



## Carbaglu Prior Authorization

<b>Drug(s) Applied:</b>	<b>Carbaglu</b> (carglumic acid) soluble tablets
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### Criteria:

Drug(s) Applied will be approved when the requested medication is being used for an FDA approved indication and all of the following criteria are met:

#### I. Initial Therapy Criteria

##### A. N-acetylglutamate synthase (NAGS) deficiency

1. Diagnosis of hyperammonemia with elevated ammonia levels according to the patient's age [Neonate: plasma ammonia level 150  $\mu\text{mol/L}$  ( $> 260 \mu\text{g/dl}$ ) or higher; Older child or adult: plasma ammonia level  $> 100 \mu\text{mol/L}$  ( $175 \mu\text{g/dl}$ )] **and**
2. Diagnosis of NAGS deficiency confirmed by enzyme analysis (via liver biopsy) or genetic testing **and**
3. Patient is unable to maintain a plasma ammonia level within the normal range with the use of a protein restricted diet and, when clinically appropriate, essential amino acid supplementation **and**
4. Prescriber is a specialist, or has consulted with a specialist, in the area of the patient's diagnosis (e.g., nephrologist, metabolic disorders)

**Approval Duration:** 12 months

##### B. Methylmalonic acidemia (MMA) or propionic acidemia (PA)

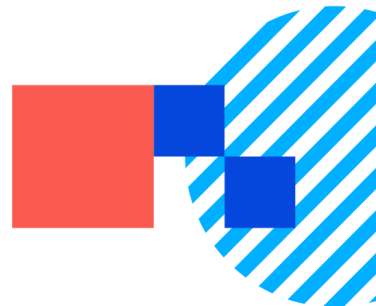
1. Requested drug will be used as adjunctive therapy to standard of care for the treatment of acute hyperammonemia **and**
2. Diagnosis of MMA or PA confirmed by enzyme and/or molecular genetic analyses **and**
3. Patient was hospitalized with a plasma ammonia level  $\geq 50 \mu\text{mol/L}$  **and**
4. Prescriber is a specialist, or has consulted with a specialist, in the area of the patient's diagnosis (e.g., nephrologist, metabolic disorders)

**Approval Duration:** 1 month

#### II. Continued Therapy Criteria

##### A. N-acetylglutamate synthase (NAGS) deficiency

1. Patient has been previously approved for the requested agent through the plan's Prior Authorization process or meets initial therapy criteria **and**
2. Patient has had clinical benefit with the requested agent as evidenced by



plasma ammonia level within the normal range **and**

3. Prescriber is a specialist, or has consulted with a specialist, in the area of the patient's diagnosis (e.g., nephrologist, metabolic disorders)

**Approval Duration:** 12 months

**B. Methylmalonic acidemia (MMA) or propionic acidemia (PA)**

1. Review under Initial Therapy Criteria for each instance

**Policy Owned by:** PBM team