curative

Carbaglu Prior Authorization

Drug(s) Applied:	**Carbaglu** (carglumic acid) soluble tablets

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 µmol/L (> 260 µg/dl) or higher; Older child or adult: plasma ammonia level > 100 µmol/L (175 µg/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 ≥50 µmol/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