AvMed

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-305-671-0200**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process can be delayed.**

Drug Requested: carglumic acid (Carbaglu®)

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member AvMed #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight: _____ **Date:** _____

Recommended Dosage:

- NAGS deficiency, acute hyperammonemia: 100 to 250 mg/kg/day given in 2 to 4 divided doses
- NAGS deficiency, chronic hyperammonemia: 10 to 100 mg/kg/day given in 2 to 4 divided doses
- Propionic acidemia or methylmalonic acidemia, acute hyperammonemia: Oral: 3.3g/m^2/day in 2 divided doses (12 hours apart) and for a maximum of 7 days

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

(Continued on next page)

❑ **N-acetylglutamate synthase (NAGS) deficiency**

Initial Authorization: 6 months

- ❒ Provider is or has consulted with a specialist in medical genetics or other specialist in treatment of urea cycle disorders
- ❒ Member has diagnosis of NAGS deficiency as confirmed by genetic testing **(submit results)**
- ❒ Member is experiencing hyperammonemia despite compliance with standard therapy **(submit current plasma ammonia lab test results and chart notes documenting therapies tried)**
- ❒ For treatment of acute hyperammonemia, carglumic acid will be used in conjunction with standard therapy (i.e. hemodialysis, intravenous sodium benzoate and phenylacetate, protein restriction)
- ❒ Prescribed dose will not exceed 250 mg/kg per day initially, followed by a maintenance dose of 100 mg/kg per day
- ❒ For approval of brand name Carbaglu: Member has had trial and intolerable life-endangering adverse event with generic carglumic acid tablets **(must submit completed MedWatch form and chart notes to document adverse event)**

❑ **N-acetylglutamate synthase (NAGS) deficiency**

Reauthorization: 12 months.

- ❒ All initial authorization criteria continues to be met
- ❒ Member's plasma ammonia levels have been sustained at or below normal limits for age **(submit current lab test results)**
- ❒ Member is **NOT** experiencing any symptoms of unacceptable toxicity associated with carglumic acid
- ❒ For approval of brand name Carbaglu: Member has had trial and intolerable life-endangering adverse event with generic carglumic acid tablets **(must submit completed MedWatch form and chart notes to document adverse event)**

❑ **Propionic Acidemia (PA) or Methylmalonic Acidemia (MMA) with acute hyperammonemia**

Authorization Criteria: 7 day length of authorization. Coverage cannot be renewed.

- ❒ Provider is or has consulted with a specialist in medical genetics or other specialist in treatment of urea cycle disorders
- ❒ Member has diagnosis of propionic acidemia or methylmalonic acidemia as confirmed by genetic testing **(submit results)**
- ❒ Member's plasma ammonia level is ≥ 70 µmol/L despite standard of care treatment, such as intravenous hydration and nutritional support **(submit current plasma ammonia lab test results and chart notes documenting therapies tried)**

(Continued on next page)

- ❒ Medication will be used in conjunction with other ammonia-lowering therapies (i.e. intravenous glucose, insulin, L-carnitine, protein restriction, hemodialysis)
- ❒ Medication will only be used until the patient's ammonia level is < 50 µmol/L and for a maximum duration of 7 days
- ❒ For approval of brand name Carbaglu: Member has had trial and intolerable life-endangering adverse event with generic carglumic acid tablets **(must submit completed MedWatch form and chart notes to document adverse event)**

Medication being provided by Specialty Pharmacy - PropriumRx

Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****

***Approved by Pharmacy and Therapeutics Committee**: 7/21/2022
REVISED/UPDATED/REFORMATTED: ~~8/10/2022;~~ **10/26/2023**