PHARMACY PRE-AUTHORIZATION CRITERIA

DRUG (S)	**Kuvan (sapropterin dihydrochloride)**
POLICY #	**22134**
INDICATIONS	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 conjuction with a Phe-restricted diet.
CRITERIA	ConnectiCare considers Kuvan to be medically necessary for patients aged 1 month and older who meet all the following criteria: • Patient has documented (BH4) responsive Phenylketonuria (PKU). • Baseline blood phenylalanine (Phe) labwork must be provided: baseline phenylalanine (Phe) levels are greater than 360 micromol/dl (6 mg/dl) with dietary interventions alone • Patient must be on a Phe-restricted diet • The dose is within the range of 5 to 20 mg/kg/day
LIMITATIONS	Initial authorization: If the above criteria are met initial authorization will be given for 2 months. Continued therapy: 1. The patient has been successfully treated with Kuvan by meeting one of the following criteria: a. The patient's blood Phe levels are being maintained within acceptable range (120-260 micromol/dl [2-6 mg/dl]); **or** b. The patient has had at least a 30% decrease in blood Phe level from baseline; **and** 2. The patient continues to use a Phe-restrictive diet in conjunction with Kuvan; **and** 3. The dose is within the range of 5 to 20 mg/kg/day. Doses greater than 20 mg/kg/day have not been evaluated in clinical trials.
REFERENCES	Kuvan full prescribing information. Novato, CA BioMarin Pharmaceutical Inc; 2016.
P&T REVIEW HISTORY	3/08, 6/08, 9/09, 9/10, 12/11, 10/12, 10/13, 10/14, 11/15, 2/16, 5/17, 5/18, 5/19
REVISION RECORD	5/17, 5/19