

## Specialty Pharmacy Program

### Carbaglu<sup>®</sup> (carglumic acid)

#### DESCRIPTION

Carbaglu acts as a replacement for the compound N-acetylglutamate (NAG), which is lacking in patients with deficiency of the enzyme NAG synthase. NAG synthase is one of several enzymes that help to break down nitrogen and remove ammonia from the body. Carbaglu is FDA-approved for acute treatment of hyperammonemia and for maintenance treatment of chronic hyperammonemia in patients with NAG synthase deficiency. Carbaglu may also be used for treating methylmalonic acidemia and propionic acidemia.

#### APPROVAL DURATION

Approval duration: 1 year

#### APPROVAL CRITERIA

Carbaglu will be approved if the patient meets the following criteria:

- I. Patient has a diagnosis of NAG synthase deficiency **AND**
- II. The diagnosis was confirmed by enzyme analysis or genetic testing **OR**
- III. Patient has a diagnosis of methylmalonic acidemia **OR**
- IV. Patient has a diagnosis of propionic acidemia.