

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	X

*FHK- Florida Healthy Kids

Kuvan (sapropterin dihydrochloride)

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

Medications
Kuvan (sapropterin dihydrochloride)

APPROVAL CRITERIA

Initial requests for Kuvan (sapropterin dihydrochloride) agents (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.

Requests for continued use of Kuvan (sapropterin dihydrochloride) agents (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 blood PHE level/dietary PHE allowance. If blood PHE levels do not decrease from baseline at a dose of 10 mg/kg/day administered for up to one month, the dose may be increased to 20 mg/kg/day. Individuals whose blood PHE does not decrease after 1 month of treatment at 20 mg/kg/day are considered non-responders and treatment should be discontinued.

State Specific Mandates		
State name	Date effective	Mandate details (including specific bill if applicable)
N/A	N/A	N/A

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This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

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Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	X

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Key References:

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2018. 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>. Accessed: July 9, 2018.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2018; 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>. Accessed on: July 9, 2018.

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