SPECIALTY GUIDELINE MANAGEMENT

CARBAGLU (carglumic acid)

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indications
1. Acute hyperammonemia in patients with NAGS deficiency
   Carbaglu is indicated as an adjunctive therapy in pediatric and adult patients for the treatment of acute hyperammonemia due to the deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS). During acute hyperammonemic episodes, concomitant administration of Carbaglu with other ammonia lowering therapies such as alternate pathway medications, hemodialysis, and dietary protein restriction is recommended.

2. Maintenance therapy for chronic hyperammonemia in patients with NAGS deficiency
   Carbaglu is indicated for maintenance therapy in pediatric and adult patients for chronic hyperammonemia due to the deficiency of the hepatic enzyme NAGS. During maintenance therapy, the concomitant use of other ammonia lowering therapies and protein restriction may be needed based on plasma ammonia levels.

B. Compendial Uses
1. Methylmalonic acidemia
2. Propionic acidemia

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. N-acetylglutamate synthase (NAGS) Deficiency
   Authorization of indefinite approval may be granted for members with diagnosis of NAGS deficiency confirmed by enzymatic or genetic testing.

B. Methylmalonic Acidemia
   Authorization of indefinite approval may be granted for members who have a diagnosis of methylmalonic acidemia.

C. Propionic Acidemia
   Authorization of indefinite approval may be granted for members who have a diagnosis of propionic acidemia.

III. CONTINUATION OF THERAPY
All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES