Premier Health Plan

POLICY AND PROCEDURE

POLICY TITLE: ***Carbaglu (carglunic acid)***
DEPARTMENT: **Clinical Pharmacy Services- Utilization Management**
ORIGINAL DATE: ***July 2010 (as adopted from UPMC Health Plan)***

Last P & T Committee Approval Date: *February 2018*

Product Applicability: *mark all applicable products below:*

COMMERCIAL	[ ] HMO [ ] PPO *Products*: [ ] Small [ ] Indiv. [ ] Large *Exchange*: [ ] Shop [ ] Indiv. [x] All
OTHER	[x] Self-funded/ASO

PURPOSE

The purpose of this policy is to define the prior authorization process for Carbaglu (carglunic acid).

Carbaglu (carglunic acid) is a carbamoyl phosphate synthetase 1 (CPS 1) activator and is intended for oral administration. It is indicated for use as adjunctive therapy for the treatment of acute hyperammonemia and maintenance therapy for the treatment of chronic hyperammonemia due to the deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS). In patients with NAGS deficiency, carglumic acid (Carbaglu) has been shown to decrease plasma ammonia levels.

DEFINITIONS

Carbamoyl phosphate synthase 1 (CPS 1) – the first enzyme of the urea cycle, which converts ammonia into urea.

N-acetylglutamate (NAG) – an essential allosteric activator of carbamoyl phosphate synthetase 1 (CPS 1) in liver mitochondria. NAG is the product of N-acetylglutamate

synthase (NAGS), a mitochondrial enzyme. Carglumic acid acts as a replacement for NAG in NAGS deficiency patients by activating CPS 1.

N-acetylglutamate synthase (NAGS) – an enzyme which converts acetylCoA and glutamate into N-acetylglutamate (NAG).

POLICY

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, and approval by the Pharmacy & Therapeutics Committee of the criteria for prior authorization, as described in RX.002 Pharmacy and Therapeutics Committee and RX.003-Prior Authorization Process.

The drug, Carbaglu (carglunic acid), is subject to the prior authorization process.

PROCEDURE

Initial Authorization Criteria:

Must meet all of the criteria listed below:

- Must be prescribed by a physician who specializes in the treatment of inherited metabolic disorders or in consultation with this specialist
- Must have a diagnosis of one of the following deficiencies. Chart documentation describing how diagnosis was confirmed (e.g. genetic testing results, enzyme assays, ammonia levels, progress notes, etc.) is required.
    - N-acetylglutamate synthase (NAGS)
    - N-acetylglutamate (NAG)
    - Carbamoyl phosphate synthetase 1 (CPS 1)

Reauthorization Criteria:

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 3 months
Reauthorization	Up to 1 year

If the established criteria are not met, the request is referred to a Medical Director for review.

REFERENCES

1. Carbaglu [package insert]. Orphan Europe. Paris, France. March 2010.
2. Haberle J, Boddaert N, Burlina A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders. *Orphanet Journal of Rare Diseases* 2012;7:32. http://www.ojrd.com/content/7/2/32. Accessed February 20, 2013
3. The Urea Cycle Disorders Conference Group. Consensus statement for the management of patients with urea cycle disorders. *J Pediatr* 2001;138:S1-S5
4. Summar M, Tuchman M. Proceeding of a consensus conference for the management of patients with urea cycle disorders. *J Pediatr* 2001;138:S6-S10
5. Wilcken B. Problems in the management of urea cycle disorders. *Molecular Genetics Metabolism* 2004;81:S86-S91

RECORD RETENTION

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual review	*02/17, 02/18*