrate)." **to** "A. Documented failure (see under Other Criteria) or contraindication to sodium phenylbutyrate." under "V. If request is for Ravicti®, must also meet both of criteria A AND B below:" **under Prior Authorization Criteria**. 2. **Added** "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]" **under Other Criteria**. 3. **Added the following table under Appendix:** **Conversion Information** Total daily dosage of sodium phenylbutyrate (g) = $\frac{\text{total daily dosage of Ravicti® (mL)}}{0.86}$ Total daily dosage of Ravicti® (mL) = total daily dosage of sodium phenylbutyrate (g) x 0.86 4. **Added** "3. Caruthers RL, Johnson CE 2007. Stability of extemporaneously prepared sodium phenylbutyrate oral suspensions. Am J Health Syst Pharm 64(14):1513-1515" **and** "4. http://www.cdc.gov/ncbddd/childdevelopment/positiveparenting/adolescence.html" **under References**.