

Pheburane®

Sodium phenylbutyrate granule

Consumer Medicine Information

What is in this leaflet

This leaflet answers some common questions about Pheburane.

It does not contain all the available information. Some of the information it contains may not apply to you.

It does not take the place of talking to your doctor or pharmacist. All medicines have benefits and risks. In deciding to give you Pheburane, your doctor has weighed the risks of you taking Pheburane against the expected benefits it will have for you.

Always follow the instructions that your doctor and pharmacist give you about Pheburane.

If you have any concerns about taking this medicine, ask your doctor or pharmacist.

What is Pheburane used for

Pheburane contains the medicine sodium phenylbutyrate. Pheburane belongs to a group of medicines called nitrogen scavengers.

Nitrogen is a building block of proteins, which are an essential part of the food we eat. Waste nitrogen is converted into ammonia and removed from the body. Liver enzymes are necessary to remove ammonia from the blood stream. In patients with urea cycle disease (UCD), ammonia builds up because the body cannot remove it. Ammonia is especially toxic for the brain and leads, in severe cases, to

reduced levels of consciousness and to coma.

Pheburane granules are indicated for the management of hyperammonaemia (elevated blood ammonia) associated with urea cycle disorders. Pheburane should be used with dietary protein restriction and, in some cases, dietary supplements (e.g. essential amino acids, arginine, citrulline, and protein-free calorie supplements).

Pheburane helps the body to eliminate waste nitrogen, reducing the amount of ammonia in your body.

Sodium phenylbutyrate can be used in infants from one month of age and needs to be taken continuously throughout the patient's lifetime.

Your doctor may have prescribed this medicine for another use. If you want more information, ask your doctor.

Pheburane is only available on a doctor's prescription

Before you take Pheburane

When you must not take it

You should not take Pheburane if:

- you are allergic to sodium phenylbutyrate or any of the ingredients listed under 'Product Description' at the end of this leaflet.
- you are pregnant
- you are breastfeeding

- the packaging shows signs of tampering.
- the expiry date on the pack has passed. If you use this product after the expiry date has passed, it may not work.

Do not take Pheburane to treat any other complaint unless your doctor says it is safe. Do not give this medicine to anyone else.

Pheburane is recommended for children.

Before you start to take Pheburane

You must tell your doctor if:

- you are pregnant or intend to become pregnant. Your doctor will discuss the possible risks and benefits of taking Pheburane during pregnancy. If you are a woman who could become pregnant, you must use reliable contraception during treatment with Pheburane and should speak with your doctor.
- you suffer from congestive heart failure (a type of heart disease where the heart cannot pump enough blood around the body);
- have decreased kidney or liver function, since Pheburane is eliminated from the body through the kidney and liver;
- are diabetic or have been diagnosed with problems (ie intolerance, malabsorption or enzyme insufficiency) relating to some sugars.
- The use of Pheburane during breast feeding has not been investigated. Ask your doctor

for advice about breast feeding.

- **Hypersensitive to the active substance or to any of the excipients**

If you have not told your doctor about any of the above, tell him/her before you start taking Pheburane.

While taking Pheburane it is still possible to experience an acute excess of ammonia in the blood. If this happens you may develop symptoms such as feeling sick (nausea), being sick (vomiting), confusion, combativeness, slurred speech, difficulty walking, and even loss of consciousness. This is a medical emergency, and medical assistance should be sought immediately. An infection can cause such a situation; therefore, if you develop a fever you should seek prompt medical assistance.

Taking other medicines

Tell your doctor or pharmacist if you are taking any other medicines, including any that you buy without a prescription from your pharmacy, supermarket or health food store.

Some medicines and Pheburane may interfere with each other. These include:

- valproate, topiramate, phenobarbital, or carbamazepine (antiepileptic medicines);
- haloperidol (used in certain psychotic disorders);
- corticosteroids (medicines that are used to provide relief for inflamed areas of the body);
- probenecid (for treatment of hyperuricaemia, high levels of uric acid in the blood, associated with gout);
- rifampicin (an antibiotic).

These medicines may change the effect of Pheburane and you may need more frequent blood tests. If you are uncertain if your medicines contain these substances, you

should check with your doctor or pharmacist.

Your doctor may need to change the amount of your medicines, or you may need to take different medicines.

If you are unsure about any medicine you are taking you should check with your doctor or pharmacist. They will have more information on medicines to be careful with or avoid while taking Pheburane.

How to take Pheburane

How to take it

Your doctor will tell you the amount of Pheburane to take each day.

Take Pheburane only as prescribed by your doctor and follow his or her directions carefully. They may differ from the information contained in this leaflet.



The daily dose of Pheburane will be based on your body weight or

body surface area and adjusted according to your protein tolerance and diet. The doses of PHEBURANE prescribed by your doctor are expressed in grams of sodium phenylbutyrate. You will need regular blood tests to determine the correct daily dose.

Use only the calibrated measuring spoon provided with this medicine to measure out each dose.

- For newborns and children who weigh less than 20kg, the usual total daily dose is up to 600 mg/kg of sodium phenylbutyrate.
- For children who weigh more than 20 kg, adolescents and adults, the usual total daily dose of sodium phenylbutyrate is up to 13.0 g/m² surface area.

Your total dose per day should not exceed 20 grams of sodium phenylbutyrate.

You should take Pheburane by mouth.

You should take Pheburane with each meal or feeding. In small children this can be 4 to 6 times per day.

A calibrated measuring spoon which dispenses up to 3 g of sodium phenylbutyrate by graduation of 250 mg is provided with the medicine. Only use this measuring spoon to measure out the dose.

To measure the dose:

- Lines on the spoon indicate the amount (in grams of sodium phenylbutyrate). Take the correct amount as prescribed by your doctor.
- Pour granules directly into the spoon as shown by the picture below.



- Tap the spoon once on a table to give a horizontal level of granules and continue filling if necessary.
- If you must take more than 3 grams of sodium phenylbutyrate at once, repeat these instructions to obtain the prescribed dose.

DO NOT USE THIS MEASURING SPOON FOR ANY OTHER MEDICINES.

The granules can be directly swallowed with a drink (water, fruit juices, protein-free infant formulas) or sprinkled on to a spoonful of solid foods (mashed potatoes or apple sauce). If you mix them with food, it is important that you take it immediately. This will keep the granules from producing any taste.

Administration by nasogastric tube or gastrostomy tube:

In certain circumstances, your doctor may decide that Pheburane should be administered through nasogastric tube (a tube that goes through the nose to the stomach) or gastrostomy tube (a tube that goes through the abdomen to the stomach). In this case, Pheburane will be prepared into a liquid (50 mg/mL of sodium phenylbutyrate) by hospital or pharmacy staff following specific instructions. Granules should not be taken directly by tube. The exact amount of liquid to measure into the syringe will be determined by your doctor. Do not use the measuring spoon provided with the product to measure the liquid as it is designed to measure the granules only.

The liquid should be stored in a refrigerator between 2 and 8 degree C and used within 7 days. The liquid should be wrapped in aluminum foil to protect it from light.

Take the liquid from the refrigerator and allow to stay at room temperature at least one hour before use.

Shake well before measuring out the prescribed amount of liquid with a syringe.

The liquid must be given with a syringe by fast push directly through the tube. Rinse with water to clear the nasogastric or gastrostomy tube.

Use in children

Sodium phenylbutyrate has not been tested for safety and effectiveness in children under one month of age.

In young infants, from one month of age, the granules can be mixed with a drink (water, fruit juices, protein-free infant formulas) or sprinkled on to a spoonful of solid foods (mashed potatoes or apple sauce). If you mix them with food, it is important that you give it immediately. This will keep the granules from producing any taste.

Use in the elderly

Sodium phenylbutyrate has not been tested for safety and effectiveness in people over 65 years of age.

Take Pheburane at about the same time each day. Taking your granules at the same time each day will have the best effect. It will also help you to remember when to take it.

You should take Pheburane for as long as your doctor continues to prescribe it. This medicine is intended for long-term use. If you stop taking it the levels of ammonia may increase to the levels before treatment was started, potentially causing damage to your liver.

If you forget to take Pheburane

You should take a dose as soon as possible with your next meal. Make sure that there are at least 3 hours between two doses. Do not take a double dose to make up for a forgotten dose.

If you are not sure what to do, ask your doctor or pharmacist.

If you have trouble remembering to take your medicine, ask your pharmacist for some hints.

If you take too much (overdose)

You may experience the following symptoms if you take more Pheburane than you should:

- sleepiness,
- tiredness
- light-headedness

And less frequently:

- confusion,
- headache,
- changes in taste (taste disturbances),
- decrease in hearing,
- may have an effect on your memory
- impaired memory, and
- worsening of existing neurological conditions.

In case of drug overdose, contact a health care practitioner, hospital emergency department or regional Poison Control Centre immediately, even if there are no symptoms.

While you are taking Pheburane

Things you must do

- **Take Pheburane exactly as your doctor has prescribed**
- **Tell your doctor if you become pregnant or plan to become pregnant**
- **Tell your doctor if you decide to breast feed your baby. Your doctor may want to discuss this and change your medicine.**
- **Tell your doctor that you are taking Pheburane if you are about to start on any new medicine.**

- If you are a woman who could become pregnant, you must use reliable contraception during treatment with Pheburane and should speak with your doctor.

If you become pregnant, your doctor will monitor your pregnancy frequently and discuss with you any risks and benefits of Pheburane treatment.

Keep all your doctor or clinic appointments.

Your doctor will do laboratory tests regularly while you are Pheburane to check levels of ammonia, amino acids and serum proteins.

Your doctor will also provide nutritional advice regarding your diet while you are taking Pheburane. You must follow these instructions carefully.

Do not suddenly stop taking Pheburane without telling your doctor.

Side effects

Tell your doctor or pharmacist as soon as possible if you do not feel well while you are taking Pheburane.

Like other medicines Pheburane can cause some side effects. Most are likely to be minor and temporary. However, some may be serious and need medical attention.

Ask your doctor or pharmacist to answer any questions you may have.

Tell your doctor immediately or go to Emergency at your nearest hospital if you notice any of the following:

- allergic reaction such as rash, hives, swelling of the face, lips, tongue or throat, difficulty swallowing or breathing
- feeling sick (nausea), being sick (vomiting), confusion or drowsiness

These are serious side effects. You may need urgent medical attention.

Tell your doctor if you notice any of the following and they worry you:

- changes in menstruation or cessation of a woman's period
- reduced appetite
- body odour
- changes in taste
- low red blood cells
- low or high platelets
- low or high white blood cells
- mood changes
- headache
- stomach ache
- rash
- changes in the blood including levels of pH (more or less acidic than normal), proteins, enzymes and electrolytes.

Other side effects not listed above may also occur in some patients. Tell your doctor if you notice any other effects.

Do not be alarmed by this list of side effects. Most people do not experience any of them.

After using Pheburane

Storage

Keep your granules in the bottle until it is time to take them.

Once the bottle is opened, the granules must be used within 45 days.

If you take your granules out of the bottle they will not keep well.

Keep Pheburane in a cool, dry place where the temperature stays below 30 degrees C.

Pheburane solution for nasogastric or gastrostomy administration should be stored between 2 and 8 degrees C and used within 7 days.

Protect from light.

Do not store Pheburane or any other medication in the bathroom or near a sink.

Keep Pheburane where young children cannot reach it.

A locked cupboard at least one and a half metres above the ground is a good place to store medicines.

Do not use after the expiry date which is stated on the carton and bottle label after "EXP". The expiry date refers to the last day of that month.

Disposal

If your doctor tells you to stop taking Pheburane, ask your pharmacist what to do with any granules that are left over.

Product description

What it looks like

Pheburane consists of white to off-white tasteless coated granules. Each gram of granules contains 483 mg/g of sodium phenylbutyrate.

AUST R 273750

Active Ingredient:

Pheburane contains sodium phenylbutyrate 483mg/g

Other Ingredients:

Ethylcellulose
 Hypromellose
 Macrogol 1500
 Maize starch
 Povidone
 Sucrose

Manufacturer

Pheburane is supplied in Australia by:

Orpharma Pty Ltd
 Level 1,
 530 Little Collins St,
 Melbourne VIC 3000,
 AUSTRALIA

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