PRIOR AUTHORIZATION (PA) CHECKLIST FOR RAVICTI

This checklist is for informational purposes only. For health plan–specific criteria, please contact a representative from **Horizon By Your Side**, a patient support program. Initiate your patient's enrollment in Horizon By Your Side by submitting the Patient Enrollment Form. Your patient must complete enrollment to access these patient-focused services and resources.

Although requirements vary by plan, below are the common criteria that may be requested for RAVICTI® (glycerol phenylbutyrate) Oral Liquid. Case Managers can provide education about navigating insurance processes and accessing treatment during your patient's access journey.

1 BENEFITS INVESTIGATION

- ☐ PA requirements vary between plans. Contact the health plan to understand the process, step therapy requirements, duration or approval, and other relevant information.

2 PA REQUIREMENTS

Patient/Provider Information

- ☐ Name
- ☐ Date of birth
- ☐ Health plan
- ☐ Provider name
- ☐ Provider identification number

Some plans may require documentation of specific information, while some may require physician attestation.

Diagnosis Information

- ☐ Diagnosis/ICD-10-CM for one of these urea cycle disorders (UCDs):
  - UCD E72.20
  - OTCD E72.4
  - ASLD E72.22
  - ARGD E72.21
  - CPS1D E72.29
  - ASSD E72.23
  - HHH E72.4
- ☐ UCD diagnosis confirmed through enzymatic, biochemical, or genetic testing
- ☐ Laboratory reports, including plasma amino acid/urine orotic acid analysis
- ☐ Chronic hyperammonemia
- ☐ Ammonia levels
- ☐ Documentation of cognitive and/or functional impairment

Be sure to provide relevant clinical support, such as clinical notes, genetic testing results, etc.

ARGD, arginase deficiency; ASLD, argininosuccinate lyase deficiency; ASSD, argininosuccinate synthetase; CPS1D, carbamoyl phosphate synthetase 1 deficiency; HHH, hyperornithinemia-hyperammonemia-homocitrullinuria; ICD-10-CM, *International Classification of Diseases, Tenth Revision, Clinical Modification*; OTCD, ornithine transcarbamylase deficiency.

INDICATION

RAVICTI (glycerol phenylbutyrate) Oral Liquid is indicated for use as a nitrogen-binding agent for chronic management of patients with urea cycle disorders (UCDs) who cannot be managed by dietary protein restriction and/or supplementation alone. RAVICTI must be used with dietary protein restriction and, in some cases, dietary supplements (e.g. essential amino acids, arginine, citrulline, protein-free calorie supplements).

LIMITATIONS OF USE

- RAVICTI is not indicated for the treatment of acute hyperammonemia in patients with UCDs because more rapidly acting interventions are essential to reduce plasma ammonia levels.
- The safety and efficacy of RAVICTI for the treatment of *N*-acetylglutamate synthase (NAGS) deficiency has not been established.

Please see Important Safety Information on page 3 and accompanying Full Prescribing Information or visit RAVICTIhcp.com.

RAVICTI®
(glycerol phenylbutyrate) Oral Liquid

PA CHECKLIST FOR RAVICTI (CONT'D)

2 PA REQUIREMENTS (CONT'D)

Treatment Information

- ☐ Provide documentation if the patient is on a protein-restricted diet and/or amino acid supplementation.
- ☐ Note all previous medications, including name, dosage, and dates or duration of treatment.
- ☐ Note any contraindication, intolerance, and/or treatment failure with other nitrogen-scavenging medicines/medications.
  - – If medication was discontinued, list all reasons for discontinuation, including side effects, intolerability information, or comorbidities, if applicable.
- ☐ Documentation of infections, hospital admissions, and any additional clinical notes.
- ☐ Include a letter of medical necessity.
- ☐ Note reauthorization criteria, including documentation that shows patient improvement with treatment.

Step therapy requirements may vary between plans.

3 PA SUBMISSION

- ☐ Submit the PA directly to the health plan or by using an an electronic PA submission service such as CoverMyMeds®.
- ☐ Verify that the PA (including the number of pages) was received.
- ☐ Check with the patient's plan to see how long it typically takes for a PA to be reviewed.
- ☐ Communicate with the Horizon By Your Side team to follow up on status and to see if any additional information is required.

HORIZON BY your side

Available Monday through Friday, 8 AM to 8 PM ET

1-844-4MY-HBYS (1-844-469-4297)

SELECT IMPORTANT SAFETY INFORMATION

CONTRAINDICATIONS

- *Patients with known hypersensitivity to phenylbutyrate:* Reactions include wheezing, dyspnea, coughing, hypotension, flushing, nausea, and rash.

Please see additional Important Safety Information on page 3 and accompanying Full Prescribing Information or visit RAVICTIhcp.com.

RAVICTI® (glycerol phenylbutyrate) Oral Liquid

INDICATION and IMPORTANT SAFETY INFORMATION

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WARNINGS AND PRECAUTIONS

- *Neurotoxicity:* Phenylacetate (PAA), the major metabolite of RAVICTI, may be toxic at levels of 500 micrograms/mL or greater. If symptoms of vomiting, nausea, headache, somnolence, or confusion, are present in the absence of high ammonia or other intercurrent illness which explains these symptoms, consider the potential for PAA neurotoxicity which may need reduction in the RAVICTI dosage.
- *Pancreatic Insufficiency or Intestinal Malabsorption:* Low or absent pancreatic enzymes or intestinal disease resulting in fat malabsorption may result in reduced or absent digestion of RAVICTI and/or absorption of phenylbutyrate and reduced control of plasma ammonia. Monitor ammonia levels closely.

ADVERSE REACTIONS

The most common adverse reactions reported in clinical trials (at least 10% of patients) were:

- *Adult patients:* diarrhea, flatulence, and headache occurred during 4-week treatment (n=45) with RAVICTI; nausea, vomiting, diarrhea, decreased appetite, dizziness, headache, and fatigue occurred during 12-month treatment (n=51) with RAVICTI.
- *Pediatric patients ages 2 to 17 years:* upper abdominal pain, rash, nausea, vomiting, diarrhea, decreased appetite, and headache occurred during 12-month treatment (n=26) with RAVICTI.
- *Pediatric patients ages 2 months to less than 2 years:* neutropenia, vomiting, constipation, diarrhea, pyrexia, hypophagia, cough, nasal congestion, rhinorrhea, rash, and papule occurred during 12-month treatment (n=17) with RAVICTI.
- *Pediatric patients less than 2 months of age:* vomiting, rash, gastroesophageal reflux, increased hepatic enzymes, feeding disorder (decreased appetite, hypophagia), anemia, cough, dehydration, metabolic acidosis, thrombocytosis, thrombocytopenia, neutropenia, lymphocytosis, diarrhea, flatulence, constipation, pyrexia, lethargy, and irritability/agitation occurred during 24-month treatment (n=16) with RAVICTI.

DRUG INTERACTIONS

- Corticosteroids, valproic acid, or haloperidol may increase plasma ammonia level. Monitor ammonia levels closely.
- Probenecid may affect renal excretion of metabolites of RAVICTI, including phenylacetylglutamine (PAGN) and PAA.
- CYP3A4 substrates with narrow therapeutic index (eg, alfentanil, quinidine, cyclosporine): RAVICTI may decrease exposure to the concomitant drug.
- Midazolam: Use of RAVICTI decreased exposure of midazolam with concomitant use.

USE IN SPECIFIC POPULATIONS

- *Pregnancy:* RAVICTI should be used with caution in patients who are pregnant or planning to become pregnant. Based on animal data, RAVICTI may cause fetal harm. Report pregnancies to Horizon at 1-866-479-6742.
- *Lactation:* breastfeeding is not recommended during treatment with RAVICTI. There are no data on the presence of RAVICTI in human milk, the effects on the breastfed infant, nor the effects on milk production.

Please see accompanying Full Prescribing Information or visit RAVICTIhcp.com.

HORIZON.

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