

### Clinical Pharmacy Program Guidelines for Carbaglu

Program	Prior Authorization
Medication	Carbaglu™ (carglumic acid)
Issue Date	7/2017
Pharmacy and Therapeutics Approval Date	7/2017
Effective Date	9/2017

**1. Background:**

Carbaglu (carglumic acid) is a Carbamoyl Phosphate Synthetase 1 (CPS 1) activator indicated for maintenance therapy in pediatric and adult patients for chronic hyperammonemia due to the deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS). Carbaglu is also 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.<sup>1</sup>

**2. Coverage Criteria:**

<p><b>A. <u>Initial Authorization</u></b></p> <p><b>1. Carbaglu</b> will be approved based on the following criteria:</p> <p style="padding-left: 40px;">a. Diagnosis of N-acetylglutamate synthase (NAGS) deficiency</p> <p style="padding-left: 40px;"><b>Authorization will be issued for 12 months.</b></p> <p><b>B. <u>Reauthorization</u></b></p> <p><b>1. Carbaglu</b> will be approved based on the following criteria:</p> <p style="padding-left: 40px;">a. Documentation of positive clinical response to Carbaglu therapy</p> <p style="padding-left: 40px;"><b>Authorization will be issued for 12 months.</b></p>
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**3. References:**

1. Carbaglu® [package insert], Memphis, TN: Accredo Health Group, Inc. ; August 2013.

Program	Prior Authorization
<b>Change Control</b>	
Date	Change
7/2017	New program