

Policies Repository



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Policy Title Carglumic Acid (Carbaglu)

Policy Number

Application of pharmacy policy is determined by benefits and contracts. Benefits may vary based on product line, group, or contract. Some medications may be subject to precertification and age, gender or quantity restrictions. Individual member benefits must be verified.

This pharmacy policy document describes the status of pharmaceutical information and/or technology at the time the document was developed. Since that time, new information relating to drug efficacy, interactions, contraindications, dosage, administration routes, safety, or FDA approval may have changed. If the Medical/Pharmacy Reviewer is aware of any new information on the subject of this document, please provide it promptly to the Medical/Pharmacy Policy Department. This information may include new FDA approved indications, withdrawals or other FDA alerts. This type of information is relevant not only when considering whether this policy should be updated, but also when applying it to current requests for coverage.

Members are advised to use participating pharmacies in order to receive the highest level of benefits.

Policy	<p>Carglumic Acid (Carbaglu) is indicated for Adjunctive therapy in the treatment of acute hyperammonemia and maintenance therapy of chronic hyperammonemia due to the deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS).</p> <p>The use of Carglumic Acid (Carbaglu) requires prior authorization (i.e. clinical pharmacy and/or medical director review).</p>					
Policy description	<p>Carglumic acid (Carbaglu) is a synthetic structural analogue of N-acetylglutamate (NAG), which is an essential allosteric activator of carbamoyl phosphate synthetase 1 (CPS 1) in liver mitochondria. CPS 1 is the first enzyme of the urea cycle, which converts ammonia into urea. NAG is the product of NAGS, a mitochondrial enzyme. Carglumic acid acts as a replacement for NAG in NAGS deficiency patients by activating CPS 1.</p>					
Policy guideline inclusion	<p>Carglumic Acid (Carbaglu) is approved when the following inclusion criteria is met:</p> <ul style="list-style-type: none"> Documentation of a diagnosis of N-acetylglutamate synthase (NAGS) deficiency 					
Policy guideline exclusion	<p>Carglumic Acid (Carbaglu) is denied when the following exclusion criteria is present:</p> <ul style="list-style-type: none"> No documentation of a diagnosis of N-acetylglutamate synthase (NAGS) deficiency 					
Policy List of Applicable Drugs	<table border="1"> <thead> <tr> <th>Brand Name</th> <th>Generic Name</th> </tr> </thead> <tbody> <tr> <td>Carbaglu</td> <td>Carglumic Acid</td> </tr> </tbody> </table>	Brand Name	Generic Name	Carbaglu	Carglumic Acid	
Brand Name	Generic Name					
Carbaglu	Carglumic Acid					
Dosing and administration	<p>Refer to the specific manufacturer's prescribing information for administration and dosage details for each specific agent.</p>					
Policy references	<p>Carbaglu [package insert]. Paris, France: Orphan Europe SARL; March 2010.</p> <p>Caldovic L, Ah Mew NA, Shi D, Morizono H, Yudkoff M, Tuchman M. N-acetylglutamate synthase: structure, function and defects [published online ahead of print February 26, 2010]. Mol Genet Metab 2010;100(suppl 1):S12-S19. doi:10.1016/j.ymgme.2010.02.018</p>					

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Drug details: Carbaglu (NDA 022562). Drugs@FDA Web site. <http://www.accessdata.fda.gov/scripts/cder/drugsatfda/index.cfm?fuseaction=Search DrugDetails>. Published March 18, 2010. Accessed April 6, 2010.

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Policy link to related policies

Version effective date

07/01/2001

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