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
      i. 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 are recommended.
   2. Maintenance therapy for chronic hyperammonemia in patients with NAGS deficiency
      i. 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 reduced or discontinued 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. REQUIRED DOCUMENTATION

The following information is necessary to initiate the prior authorization review: Documentation of enzymatic or genetic testing to confirm the diagnosis of NAGS deficiency (NAGS deficiency only)

III. CRITERIA FOR APPROVAL

A. 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.
IV. DOSAGE AND ADMINISTRATION

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

V. REFERENCES