

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

*FHK- Florida Healthy Kids

Kuvan (sapropterin dihydrochloride)

Override(s)
Prior Authorization

Approval Duration
<p>Initial Requests: 8 weeks</p> <p>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.</p> <p>Continued Therapy Requests: 1 year</p>

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 continues to meet the initial request approval criteria; **AND**

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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- II. Individual is showing signs of continuing improvement, as evidenced by blood Phe level/dietary Phe allowance.

Note: Treatment with Kuvan should be directed by physicians knowledgeable in the management of PKU. The initiation of therapy does not eliminate the need for appropriate monitoring by trained professionals. Individuals being treated with Kuvan should have frequent blood (PHE) measurements and nutritional counseling with their physician and other individuals of the health care team to encourage maintenance of blood (PHE) levels in the desirable range.

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

Key References:

1. 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: March 9, 2017.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Kuvan [Package insert]. Novato, CA. BioMarin Pharmaceutical Inc., 2016. Available from: http://www.accessdata.fda.gov/drugsatfda_docs/label/2016/205065s002lbl.pdf. Accessed on: March 9, 2017.
4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2017; Updated periodically.

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