Sapropterin (Javygtor, Kuvan)

Override(s)	Approval Duration
Prior Authorization	Initial requests: 8 weeks Continued/Subsequent requests: 1 year

Medications
Javygtor (sapropterin dihydrochloride) Kuvan (sapropterin dihydrochloride)

APPROVAL CRITERIA

Initial requests for Javygtor (sapropterin dihydrochloride) or Kuvan (sapropterin dihydrochloride) (tablet, oral packet) may be approved if the following criteria are met:

I. Individual has a diagnosis of hyperphenylalaninemia (HPA) due to tetrahydrobiopterin-(BH4-) responsive* phenylketonuria (PKU); **AND**
II. Individual is using in conjunction with a phenylalanine-(PHE-) restricted diet.

*BH4-responsiveness is known or will be determined by a trial of Javygtor or Kuvan

Requests for continued use of Javygtor (sapropterin dihydrochloride) or Kuvan (sapropterin dihydrochloride) (tablet, oral packet) after the initial 8 weeks of therapy may be approved if the following criteria are met:

I. Individual is using in conjunction with a PHE-restricted diet; **AND**
II. Documentation is provided for positive response to therapy as evidenced by reduction in blood PHE levels from baseline.

Subsequent requests for continued use of Javygtor (sapropterin dihydrochloride) or Kuvan (sapropterin dihydrochloride) (tablet, oral packet) may be approved if the following criteria are met:

I. Individual is using in conjunction with a PHE-restricted diet; **AND**
II. Individual is showing signs of continuing improvement, as evidenced by maintaining acceptable blood PHE levels and/or dietary PHE allowance.

Requests for **brand** Kuvan must also meet the following criteria, in addition to the above Prior Authorization criteria:

I. Documentation is provided that individual has failed an adequate trial of one chemically equivalent generic sapropterin dihydrochloride agent. Medication samples/coupons/discount cards are excluded from consideration as a trial.; **AND**

A. Documentation is provided that generic sapropterin dihydrochloride had inadequate response; **OR**
B. Documentation is provided that generic sapropterin dihydrochloride caused adverse outcome; **OR**
C. Documentation is provided that the individual has a genuine allergic reaction to an inactive ingredient in generic agent. Allergic reaction(s) must be clearly documented in the individual's medical record.

Requests for Javygtor (sapropterin dihydrochloride) or Kuvan (sapropterin dihydrochloride) (tablet, oral packet) may not be approved for the following:

I. Individual is using in combination with Palynziq (pegvaliase-pqpz).

Key References:

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2023. URL: http://www.clinicalpharmacology.com. Updated periodically.
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2023; Updated periodically.
5. American College of Medical Genetics and Genomics Therapeutic Committee. Phenylalanine hydroxylase deficiency: diagnosis and management guideline. *Genet Med.* 2014; 16(2):188-200. doi:10.1038/gim.2013.157. Available from: http://www.nature.com/gim/journal/v16/n2/pdf/gim2013157a.pdf.