

Urea Cycle Disorders Prior Authorization

Drug(s) Applied:	Sodium phenylbutyrate tablets
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Criteria:

Drug(s) Applied will be approved when the requested medication is being used for an FDA approved indication and all of the following criteria are met:

I. Initial Therapy Criteria

A. Urea Cycle Disorder as indicated by chart notes within past 180 days

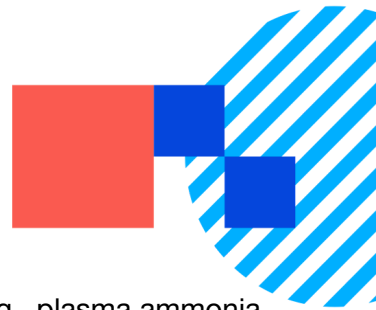
1. Diagnosis of hyperammonemia with ALL of the following:
 - a) Elevated ammonia levels according to the patient's age [Neonate: plasma ammonia level 150 $\mu\text{mol/L}$ (greater than 260 $\mu\text{g/dL}$) or higher; Older child or adult: plasma ammonia level greater than 100 $\mu\text{mol/L}$ (175 $\mu\text{g/dL}$)] **and**
 - b) Normal anion gap **and**
 - c) Normal blood glucose level **and**
2. Urea cycle disorder of CPSID, OTCD, ASSD, ASLD, or ARGD is confirmed by enzyme analysis or genetic testing **and**
3. Patient weighs at least 20 kg **and**
4. Patient is unable to maintain a plasma ammonia level within the normal range with the use of a protein restricted diet and, when clinically appropriate, essential amino acid supplementation **and**
5. Patient will be using the requested agent as adjunctive therapy to dietary protein restriction **and**
6. Chart notes and/or prescriber do not show documentation that the requested drug will be used for treatment of acute hyperammonemia **and**
7. Chart notes and/or prescriber do not show documentation that patient will be taking the requested agent concurrently with haloperidol or valproic acid or prescriber has provided a statement of benefit over risk **and**
8. Prescriber is a specialist or has consulted with a specialist in the area of the patient's diagnosis (e.g., metabolic disorders)

Approval Duration: 12 months

II. Continued Therapy Criteria

A. Urea Cycle Disorder as indicated by chart notes within past 12 months

1. Patient has been previously approved for the requested agent through Curative's Prior Authorization process or meets the initial therapy criteria above **and**



2. Patient has had clinical benefit with the requested agent (e.g., plasma ammonia level within the normal range) **and**
3. Chart notes and/or prescriber do not show documentation that the requested drug is being used for treatment of acute hyperammonemia **and**
4. Patient will be using the requested agent as adjunctive therapy to dietary protein restriction **and**
5. Prescriber is a specialist or has consulted with a specialist in the area of the patient's diagnosis (e.g., metabolic disorders)

Approval Duration: 12 months

Policy Owned by: PBM team