



Drug Name: Kuvan® (sapropterin)

Effective Date: 12/12/2018

Last Revision Date: 6/2019

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Drug Name:	Kuvan® (sapropterin)
Prescriber Restrictions:	Must be prescribed by a physician with knowledge and experience in metabolic disease
Age Restrictions:	1 month and older
Exclusion Criteria:	
Required Medical Information:	• Patient must have a diagnosis of phenylketonuria (PKU) • Patient must see a dietician that specializes in PKU/metabolic disease (submit supporting clinical documentation) <u>For Initial Criteria:</u> • The patient failed phenylalanine restricted diet alone despite strict compliance AND • The baseline Phe level while on a Phe restricted diet Phe level [60 µmol/L = 1 mg/dL] is: ○ Greater than 6 mg/dL (360 µM/L) if less than 12 years of age OR ○ Greater than 10 mg/dL (600 µM/L) if greater than or equal to 12 and less than 18 years of age, OR ○ Greater than 15 mg/dL (900 µM/L) if greater than or equal to 18 years of age AND • The patient will have Phe blood levels measured after 1 week of therapy and periodically for up to 2 months of therapy to determine response. <u>For Continuation of therapy Criteria:</u> • The patient continues to maintain a Phe restricted diet AND • The patient has had an objective response to therapy which resulted in a 30% or more decrease in blood phenylalanine levels from baseline OR • The patient has had an objective response to therapy which resulted in blood phenylalanine levels decreasing less than 30%, but meeting NIH recommended target phenylalanine levels [patients less than 12 years of age: 2 to 6 mg/dL (120–360 µM/L); patients greater than or equal to 12 years of age: 2 to 10 mg/dL (120–600 µM/L)] AND • The dose of Kuvan does not exceed 20mg/kg/day



Notes:	If blood Phe levels do not decrease after one month, the dose should be increased to 20 mg/kg daily. If blood Phe levels do not decrease after one month of therapy with 20 mg/kg daily, sapropterin should be discontinued. A responsive patient may receive a dosage within the range of 5-20 mg/kg daily. Kuvan is to be used in conjunction with a Phe-restricted diet
Coverage Duration:	Initial: 2 months Renewals: 6 months