A mobile display of creative PKU-friendly options & helpful resources

A low-Phe diet doesn’t have to be boring! Finding low-Phe options can be daunting and challenging. The PKU Pantry Table is an educational display provided by your BioMarin representative and includes PKU-friendly foods available at local grocery stores, specialty low-Phe foods, and helpful resources for PKU management, including:

- Take-home low-protein cookbooks & seasonal recipe ideas
- Nutrition tips & diet journals
- Resources focused on the importance of keeping blood Phe levels low and stable
- Information on important PKU expert guidelines
- Information on KUVAN® (sapropterin dihydrochloride) Tablets or Powder for Oral Solution

Please see reverse for Important Safety Information.
**Indication**

KUVAN® (sapropterin dihydrochloride) Tablets for Oral Use and Powder for Oral Solution are approved 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.

**Important Safety Information**

Treatment with KUVAN should be directed by physicians knowledgeable in the management of PKU. Prolonged exposure to elevated blood Phe levels in PKU patients can result in severe neurologic damage. Treat all patients with a Phe-restricted diet. The initiation of KUVAN therapy does not eliminate the need for careful monitoring of blood Phe levels and ongoing dietary management to ensure adequate Phe control and nutritional balance. Not all patients with PKU respond to treatment with KUVAN. Response to treatment can only be determined by a therapeutic trial of KUVAN.

KUVAN is not recommended in patients with a history of anaphylaxis to KUVAN. Hypersensitivity reactions, including anaphylaxis and rash have occurred. Discontinue KUVAN treatment in patients who experience anaphylaxis and initiate appropriate medical treatment. Continue dietary Phe restrictions in patients who experience anaphylaxis.

During clinical studies, gastritis was reported as a serious adverse reaction. Monitor patients for signs and symptoms of gastritis.

Monitor patients for hyperactivity.

KUVAN has not been studied in patients with liver or renal impairment. Patients who have these conditions should be carefully monitored when receiving KUVAN.

Monitor patients when co-administering KUVAN with medications known to inhibit folate metabolism, or with levodopa. Monitor patients for hypotension when co-administering KUVAN with medications known to affect nitric oxide-mediated vasorelaxation. Due to a potential for KUVAN to inhibit p-glycoprotein (P-gp) and breast cancer resistance protein (BCRP) in the gut at the therapeutic doses, monitor patients for increased systemic exposure when co-administering KUVAN with medications that are BCRP or P-gp substrates.

Frequent blood monitoring is recommended in the pediatric population.

Some patients receiving KUVAN can experience significant drops in blood Phe levels, and children younger than 7 years old treated with KUVAN doses of 20 mg/kg per day are at an increased risk for low levels of blood Phe compared with children 7 years and older. All patients should be monitored closely to ensure that blood Phe levels do not fall too low.

Patients should be advised to notify their physicians in cases of overdose.

The most common adverse reactions (incidence ≥4%) were headache, rhinorrhea, pharyngolaryngeal pain, diarrhea, vomiting, cough, and nasal congestion. Additional adverse reactions reported in connection with worldwide marketing include pharyngitis, esophageal pain, gastritis, dyspepsia, abdominal pain, nausea, and rhinitis.

To report SUSPECTED ADVERSE REACTIONS, contact BioMarin Pharmaceutical Inc. at 1-866-906-6100, or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

**Please read the attached full Prescribing Information.**