KUVAN Dosing and Administration

- Treatment with KUVAN should be administered orally once a day with food and at the same time of day; the tablets should be dissolved in 4–8 oz. of water or apple juice.[1]
- The starting dose of KUVAN is 10 mg/kg/day; blood Phe levels should be checked before initiation of KUVAN, after 1 week of treatment, then periodically for up to 1 month to determine response.[1]
- If blood Phe levels do not decrease from baseline at 10 mg/kg/day, the dose may be increased to 20 mg/kg/day for up to 1 month to determine response.[1]
- Once responsiveness to KUVAN has been established, KUVAN dosage may be adjusted within the range of 5 to 20 mg/kg/day, according to response to therapy.[1]
- After the dose of KUVAN is established, continued active management of dietary Phe intake is required to ensure blood Phe control and adequate nutritional balance.[1]

KUVAN is indicated to reduce blood phenylalanine (Phe) levels in patients with hyperphenylalaninemia (HPA) due to tetrahydrobiopterin- (BH4-) responsive Phenylketonuria (PKU). KUVAN is to be used in conjunction with a Phe-restricted diet.[1]

Important Safety Information

Prolonged exposure to elevated blood Phe levels in PKU patients can result in severe neurologic damage. The initiation of KUVAN therapy does not eliminate the need for careful monitoring of blood Phe levels and ongoing dietary management.[1]

Some patients receiving KUVAN can experience significant drops in blood Phe levels. Patients should be monitored closely to ensure that blood Phe levels do not fall too low.[1]

Not all patients with PKU respond to treatment with KUVAN. Response to treatment can only be determined by a therapeutic trial of KUVAN.[1]

KUVAN has not been studied in patients with liver or renal impairment. Patients who have these conditions should be carefully monitored when receiving KUVAN. Caution should be used with the administration of KUVAN to patients who are receiving levodopa and drugs that affect nitric oxide–mediated vasorelaxation or folate metabolism.[1]

The most serious adverse reactions reported during KUVAN administration (regardless of relationship to treatment) were gastritis, spinal cord injury, streptococcal infection, testicular carcinoma, and urinary tract infection. Mild to moderate neutropenia was also noted. The most common adverse reactions were headache, diarrhea, abdominal pain, upper respiratory tract infection, pharyngolaryngeal pain, vomiting, and nausea.[1]

KUVAN™

(sapropterin dihydrochloride) Tablets

KUVAN Dosing Charts

KUVAN Dose: 5 mg/kg/day	
Weight Range (kg)	Number of Tablets Required
15–29	1
30–49	2
50–69	3
70–89	4
90–109	5

KUVAN Dose: 10 mg/kg/day	
Weight Range (kg)	Number of Tablets Required
15–24	2
25–34	3
35–44	4
45–54	5
55–64	6
65–74	7
75–84	8
85–94	9
95–104	10

KUVAN Dose: 20 mg/kg/day	
Weight Range (kg)	Number of Tablets Required
15–17	3
18–22	4
23–27	5
28–32	6
33–37	7
38–42	8
43–47	9
48–52	10
53–57	11
58–62	12
63–67	13
68–72	14
73–77	15
78–82	16
83–87	17
88–92	18
93–97	19
98–102	20

*Doses above 20 mg/kg/day have not been evaluated in clinical trials.

Calculating KUVAN dose for body weights not found on chart

KUVAN is supplied in 100 mg tablets.[1] Therefore, calculate the amount of KUVAN needed based on total body weight in kilograms and divide by 100. Round to the closest number of whole tablets (eg, 110 kg at 10 mg/kg/day = 110 x 10 ÷ 100 = 11).

Reference: 1. KUVAN prescribing information, BioMarin Pharmaceutical Inc.

Please see attached full Prescribing Information.

BIOMARIN

KUVAN™
(sapropterin dihydrochloride) Tablets