

Criteria-Based Consultation Prescribing Program

CRITERIA FOR DRUG COVERAGE

Glycerol phenylbutyrate (Ravicti)

Notes:

^ Definition of sodium phenylbutyrate ineffectiveness and tolerability:

- **Ineffectiveness:** Ammonia and glutamine levels greater than those achieved with glycerol phenylbutyrate in last 12 months, despite:
 - i. Sodium phenylbutyrate dosage titration
 - ii. Increasing dosage frequency (i.e., splitting total daily dose over four times daily instead of three times daily)
 - iii. Medication adherence greater than 75%
 - iv. Maintaining same dietary protein restriction
- **Tolerability:** Documentation of loss of work, classes, or ill days due to side effects reported with sodium phenylbutyrate despite:
 - i. Sodium phenylbutyrate dosage titration
 - ii. Increasing dosage frequency (i.e., splitting total daily dose over four times daily instead of three times daily)
 - iii. Medication adherence greater than 75%
 - iv. Maintaining same dietary protein restriction
 - v. Dizziness towards end of dosage interval despite increasing dosage frequency (taking sodium phenylbutyrate up to 6 times daily)

Initial approval criteria: Non-formulary **glycerol phenylbutyrate (Ravicti)** will be covered for 12 months on the prescription drug benefit when the following criteria are met:

- Diagnosis of a urea cycle disorder
- At least 2 months of age
- At least 3-month treatment with sodium phenylbutyrate was ineffective[^], not tolerated[^], or is contraindicated
- Planned to be used concomitantly with dietary protein restriction and, in some cases, dietary supplements (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements)

Continued use criteria: Non-formulary **glycerol phenylbutyrate (Ravicti)** will continue to be covered on the prescription drug benefit for 12 months when the following criteria are met:

- Documentation of positive clinical response to glycerol phenylbutyrate therapy:
 - Clinically significant reduction blood ammonia level
- Patient is actively on dietary protein restriction and, in some cases, dietary supplements (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements)

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