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

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

I. Documentation is provided that 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 is 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.

Continuation requests for Buphenyl (sodium phenylbutyrate) may be approved if the following criteria are met:

I. Documentation is provided that there is clinically significant improvement or stabilization in plasma ammonia level; **AND**
II. Use in combination with dietary protein restriction.

Requests for **brand** Buphenyl must also meet the following criteria, in addition to the above Prior Authorization criteria:

I. Documentation is provided that individual has failed an adequate trial (medication samples/coupons/discount cards are excluded from consideration as a trial) of one chemically equivalent generic sodium phenylbutyrate agent; **AND**
   A. Documentation is provided that generic sodium phenylbutyrate had inadequate response; **OR**
   B. Documentation is provided that generic sodium phenylbutyrate caused adverse outcome; **OR**

C. Documentation is provided that the individual has a genuine allergic reaction to an inactive ingredient in generic agent. Allergic reaction(s) must be clearly documented in the individual's medical record.

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

I. Management of acute hyperammonemia; **OR**
II. Treatment of malignancy; **OR**
III. Use in combination with Ravicti.

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 9, 2022.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Haberle J, Boddaert N, Burlina A, et al. Suggested guidelines for the diagnosis and management of urea cycle disorders. *Orphanet J Rare Dis*. 2012;7.
4. Lee B. Urea cycle disorders: Management. Last updated: August 6, 2021. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: December 9, 2022.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.