Carbaglu (carglumic acid)

Override(s)	Approval Duration
Prior Authorization	N-acetylglutamate synthase (NAGS) deficiency: 1 year Propionic Acidemia (PA) or Methylmalonic Acidetrimmia (MMA): 1 month

Medications	Comments
carglumic acid tablets	Preferred
Carbaglu tablets	Non-Preferred

APPROVAL CRITERIA

Initial requests for Carbaglu (carglumic acid) for N-acetylglutamate synthase (NAGS) deficiency may be approved for the following:

I. Documentation is provided that individual has a diagnosis of *acute* hyperammonemia due to the deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS); **AND**
II. Using as adjunctive therapy with other ammonia lowering therapies, including but not limited to the following:
   A. Alternate pathway medications to eliminate nitrogen waste (such as, sodium phenylacetate); **OR**
   B. Hemodialysis; **OR**
   C. Dietary protein restriction;

OR

III. Documentation is provided that individual has a diagnosis of *chronic* hyperammonemia due to the deficiency of the hepatic enzyme NAGS; **AND**
IV. Using as maintenance therapy;

AND

V. Individual has had a trial (medication samples/coupons/discount cards are excluded from consideration as a trial) and inadequate response or intolerance to generic carglumic acid tablet.

Continuation requests for Carbaglu (carglumic acid) for N-acetylglutamate synthase (NAGS) deficiency may be approved if the following criterion is met:

I. Documentation is provided that there is clinically significant improvement or stabilization in plasma ammonia level.

Requests for Carbaglu (carglumic acid) for propionic acidemia (PA) or methylmalonic acidetrimmia (MMA) may be approved for the following:

I. Documentation is provided that individual has a diagnosis of *acute* hyperammonemia due to propionic acidemia (PA) or methylmalonic acidemia (MMA); **AND**
II. Using as adjunctive therapy with other ammonia lowering therapies including but not limited to the following:
   A. Intravenous glucose;
   B. Insulin;
   C. L-carnitine;
   D. Hemodialysis;
   E. Dietary protein restriction.

AND

III. Individual has had a trial (medication samples/coupons/discount cards are excluded from consideration as a trial) and inadequate response or intolerance to generic carglumic acid tablet.

Key References:

1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: December 13, 2021.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Haberle J, Boddaert N, Burlina A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders. *Orphanet J Rare Dis*. 2012;7.
4. Lee B. Urea cycle disorders: Management. Last updated: August 6, 2021. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: December 12, 2021.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2021; Updated periodically.