



Prior Authorization Approval Criteria

Carbaglu (carglumic acid)

Generic name:	carglumic acid
Brand name:	Carbaglu
Medication class:	CPS 1 activator
FDA-approved uses:	for acute and chronic treatment of hyperammonemia due to N-acetylglutamate synthase (NAG) deficiency.
Available dosage forms:	200mg scored tablet.
Usual dose:	<p>In adults and children:</p> <ul style="list-style-type: none">-Initial dosage for acute hyperammonemia is 100 to 250 mg/kg/day, with the dosage adjusted to maintain normal levels based on age.-Maintenance dosage is usually less than 100 mg/kg/day. The total daily dosage should be divided into 2 to 4 doses to be given immediately before meals or feedings. In adults, each divided dose should be rounded to the nearest 100 mg (half tablet).-The tablets should not be swallowed whole or crushed. Carglumic acid can be administered orally or through a nasogastric tube.-For oral administration in adults, each 200 mg tablet should be dispersed in a minimum of 2.5 mL of water and taken immediately. The mixing container should be rinsed with additional water and the contents swallowed immediately. For administration through a nasogastric tube, each 200 mg tablet should be mixed with a minimum of 2.5 mL of water and shaken gently to quickly disperse. The dispersion should be immediately administered through the nasogastric tube, followed by flushing with additional water to clear the tube.-For children, each 200 mg tablet should be mixed with 2.5 mL of water to yield a concentration of 80 mg/mL in a mixing container. The appropriate volume of dispersion should be drawn up and administered either via an oral syringe or through a nasogastric tube. If an oral syringe is used, it should be refilled with at least 1 to 2 mL of water and administered immediately. For administration through a nasogastric tube, it should be flushed with additional water to clear the tube. Any unused portion of the dispersion should be discarded.-Mixing carglumic acid with foods or liquids other than water has not been studied and is not recommended.

Approximate monthly cost:
(based on AWP 2011)

based on 100mg/kg/day for a 70kg patient and \$159/pill.
Daily dose would be 7000mg/day (35pills/day)
Cost/day= \$5565.00 or \$166,950/month.

Duration of therapy:

indefinitely

Criteria for use (*bullet points below are all inclusive unless otherwise noted*):

- The indicated diagnosis (including any applicable labs and /or tests) and medication usage must be supported by documentation from the patient's medical records.
- Clinically diagnosed with hyperammonemia due to N-acetylglutamate synthase (NAG) deficiency.
- therapy should be initiated by a health care provider experienced in the treatment of metabolic disorders.

Monitoring:

- Monitoring of plasma ammonia levels, neurological status, and clinical response is necessary to assess patient response. Plasma ammonia levels should be maintained within the normal range for the age of the patient.

Contraindication:

- None reported at this time.

Not approved if:

- Patient does not meet the above stated criteria.

Special considerations:

- In acute therapy, carglumic acid should be administered with other ammonia-lowering therapies, including alternate pathway medications, hemodialysis, and dietary protein restriction.
- During maintenance therapy, the concomitant use of other ammonia-lowering therapies and protein restriction may be reduced or discontinued.
- Current available therapies used to decrease hyperammonemia include sodium phenylbutyrate and sodium phenylacetate/sodium benzoate, as well as hemodialysis, arginine citrulline, carnitine, and dietary protein restriction.

FCHP Pharmacy and Therapeutics Committee approval: _____

Date: _____

Adopted: 06/08/11