Consider monitoring blood Phe levels more frequently during concomitant administration.

11.2 Pharmacodynamics

Sapropterin has a sigmoidal doseresponse relationship at the active site, with an apparent V_{max} of 660 μmol/L (mean 747 ± 153 μmol/L) and an AUC of 87% (dissolved in water). However, there was extensive variability among patients, with a range of 84% and an AUC of 87% (dissolved in water). The absorption of intact tablets under fed conditions resulted in an approximately 43% increase in the extent of exposure compared to fasting conditions.

Sapropterin dihydrochloride is a saline solution containing 10 mg of sapropterin dihydrochloride in each mL. The solution is clear, colorless, and nonpyrogenic.

Sapropterin dihydrochloride is a synthetic preparation of the dihydrochloride salt of naturally occurring tetrahydrobiopterin (BH4). BH4 is a metabolic intermediate produced in the body from dietary sources and is involved in the biosynthesis of several neurotransmitters, including serotonin, dopamine, and norepinephrine. BH4 is also a cofactor for several enzymes involved in the synthesis of neurotransmitters such as dopamine, norepinephrine, and serotonin.

12.2 Pharmacodynamics

Sapropterin dihydrochloride is a saline solution containing 10 mg of sapropterin dihydrochloride in each mL. The solution is clear, colorless, and nonpyrogenic.
Monitoring of blood Phe levels is important during sapropterin dihydrochloride powder for oral solution treatment.

- The mean (±SD) blood Phe levels were 744 (±384) μmol/L at baseline and 248 (±125) μmol/L at Week 6 (Figure 2).

### Precautions

- **Severe allergic reactions.**
  - As with any medicine, there is a risk of an allergic reaction. If you develop any of these symptoms of a severe allergic reaction:
    - rash
    - swelling of the face
    - breathing difficulties
    - shock
  - Call your doctor or go to the nearest emergency department right away.

### Administration

#### Preparation and Administration

- **Empty the contents of the packet(s) into 4 ounces to 8 ounces (1/2 cup to 1 cup) of water or apple juice.**
- **Step 2:**
  - Have the powder and water ready and sit with it. It is best to take sapropterin dihydrochloride powder for oral solution in the morning on an empty stomach.
- **Step 3:**
  - Take the powder in suspension. Do not chew or swallow it whole.

#### Instructions for Use

- **Read this Instructions for Use before you start taking sapropterin dihydrochloride powder for oral solution.**
- **Tell your doctor how much to give and when to give it.**
- **Step 7:**
  - Take the dose of sapropterin dihydrochloride powder for oral solution exactly as your doctor tells you. Do not take more or less.
- **Step 8:**
  - Store sapropterin dihydrochloride powder for oral solution at room temperature between 68° to 77°F (20° to 25°C).

### What to do in case of overdose

- **If you take too much sapropterin dihydrochloride powder for oral solution:***
  - Call your doctor or go to the nearest emergency department right away.

### What to do in case of missed dose

- **If you forget to take your dose of sapropterin dihydrochloride powder for oral solution:**
  - Take as soon as you remember it. Do not take 2 doses in a day.

### Side effects

- **Common side effects:***
  - Runny nose and nasal congestion
  - Hyperactivity
  - Wheezing, dyspnea, coughing, hypotension, flushing, nausea, rash

### Contraindications

- **Sapropterin dihydrochloride is contraindicated in patients:**
  - Who are allergic to sapropterin dihydrochloride or any of the ingredients in sapropterin dihydrochloride powder for oral solution
  - Who have had a severe allergic reaction to sapropterin dihydrochloride powder for oral solution

### Warnings and Precautions

- **Sapropterin dihydrochloride is not recommended in patients:**
  - With severe liver, kidney, or heart disease
  - With a history of seizures, migraine headaches, head injury, or a stroke

### Interactions

- **Tell your doctor and pharmacist about all the medicines you take including prescription and nonprescription (over-the-counter) medicines, vitamins, and herbal or dietary supplements.**

### Dosing and Monitoring

- **Sapropterin dihydrochloride powder for oral solution is available in 100 mg packets.**
  - Babies who weigh 10-20 kg: 1 packet
  - Babies who weigh 20-30 kg: 2 packets
  - Babies who weigh 30-40 kg: 3 packets

### Supplied as

- **16 packets of 100 mg (2 packets of 50 mg) for oral dosing and 4g of ascorbic acid, mannitol, potassium citrate and sucralose.**

### Additional information

- **See the detailed "Instructions for Use" that comes with sapropterin dihydrochloride powder for oral solution.**

### How should I store sapropterin dihydrochloride powder for oral solution?

- **How long will I take sapropterin dihydrochloride powder for oral solution?***
  - Treatment with sapropterin dihydrochloride powder for oral solution is given indefinitely. It may harm them.

### How should I take sapropterin dihydrochloride powder for oral solution?

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.

### How long will I take sapropterin dihydrochloride powder for oral solution?***

- **How should I take sapropterin dihydrochloride powder for oral solution?***
  - Take the powder in suspension. Do not chew or swallow it whole.