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®, Pheburane®), carglumic acid (Carbaglu®), and glycerol phenylbutyrate (Ravicti®).

Additional Point-of-Sale edits may apply.

*These agents may have **Black Box Warnings** and/or may be subject to **Risk Evaluation and Mitigation Strategy (REMS)** under FDA safety regulations. Please refer to individual prescribing information for details.*

Sodium Phenylbutyrate (Buphenyl®, Pheburane®)
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 medication is prescribed by, or the request states that this medication is being prescribed in consultation with, a metabolic disease/medical genetic specialist; **AND**
- By submitting the authorization request, the prescriber attests to the following:
  - Sodium phenylbutyrate ~~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 and will not receive the requested medication in combination with any medication that is contraindicated or not recommended per FDA labeling.

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 **ONE** of the following diagnoses which is **stated on the request**:
    - Acute or chronic hyperammonemia due to N-acetylglutamate Synthase (NAGS) Deficiency; **OR**
    - Acute hyperammonemia due to propionic acidemia (PA) or methylmalonic acidemia (MMA); **AND**
- The medication is prescribed by, or the request states that this medication is being prescribed in consultation 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 due to NAGS deficiency, the initial dose does not exceed 250mg/kg/day; **OR**
        - For chronic hyperammonemia due to NAGS deficiency, the maintenance dose does not exceed 100mg/kg/day; **OR**
        - For acute hyperammonemia due to PA or MMA, the daily dose does not exceed 3.3 g/m^2/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 and will not receive the requested medication in combination with any medication that is contraindicated or not recommended per FDA labeling.

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

Glycerol phenylbutyrate (Ravicti®)

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 medication is prescribed by, or the request states that this medication is being prescribed in consultation 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 and will not receive the requested medication in combination with any medication that is contraindicated or not recommended per FDA labeling.

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; JulyFebruary 20220. https://www.hzndocs.com/BUPHENYL-Prescribing-Information.pdf

Carbaglu (carglumic acid) [package insert]. Lebanon, NJ: Recordati Rare Diseases Inc; January 2021. https://www.carbaglu.net/wp-content/uploads/2020/01/carbaglu-prescribing-information.pdf

Pheburane (sodium phenylbutyrate) [package insert]. Bryn Mawr, PA: Medunik USA, Inc; June 2022. https://files.medunik.com/usa/pheburane/prescribing-information.pdf

Ravicti (glycerol phenylbutyrate) [package insert]. Lake Forest, IL: Horizon Therapeutics USA, Inc; September November 20211219. https://www.hzndocs.com/RAVICTI-Prescribing-Information.PDF

Revision / Date	Implementation Date
Policy created / January 2020	May 2020
Updated diagnoses and maximum dose for Carbaglu®, updated references / February 2021	July 2021
Added Phenburane®, updated references / September 2022	January 2023