Louisiana Medicaid
Urea Cycle Disorder Agents

The *Louisiana Uniform Prescription Drug Prior Authorization Form* should be utilized to request clinical authorization for sodium phenylbutyrate (Buphenyl®), carglumic acid (Carbaglu®), and glycerol phenylbutyrate (Ravicti®).

Additional Point-of-Sale edits may apply.

Sodium Phenylbutyrate (Buphenyl®)

Approval Criteria

- The recipient has a diagnosis of urea cycle disorder involving deficiencies of:
  - Carbamylphosphate synthetase (CPS); **OR**
  - Ornithine transcarbamylase (OTC); **OR**
  - Argininosuccinic acid synthetase (AS); **AND**
- The prescriber is, or it is **stated on the request** that the prescriber has consulted with, a metabolic disease/medical genetic specialist; **AND**
- By submitting the authorization request, the prescriber attests to the following:
  - Buphenyl® will be combined with dietary protein restriction and, if appropriate, essential amino acid supplementation; **AND**
  - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
  - The maximum total daily dosage does not exceed 20 grams per day; **AND**
  - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
  - The recipient has no concomitant drug therapies or disease states that limit the use of the requested medication.

Reauthorization Criteria

- The recipient continues to meet initial approval criteria; **AND**
- The prescriber **states on the request** that there is evidence of a positive response to treatment.

*******Duration of initial and reauthorization approval**: 12 months*****

Carglumic Acid (Carbaglu®)

Approval Criteria

- The recipient has a diagnosis of N-acetylglutamate synthase (NAGS) deficiency, with **ONE** of the following conditions **stated on the request**:
    - Acute hyperammonemia; **OR**
    - Chronic hyperammonemia; **AND**
- The prescriber is, or it is **stated on the request** that the prescriber has consulted with, a metabolic disease/medical genetic specialist; **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - During acute hyperammonemic episodes, Carbaglu® will be used concomitantly with other ammonia lowering therapies, such as alternate pathway medications, hemodialysis, and dietary protein restriction; **AND**
    - During maintenance therapy, Carbaglu® will be used concomitantly with other ammonia lowering therapies and protein restriction, if appropriate based on plasma ammonia levels; **AND**
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - Dosing does not exceed the following:
        - For acute hyperammonemia, the initial dose does not exceed 250mg/kg/day; **OR**
        - For chronic hyperammonemia, the maintenance dose does not exceed 100mg/kg/day; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of the requested medication.

Reauthorization Criteria

- The recipient continues to meet initial approval criteria; **AND**
- The prescriber **states on the request** that there is evidence of a positive response to treatment.

*******Duration of initial and reauthorization approval**: 12 months*****

<u>Glycerol phenylbutyrate (Ravicti®)</u>
Approval Criteria

- Both of the following are true and **stated on the request:**
    - The recipient has a diagnosis of urea cycle disorder (UCD); **AND**
    - The recipient does **NOT** have N-acetylglutamate synthase (NAGS) deficiency; **AND**
- The prescriber is, or it is **stated on the request** that the prescriber has consulted with, a metabolic disease/medical genetic specialist; **AND**
- By submitting the authorization request, the prescriber attests to the following:

- Ravicti® will **NOT** be used for the treatment of **acute** hyperammonemia in recipients with UCD; **AND**
- Ravicti® will be used with dietary protein restriction and, if appropriate, dietary supplements; **AND**
- The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
- The maximum total daily dosage does not exceed 19 grams per day; **AND**
- All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
- The recipient has no concomitant drug therapies or disease states that limit the use of the requested medication.

Reauthorization Criteria

- The recipient continues to meet initial approval criteria; **AND**
- The prescriber **states on the request** that there is evidence of a positive response to treatment.

Duration of initial and reauthorization approval**: 12 months**

References

Buphenyl (sodium phenylbutyrate) [package insert]. Lake Forest, IL: Horizon Pharma USA, Inc; March 2018. https://www.hzndocs.com/BUPHENYL-Prescribing-Information.pdf

Carbaglu (carglumic acid) [package insert]. Lebanon, NJ: Recordati Rare Diseases Inc; December 2019. https://www.carbaglu.net/wp-content/uploads/2020/01/carbaglu-prescribing-information.pdf

Ravicti (glycerol phenylbutyrate) [package insert]. Lake Forest, IL: Horizon Therapeutics USA, Inc; November 2019. https://www.hzndocs.com/RAVICTI-Prescribing-Information.PDF

Revision	Date
Policy created.	January 2020
Policy implemented.	May 2020