

## PHARMACY POLICY STATEMENT

### Marketplace

<b>DRUG NAME</b>	<b>Carbaglu (carglumic acid)</b>
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
STATUS	Prior Authorization Required

Carbaglu is a carbamoyl phosphate synthetase 1 (CPS 1) activator initially approved by the FDA in 2010. It is indicated in pediatric and adult patients as adjunctive therapy to standard of care for the treatment of acute hyperammonemia due to N-acetylglutamate synthase (NAGS) deficiency. Carbaglu is also indicated for maintenance therapy for the treatment of chronic hyperammonemia due to NAGS deficiency. Adults and pediatric patients may also use Carbaglu for adjunctive therapy to standard of care for the treatment of acute hyperammonemia due to propionic acidemia (PA) or methylmalonic acidemia (MMA).

Carbaglu (carglumic acid) will be considered for coverage when the following criteria are met:

#### **N-acetylglutamate Synthase (NAGS) Deficiency with Hyperammonemia**

For **initial** authorization:

1. Medication must be prescribed by or in consultation with a geneticist, hepatologist or a metabolic disorder specialist; AND
2. Member has confirmed hepatic N-acetylglutamate synthase deficiency confirmed by enzyme analysis or DNA mutation analysis; AND
3. Member has an elevated plasma ammonia level based on the patient's age; AND
4. Member has a normal anion gap and a normal blood glucose level; AND

2. Member has a diagnosis of Propionic Acidemia (PA) or Methylmalonic Acidemia (MMA), confirmed by genetic, urine or enzymatic analysis;
3. Member has a documented plasma ammonia level of  $\geq 50$  micromol/L; AND
- 4.