



# How KUVAN Works



## In people without PKU



## In people with PKU



## In people who respond, KUVAN works like your body's own BH4



## PKU and you

- In people without PKU, the PAH enzyme breaks down Phe
- In people *with* PKU, the PAH enzyme is either missing or not working properly, which can result in a buildup of Phe in the blood. High Phe can cause problems in the way you think, feel, and act
- KUVAN® (sapropterin dihydrochloride) Tablets for Oral Use or Powder for Oral Solution works like your body's own BH4. In people who respond, it helps the PAH enzyme convert more Phe into tyrosine and lower blood Phe levels

## Is KUVAN right for you?

KUVAN is available as Tablets for Oral Use or Powder for Oral Solution. Along with a low-Phe diet, KUVAN has been prescribed to lower blood Phe levels in many people with PKU of all ages and types.

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**Talk with your doctor to find out if KUVAN is right for you!**  
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Please see other side for Important Safety Information.

## Indication

KUVAN® (sapropterin dihydrochloride) Tablets for Oral Use and Powder for Oral Solution are approved to reduce blood Phe levels in people with a certain type of Phenylketonuria (PKU). KUVAN is to be used with a Phe-restricted diet.

## Important Safety Information

It is not possible to know if KUVAN will work for you without a trial of the medicine. Your doctor will check your blood Phe levels when you start taking KUVAN to see if the medicine is working.

Starting KUVAN does not eliminate the need for ongoing dietary management. Any change to your diet may impact your blood Phe level. Follow your doctor's instructions carefully. Your doctor and dietitian will continue to monitor your diet and blood Phe levels throughout your treatment with KUVAN **to make sure your blood Phe levels are not too high or too low**. If you have a fever, or if you are sick, your Phe level may go up. Tell your doctor and dietitian as soon as possible so they can make any necessary changes to your treatment.

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. Frequent blood monitoring is recommended in this population to ensure that blood Phe levels do not fall too low.

Tell your doctor if you have ever had liver or kidney problems, have poor nutrition or have a loss of appetite, are pregnant or plan to become pregnant, or are breastfeeding or plan to breastfeed.

KUVAN is a prescription medicine and should not be taken by people who are allergic to any of its ingredients. KUVAN and other medicines may interact with each other. Tell your doctor about **all the medicines you take**, including prescription and over-the-counter medicines, vitamins, herbal and dietary supplements.

If you forget to take your dose of KUVAN, take it as soon as you remember that day. Do not take 2 doses in a day. If you take too much KUVAN, call your doctor for advice.

The most common side effects reported when using KUVAN are headache, runny nose and nasal congestion, sore throat, diarrhea, vomiting, and cough. Additional adverse reactions reported in connection with worldwide marketing include sore throat, heartburn or pain in the esophagus, inflammation of the lining of the stomach, indigestion, stomach pain, and nausea. These are not all the possible side effects seen with KUVAN. Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

KUVAN can cause serious side effects, including:

- **Severe allergic reactions.** Stop taking KUVAN and **get medical help right away** if you develop any of these symptoms of a severe allergic reaction:
  - Wheezing or trouble breathing
  - Nausea
  - Flushing
  - Lightheadedness or fainting
  - Coughing
  - Rash
- **Inflammation of the lining of the stomach (gastritis).** Gastritis can happen with KUVAN and may be severe. **Call your doctor right away if you have any:**
  - Severe upper stomach-area discomfort or pain
  - Blood in your vomit or stool
  - Black, tarry stools
  - Nausea and vomiting
- **Too much or constant activity (hyperactivity) can happen with KUVAN.** Tell your doctor if you have any signs of hyperactivity, including fidgeting, moving around or talking too much.

For more information, call BioMarin Patient and Physician Support (BPPS) at 1-866-906-6100.

Please read the attached full Patient Information.

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**KUVAN**<sup>®</sup>  
(sapropterin dihydrochloride)  
Tablets or Powder for Oral Solution