Kuvan® (sapropterin dihydrochloride)

BENEFIT APPLICATION

Benefit determinations are based on the applicable contract language in effect at the time the services were rendered. Exclusions, limitations or exceptions may apply. Benefits may vary based on contract, and individual member benefits must be verified. Wellmark determines medical necessity only if the benefit exists and no contract exclusions are applicable. This medical policy may not apply to FEP. Benefits are determined by the Federal Employee Program.

DESCRIPTION

The intent of the Kuvan® (sapropterin dihydrochloride) drug policy is to ensure appropriate selection of patients for therapy based on product labeling, clinical guidelines and clinical studies.

FDA-Approved Indication

- Kuvan is indicated to reduce blood phenylalanine (Phe) levels in patients with hyperphenylalaninemia (HPA) due to tetrahydrobiopterin- (BH4-) responsive phenylketonuria (PKU). Kuvan is to be used in conjunction with a Phe-restricted diet.

Compendial Uses

- Autosomal dominant guanine triphosphate cyclohydrolase deficiency (Segawa disease)
- Autosomal recessive guanine (GTP) cyclohydrolase deficiency
- 6-pyruvoyl-tetrahydropterin synthase (6-PTS) deficiency
- Sepiapterin reductase deficiency
- Dihydropteridine reductase (DHPR) deficiency
- Pterin-4a-carbinolamine dehydratase deficiency (also called primapterinuria)

Required Documentation

The following information is necessary to initiate the prior authorization review:

- Phenylketonuria
  - For members requesting their first course treatment following a therapeutic trial: Blood phenylalanine levels before and after the trial

POLICY

CRITERIA FOR APPROVAL

I. Phenylketonuria (PKU)

   1. Authorization of 1 month may be granted for members requesting a therapeutic trial with Kuvan when their pretreatment, including before dietary management, phenylalanine level was greater than 6 mg/dL (360 micromol/L)

   2. Authorization of 12 months may be granted for members requesting Kuvan for the first course of treatment following a therapeutic trial with Kuvan when both of the following are met:

      i. The member’s therapeutic trial meets either of the following:
a. Member experienced a reduction in blood Phe level of at least 30% during the therapeutic trial with Kuvan.
b. Member has demonstrated an improvement in neuropsychiatric symptoms during the therapeutic trial with Kuvan.

ii. Kuvan will be used in conjunction with dietary management

3. Authorization of 24 months may be granted for members requesting a second and subsequent course of treatment when Kuvan will be used in combination with dietary management

II. Biopterin Metabolic Defects

1. Authorizations of 24 months may be granted for members who have ANY of the following biopterin metabolic defects:
   - Autosomal dominant guanine triphosphate cyclohydrolase deficiency (Segawa disease)
   - Autosomal recessive guanine (GTP) cyclohydrolase deficiency
   - 6-pyruvoyl-tetrahydropterin synthase (6-PTS) deficiency
   - Sepiapterin reductase deficiency
   - Dihydropteridine reductase (DHPR) deficiency
   - Pterin-4a-carbinolamine dehydratase deficiency (also called primapterinuria)

III. Kuvan is considered not medically necessary for patients who do not meet the criteria set forth above.

CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

DOSAGE AND ADMINISTRATION

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

PROCEDURES AND BILLING CODES

To report provider services, use appropriate CPT* codes, Alpha Numeric (HCPCS level 2) codes, Revenue codes, and/or ICD diagnostic codes.

REFERENCES


POLICY HISTORY

Policy #: 05.01.65
Original Effective Date: January 2008
Reviewed: January 2017
Revised: May 2016
Current Effective Date: July 11, 2016

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