

### Clinical Pharmacy Program Guidelines for Buphenyl

Program	Prior Authorization
Medication	Buphenyl (sodium phenylbutyrate) oral solution
Issue Date	5/2016
Pharmacy and Therapeutics Approval Date	7/2017
Effective Date	9/2017

**1. Background:**

**Urea cycle disorders (UCDs)**

Buphenyl is indicated as adjunctive therapy in the chronic management of patients with urea cycle disorders (UCDs). It must be combined with dietary protein restriction with or without essential amino acid supplementation. Buphenyl is indicated in all patients with neonatal-onset deficiency. It is also indicated in patients with late-onset disease (partial enzymatic deficiency, presenting after the first month of life) who have a history of hyperammonemic encephalopathy. It is important that the diagnosis be made early and treatment initiated immediately to improve survival. Buphenyl should not be used in the management of acute hyperammonemia.

**NOTE:** This policy applies to only the oral solution.

**2. Coverage Criteria:**

A.	<b>Criteria for approval</b>
1.	Diagnosis of urea cycle disorders (UCDs)
	<b>Authorization will be issued for 12 months.</b>

**3. References:**

- BUPHENYL [package insert]. Lake Forest, IL:Horizon Therapeutics Inc; 2016.

Program	Program type – Prior Authorization
<b>Change Control</b>	
Date	Change
5/2016	New Program
7/2016	Auto-update: Applicable to only oral solution.
7/2017	Updated policy template. Updated background and template.

