



Policy Type: PA/SP

Pharmacy Coverage Policy: EOCCO034

**Description**

Glycerol phenylbutyrate (Ravicti) is an orally administered nitrogen-binding agent.

**Length of Authorization**

- Initial: 12 months
- Renewal: 12 months

**Quantity limits**

glycerol phenylbutyrate (Ravicti)	Indication	Quantity Limit	DDID
1.1 g/mL (25 mL bottle)	Urea Cycle Disorder	500 mL (20 bottles)/30 days	177929

**Initial Evaluation**

- I. Glycerol phenylbutyrate (Ravicti) may be considered medically necessary when the following criteria below are met:
  - A. Age two months and older; **AND**
  - B. A diagnosis of:
    1. **Urea Cycle Disorder; AND**
      - i. Has tried and failed to be managed by dietary protein restriction and amino acid supplementation, or has a contraindication to those therapies; **AND**
      - ii. Has tried and failed sodium phenylbutyrate (Buphenyl); **AND**
      - iii. Has plasma ammonia level >100 µmol/L

**Renewal Evaluation**

- I. Patient has previously received treatment with glycerol phenylbutyrate (Ravicti); **AND**
- II. The patient’s chronic hyperammonia is being properly manage with glycerol phenylbutyrate; **AND**
- III. Absence of unacceptable toxicity from the medication
- IV. Documentation that the patient’s plasma ammonia level is <35 µmol/L



### Supporting Evidence

- I. Glycerol phenylbutyrate (Ravicti) is indicated for use as a nitrogen-binding agent for chronic management of patient with urea cycle disorder (UCD) that cannot be managed by dietary protein restriction and/or dietary supplementation alone.
- II. An elevated plasma ammonia level of 150 µmol/L (>260 µg/dl) or higher in neonates and > 100 µmol/l (175 µg/dl) in older children and adults, is a strong indication for the presence of a urea cycle disorder.
- III. Clinical study results showed ammonia values ranged from 9-35 µmol/L following treatment with glycerol phenylbutyrate (Ravicti).

### References

1. Ravicti [Prescribing Information]. South San Francisco, CA: Hyperion Therapeutics Inc.; January 2013.
2. Lee B. Urea Cycle Disorder: Management. UpToDate [database online]. Available at
  - a. <<http://www.uptodate.com/home/index.html>> Updated March 26, 2013. [Accessed on July 22, 2013].
3. Urea Cycle Disorders Treatment Guidelines. Rare Disease Clinical Research Network. Available at < <http://rarediseasesnetwork.epi.usf.edu/ucdc/physicians/guidelines-main.htm>> [Accessed on July 22, 2013].

### Policy Implementation/Update:

Date Created	July 2013
Date Effective	August 2013
Last Updated	January 2019
Last Reviewed	

Action and Summary of Changes	Date
Criteria update: Included new FDA expanded indication for pediatric patients 2 months and older. Glycerol phenylbutyrate (Ravicti) was originally approved for pediatric patients 2 years and older. Additionally, a question was added to the renewal portion of this policy to assess for toxicity.	01/2019