Prior Authorization Review Panel

CHC-MCO Policy Submission

A separate copy of this form must accompany each policy submitted for review.
Policies submitted without this form will not be considered for review.

Plan: PA Health & Wellness	**Submission Date: N/A**
Policy Number: PHW.PDL.733	**Effective Date: 01/01/2020** **Revision Date: 07/2020**
Policy Name: Urea Cycle Disorder Agents	

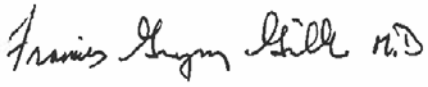
Type of Submission – Check all that apply:

- ☐ **New Policy**
- ☐ **Revised Policy***
- ✓ **Annual Review - No Revisions**
- ✓ **Statewide PDL -** *Select this box when submitting policies for Statewide PDL implementation and when submitting policies for drug classes included on the Statewide PDL.*

***All revisions to the policy must be highlighted using track changes throughout the document.**

Please provide any changes or clarifying information for the policy below:

Q3 2020 annual review: no changes.

Name of Authorized Individual (Please type or print): **Francis G. Grillo, MD**	**Signature of Authorized Individual:** 

Clinical Policy: Urea Cycle Disorder Agents

Reference Number: PHW.PDL.733
Effective Date: 01/01/2020
Last Review Date: 07/2020

Revision Log

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of health plans affiliated with PA Health and Wellness® that Urea Cycle Disorder Agents is **medically necessary** when the following criteria are met:

I. Requirements for Prior Authorization of Urea Cycle Disorder Agents

A. Prescriptions That Require Prior Authorization

Prescriptions for Urea Cycle Disorder Agents that meet any of the following conditions must be prior authorized:

1. A non-preferred Urea Cycle Disorder Agent.

2. A Urea Cycle Disorder Agent with a prescribed quantity that exceeds the quantity limit.

B. Review of Documentation for Medical Necessity

In evaluating a request for prior authorization of a prescription for a Urea Cycle Disorder Agent, the determination of whether the requested prescription is medically necessary will take into account whether the beneficiary:

1. Is prescribed the Urea Cycle Disorder Agent by or in consultation with a physician who specializes in treating metabolic disorders; **AND**

2. Is being treated for a diagnosis that is indicated in the U.S. Food and Drug Administration (FDA)-approved package labeling OR a medically accepted indication; **AND**

3. Has chart documentation supporting the diagnosis (e.g., ammonia levels, genetic testing, enzyme assays, plasma amino acid/urine orotic acid analyses, progress notes); **AND**

4. Is prescribed a dose and duration of therapy that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**

5. For a non-preferred Urea Cycle Disorder Agent**,** has a documented history of

therapeutic failure, contraindication, or intolerance to the preferred Urea Cycle Disorder Agent; **AND**

6. If a prescription for a Urea Cycle Disorder Agent is in a quantity that exceeds the quantity limit, the determination of whether the prescription is medically necessary will also take into account the guidelines set forth in PA.CP.PMN.59 Quantity Limit Override.

   NOTE: If the beneficiary does not meet the clinical review guidelines above but, in the professional judgement of the physician reviewer, the services are medically necessary to meet the medical needs of the beneficiary, the request for prior authorization will be approved

FOR RENEWALS OF PRESCRIPITONS FOR UREA CYCLE DISORDER AGENTS: The determination of medical necessity of a request for renewal of a prior authorization for a Urea Cycle Disorder Agent that was previously approved will take into account whether the beneficiary:

1. Has documentation from the prescribing provider that the beneficiary had a positive clinical response to therapy; **AND**

2. Is prescribed the Urea Cycle Disorder Agent by or in consultation with a physician who specializes in treating metabolic disorders; **AND**

3. Is prescribed a dose and duration of therapy that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**

4. If a prescription for a Urea Cycle Disorder Agent is in a quantity that exceeds the quantity limit, the determination of whether the prescription is medically necessary will also take into account the guidelines set forth in PA.CP.PMN.59 Quantity Limit Override.

   NOTE: If the beneficiary does not meet the clinical review guidelines above but, in the professional judgement of the physician reviewer, the services are medically necessary to meet the medical needs of the beneficiary, the request for prior authorization will be approved.

C. Clinical Review Process

Prior authorization personnel will review the request for prior authorization and apply the clinical guidelines in Section B. above to assess the medical necessity of a prescription for a Urea Cycle Disorder Agent. If the guidelines in Section B. are met, the reviewer will prior authorize the prescription. If the guidelines are not met, the prior authorization request will be referred to a physician reviewer for a medical necessity determination. Such a request for prior authorization will be approved when, in the professional

judgment of the physician reviewer, the services are medically necessary to meet the medical needs of the beneficiary.

D. **Approval Duration:**
- **New Request: 6 months**
- **Renewal Request: 12 months**

E. References

1. Haberle J, Boddaert N, et.al. Suggested guidelines for the diagnosis and management of urea cycle disorders. *Orphanet Journal of Rare Diseases*. 2012, 7:32.
2. Diaz G.A, Krivitzky L.S, et.al. Ammonia Control and Neurocognitive Outcome Among Urea Cycle Disorder Patients Treated with Glycerol Phenylbutyrate. *Hepatology*. 2013 June; 57(6): 2171–2179.
3. Smith W, Diaz G.A, et al. Ammonia control in children ages 2 months through 5 years with urea cycle disorders: comparison of sodium phenylbutyrate and glycerol phenylbutyrate. *Journal of Pediatrics*. 2013 June; 162(6): 1228–1234.
4. Ravicti Prescribing Information. Lake Forest, IL. Horizon Therapeutics, LLC.
5. Buphenyl Prescribing Information. Scottsdale, AZ: Ucyclyd Pharma Inc.; April 2009.

Reviews, Revisions, and Approvals	Date
Policy created	01/01/2020
Q3 2020 annual review: no changes.	07/2020