Custom Criteria for
BlueCross Blue Shield of Rhode Island Commercial

catamaran™

RAVICTI® (glycerol phenylbutyrate)

GPI CODING:

309080300009**

DESCRIPTION:

Ravicti is indicated for use as a nitrogen-binding agent for chronic management of adult and pediatric patient's ≥2 years of age with urea cycle disorders (UCDs) that cannot be managed by dietary protein restriction and/or amino acid 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). Ravicti is not indicated for treatment of acute hyperammonemia in patients with UCDs. Safety and efficacy for treatment of N-acetylglutamate synthase (NAGS) deficiency has not been established. The use of Ravicti in patients <2 months of age is contraindicated.

APPROVAL DURATION:

Indefinite

CRITERIA FOR RAVICTI

I. The member has no contraindication to therapy including:
   A. Age younger than 2 months of age
   B. Diagnosis of acute hyperammonemia
   C. Diagnosis of N-acetylglutamate synthase (NAGS) deficiency

AND

II. The member has a confirmed diagnosis of a Urea Cycle Disorder involving deficiencies of carbamoyl phosphate synthetase (CPS), ornithine transcarbamoylase (OTC) or argininosuccinic acid synthetase (AAS) confirmed via enzymatic, biochemical, or genetic testing

AND

III. A protein-restricted diet alone or amino acid supplements alone has been ineffective

AND

IV. The member has tried and had an inadequate response to at least 3 months of therapy with Buphenyl, unless the member has had an intolerance to Buphenyl

AND

V. The member will maintain a protein-restricted diet while on Ravicti therapy