PRIOR AUTHORIZATION CRITERIA

BRAND	**BUPHENYL**
(generic)	**(sodium phenylbutyrate)**
	sodium phenylbutyrate
Status: CVS caremark Criteria	***MDC***
Type: Initial Prior Authorization	***Ref # 965-A***

FDA-APPROVED INDICATION[1]

Buphenyl is indicated as adjunctive therapy in the chronic management of patients with urea cycle disorders involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS). It is indicated in all patients with neonatal-onset deficiency (complete enzymatic deficiency, presenting within the first 28 days of life). 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. Any episode of acute hyperammonemia should be treated as a life-threatening emergency.

CRITERIA FOR APPROVAL

1	Does the patient have a diagnosis of urea cycle disorder? [If no, no further questions.]	Yes	No
2	Was the diagnosis confirmed by enzymatic, biochemical, or genetic testing? [If no, no further questions.]	Yes	No
3	Will Buphenyl be used for chronic management of urea cycle disorder?	Yes	No

Guidelines for Approval	
Duration of Approval	**12 months**
Set 1: UCD	
Yes to question(s)	**No to question(s)**
1	None
2	
3	

Internal Use Only – Mapping Instructions		
	Yes	**No**
1.	Go to 2	Deny
2.	Go to 3	Deny
3.	Approve, 12 months	Deny

RATIONALE

These criteria meet the Medicare Part D definition of a medically accepted indication. This definition includes uses which are approved by the FDA or supported by a citation included, or approved for inclusion, in one of the Medicare approved compendia.

The intent of the criteria is to ensure that patients follow selection elements noted in labeling and/or practice guidelines in order to decrease the potential for inappropriate utilization.

CVS caremark™

REFERENCE

1. Buphenyl [package insert]. South San Francisco, CA: Hyperion Therapeutics Inc.; April 2016.

DOCUMENT HISTORY

Written by:	UM Development (CT) 03/2013
Revised:	Specialty Clinical Development (KW) 06/2013, ST 04/2014; JP 04/2015, TS 04/2016, JP 06/2016 (CMS), 09/2016 (CMS feedback), TS 07/2017
Reviewed:	CDPR/ KP 03/2013, LMS 06/2013; KP 05/2014; DNC 04/2015; N/C 04/2016, LMS 09/2016
External Review:	07/2013, 06/2014, 05/2015, 07/2016