

Market Applicability							
Market	DC	GA	KY	MD	NJ	NY	WA
Applicable	X	X	X	X	X	X	NA

Buphenyl (sodium phenylbutyrate)

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year

Medications	Quantity Limit
Buphenyl (sodium phenylbutyrate) tablets Buphenyl (sodium phenylbutyrate) powder	May be subject to quantity limit

APPROVAL CRITERIA

Requests for Buphenyl (sodium phenylbutyrate) may be approved for members who meet the following criteria:

- I. Individual has a diagnosis of urea cycle disorder (UCD), involving enzyme deficiencies of **one** of the following:
 - A. Carbamylphosphate synthetase (CPS); **OR**
 - B. Ornithine transcarbamylase (OTC); **OR**
 - C. Argininosuccinic acid synthetase (AS);

AND

- II. Individual has **one** of the following:
 - A. Neonatal-onset deficiency with complete enzymatic deficiency presenting within the first 28 days of life; **OR**
 - B. Late-onset disease defined as the following:
 1. Partial enzymatic deficiency presenting after the first month of life; **AND**
 2. History of hyperammonemic encephalopathy;

AND

- III. Using as adjunctive therapy for chronic management of hyperammonemia, including but not limited to the following:
 - A. In combination with dietary protein restriction and, in some cases, essential amino acid supplementation.

Requests for Buphenyl (sodium phenylbutyrate) may not be approved for the following:

- I. Management of acute hyperammonemia.

Market Applicability							
Market	DC	GA	KY	MD	NJ	NY	WA
Applicable	X	X	X	X	X	X	NA

Key References:

1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. <http://dailymed.nlm.nih.gov/dailymed/about.cfm>. Accessed: December 3, 2019.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.