curative

Urea Cycle Disorders Prior Authorization

Drug(s) Applied:	**Sodium phenylbutyrate tablets**

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 µmol/L (greater than 260 µg/dL) or higher; Older child or adult: plasma ammonia level greater than 100 µmol/L (175 µg/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