Carbaglu (carglumic acid)  
Effective 10/01/2020

<table>
<thead>
<tr>
<th>Plan</th>
<th>Program Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>☑ MassHealth</td>
<td>☑ Prior Authorization</td>
</tr>
<tr>
<td>☑ Commercial/Exchange</td>
<td>☐ Quantity Limit</td>
</tr>
<tr>
<td>☑ Commercial/Exchange</td>
<td>☐ Step Therapy</td>
</tr>
<tr>
<td>☑ Pharmacy Benefit</td>
<td></td>
</tr>
<tr>
<td>☐ Medical Benefit (NLX)</td>
<td></td>
</tr>
</tbody>
</table>

**Specialty Limitations**

This medication has been designated specialty and must be filled at a contracted specialty pharmacy.

**Contact Information**

<table>
<thead>
<tr>
<th>All Plans</th>
<th>Specialty Medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phone: 866-814-5506</td>
<td>Fax: 866-249-6155</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>MassHealth</th>
<th>Phone: 877-433-7643</th>
<th>Fax: 866-255-7569</th>
</tr>
</thead>
<tbody>
<tr>
<td>Commercial</td>
<td>Phone: 800-294-5979</td>
<td>Fax: 888-836-0730</td>
</tr>
<tr>
<td>Exchange</td>
<td>Phone: 855-582-2022</td>
<td>Fax: 855-245-2134</td>
</tr>
</tbody>
</table>

**Medical Specialty Medications (NLX)**

| All Plans | Phone: 844-345-2803 | Fax: 844-851-0882 |

**Exceptions**

N/A

---

**Overview**

Carbaglu is used for the adjunctive treatment of acute hyperammonemia and maintenance therapy of chronic hyperammonemia due to the deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS) in adult and pediatric patients.

**Coverage Guidelines**

Authorization may be granted for members who are currently receiving treatment with Carbaglu, excluding when the product is obtained as samples or via manufacturer’s patient assistance programs OR

Authorization may be granted when one of the following criteria is met:

1. Diagnosis is hyperammonemia due to the N-acetylglutamate synthetase (NAGS) deficiency AND

**Continuation of Therapy**

Reauthorization may be granted when improvement per physician assessment/evaluation and documentation of improved and/or normalized blood ammonia levels for age is received.

**Limitations**

1. Initial approvals will be granted for 12 months.
2. Reauthorizations will be granted for 36 months.

**Appendix**

<table>
<thead>
<tr>
<th>Recommended Dosing:</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute hyperammonemia (adult &amp; pediatric)</td>
<td>Initial: 100 to 250 mg/kg/day</td>
</tr>
<tr>
<td>Chronic hyperammonemia (adult &amp; pediatric)</td>
<td>Initial: ≤ 100mg/kg/day</td>
</tr>
</tbody>
</table>
Maintenance: dose titrated to the normal plasma ammonia level for age (generally less than 100 mg/kg/day); total daily dose should be divided into 2 to 4 doses and rounded to the nearest 100 mg.

Note: tablets should not be swallowed whole or crushed. Please refer to the prescribing information for adult and pediatric oral administration recommendations as well as nasogastric tube administration directions.

Pharmacist’s Notes:
1. Any episode of acute symptomatic hyperammonemia should be treated as a life-threatening emergency & treatment may require hemodialysis in some instances.
2. The management of hyperammonemia due to NAGS deficiency should be done in coordination with medical personnel experienced in metabolic disorders.
3. Plasma ammonia levels should also be maintained within normal range for age through individual dose adjustment.
4. During acute hyperammonemia episodes, protein restrictions and hyper-caloric intake is recommended to block ammonia-generating catabolic pathways. Protein intake can subsequently be increased when ammonia levels have normalized.

References

Review History
06/25/2012 – Reviewed
06/24/2013 – Reviewed
06/23/2014 – Reviewed
06/22/2015 – Reviewed
06/27/2016 – Reviewed
06/26/2017 – Reviewed
06/25/2018 – Reviewed
06/19/2019 – Reviewed

Disclaimer
AllWays Health Partners complies with applicable federal civil rights laws and does not discriminate or exclude people on the basis of race, color, national origin, age, disability, or sex.