Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	X

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.

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

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

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	X

Key References:

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2019. URL: http://www.clinicalpharmacology.com. Updated periodically.
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3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
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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: June 7, 2019.