

## PHENYBUTYRATE AGENTS

### Applies To:

sodium phenylbutyrate 500 MG tab  
 BUPHENYL 500 MG TAB  
 sodium phenylbutyrate 3 GM/TSP powder  
 BUPHENYL 3 GM/TSP POWDER  
 RAVICTI (glycerol phenylbutyrate) 1.1 GM/ML LIQUID  
 PHEBURANE 483 MG/GM PELLETT  
 Olpurva

### Diagnosis Considered for Coverage:

- Chronic Hyperammonemia due to a chronic urea cycle disorder (UCD) involving deficiencies of:
  - Argininosuccinic Acid Synthetase (AAS)
  - Carbamyl Phosphate Synthetase (CPS)
  - Ornithine Transcarbamylase (OTC)

### Coverage Criteria:

#### For diagnosis listed above:

- Does not exceed FDA label maximum, **and**
- Confirmation of a urea cycle disorder by one of the following:
 

- Screening the urine and blood for abnormal metabolites (substance produced by metabolism), and high ammonia levels
  - A liver biopsy can be done to confirm the diagnosis since it can show low levels of enzyme activity
  - Genetic tests also can be done to show whether there is a problem with one of the genes needed to break down proteins of the urea cycle to identify the particular type of urea cycle disorder
  - An MRI or CT scan may be done to see if there is swelling of the brain caused by the ammonia in the blood

### Coverage Duration: one year

#### References:

1. Buphenyl (sodium phenylbutyrate). Prescribing Information. Ucylyd Pharma Inc., Scottsdale, AZ. 2009.
2. Ravicti. Prescribing Information. Horizon Therapeutics USA, Lake Forest, IL. 2021.
3. Pheburane. Prescribing Information. Medunik USA Inc. 2022.
4. Olpruva. Prescribing Information. Acer Therapeutics Inc. 2022.

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