Package leaflet: Information for the patient

PHEBURANE 483 mg/g granules
Sodium phenylbutyrate

Read all of this leaflet carefully before you start taking this medicine because it contains important information for you.
- Keep this leaflet. You may need to read it again.
- If you have any further questions, ask your doctor or pharmacist.
- This medicine has been prescribed for you only. Do not pass it on to others. It may harm them, even if their signs of illness are the same as yours.
- If you get any side effects, talk to your doctor or pharmacist. This includes any possible side effects not listed in this leaflet. See section 4.

What is in this leaflet:

1. What PHEBURANE is and what it is used for

PHEBURANE contains the active substance sodium phenylbutyrate which is used to treat patients of all ages with urea cycle disorders. These rare disorders are due to a deficiency of certain liver enzymes which are necessary to eliminate waste nitrogen in the form of ammonia.

Nitrogen is a building block of proteins, which are an essential part of the food we eat. As the body breaks down protein after eating, waste nitrogen, in the form of ammonia, accumulates because the body cannot eliminate it. Ammonia is especially toxic for the brain and leads, in severe cases, to reduced levels of consciousness and to coma.

PHEBURANE helps the body to eliminate waste nitrogen, reducing the amount of ammonia in your body. However PHEBURANE must be used along with a diet reduced in proteins, designed especially for you by the doctor and the dietician. You must follow this diet carefully.

2. What you need to know before you take PHEBURANE

Do not take PHEBURANE if you:
- are allergic to sodium phenylbutyrate or any of the other ingredients of this medicine (listed in section 6).
- are pregnant.
- are breast-feeding.

Warnings and precautions
Talk to your doctor or pharmacist before taking PHEBURANE if you:
- suffer from congestive heart failure (a type of heart disease where the heart cannot pump enough blood around the body) or a decrease in your kidney function.
- have decreased kidney or liver function, since PHEBURANE is eliminated from the body through the kidney and liver.

PHEBURANE will not prevent the occurrence of an acute excess of ammonia in the blood, a condition which usually constitutes a medical emergency. If this happens you will develop symptoms such as feeling sick (nausea), being sick (vomiting), confusion and will need to get urgent medical help.

If you need laboratory tests, it is important to remind your doctor that you are taking PHEBURANE, since sodium phenylbutyrate may interfere with certain laboratory test results (such as blood electrolytes or protein, or liver function tests)

In case of any doubt, ask your doctor or pharmacist.

Other medicines and PHEBURANE
Tell your doctor or pharmacist if you are taking, have recently taken or might take any other medicines.

It is especially important to tell your doctor if you are taking medicines containing:
- valproate (an antiepileptic medicine),
- 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)

These medicines may change the effect of PHEBURANE and you will need more frequent blood tests. If you are uncertain if your medicines contain these substances, you should check with your doctor or pharmacist.

Pregnancy and breast-feeding
Do not use PHEBURANE if you are pregnant, because this medicine can harm your unborn baby.

If you are a woman who could get pregnant, **you must use reliable contraception, during treatment with PHEBURANE**. Talk to your doctor for the details.

Do not use PHEBURANE if you are breast-feeding, because this medicine can pass into the breast milk and may harm your baby.

Driving and using machines
PHEBURANE is unlikely to affect your ability to drive and use machines.

PHEBURANE contains sodium and sucrose
This medicine contains 124 mg (5.4 mmol) of sodium per 1 g of sodium phenylbutyrate. Talk to your doctor or pharmacist if you need 3 or more grams daily for a prolonged period, especially if you have been advised to follow a low salt (sodium) diet.

This medicine contains 768 mg of sucrose per 1 g of sodium phenylbutyrate. This should be taken into account if you have diabetes mellitus. If you have been told by your doctor that you have an intolerance to some sugars, contact your doctor before taking this medicine.

3. How to take PHEBURANE

Always take this medicine exactly as your doctor has told you. Check with your doctor or pharmacist if you are not sure.

Dosage
The daily dose of PHEBURANE will be based on your body weight or body surface and adjusted according to your protein tolerance and diet. You will need regular blood tests to determine the correct daily dose. Your doctor will tell you the amount of granules you should take.

Method of administration
You should take PHEBURANE by mouth. Because it dissolves slowly, PHEBURANE should not be administered through a gastrostomy (tube that goes through the abdomen to the stomach) or through a nasogastric tube (tube that goes through the nose to the stomach).

PHEBURANE must be taken with a special diet reduced in protein.

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

A calibrating measuring spoon which dispenses up to 3 g of sodium phenylbutyrate 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 gram of sodium phenylbutyrate). Take the correct amount as prescribed by your doctor.
- Pour granules directly into the spoon as shown by the picture (on the outer carton and at the end of/page 2 of this leaflet)
- Tap the spoon once on a table to give a horizontal level of granules and continue filling if necessary

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.

You will need to take this medicine and to follow a diet throughout your life.

If you take more PHEBURANE than you should
Patients who have taken very high doses of sodium phenylbutyrate experienced:
- sleepiness, tiredness, light-headedness and less frequently confusion,
- headache,
- changes in taste (taste disturbances),
- decrease in hearing,
- disorientation,
- impaired memory,
- worsening of existing neurological conditions.

If you experience any of these symptoms, you should immediately contact your doctor or the nearest hospital emergency department for supportive treatment.

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 have any further questions on the use of this medicine, ask your doctor or pharmacist.

4. Possible side effects

Like all medicines, this medicine can cause side effects, although not everybody gets them.

If persistent vomiting occurs, you should contact your doctor immediately.

Very common side effects (may affect more than 1 in 10 people): irregular menstrual periods and stopping of menstrual periods in fertile women.
If you are sexually active and your period stops altogether, do not assume that this is caused by PHEBURANE. If this occurs, please discuss it with your doctor, because the absence of your period may be caused by pregnancy (see 'Pregnancy and breast-feeding' section above) or by menopause.

Common side effects (may affect more than 1 in 100 people): changes in number of blood cells (red cells, white cells and platelets), changes in the amount of bicarbonate in the blood, reduced appetite, depression, irritability, headache, fainting, fluid retention (swelling), changes in taste (taste disturbances), stomach ache, vomiting, nausea, constipation, abnormal skin odor, rash, abnormal kidney function, weight gain, altered laboratory test values.

Uncommon side effects (may affect more than 1 in 1,000 people): deficiency in red blood cells due to failure of the bone marrow, bruising, altered heart rhythm, rectal bleeding, inflammation of the stomach, stomach ulcer, inflammation of the pancreas.

Reporting of side effects
If you get any side effects, talk to your doctor or pharmacist. This includes any possible side effects not listed in this leaflet. You can also report side effects directly via Yellow Card Scheme Website: www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store.

By reporting side effects you can help provide more information on the safety of this medicine.

5. How to store PHEBURANE

Keep this medicine out of the sight and reach of children.

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

After the first opening, PHEBURANE can be used within 45 days.

Do not throw away any medicines via wastewater or household waste. Ask your pharmacist how to throw away medicines you no longer use. These measures will help protect the environment.

6. Contents of the pack and other information

What PHEBURANE contains
The active substance is sodium phenylbutyrate.
Each gram of granules contains 483 mg of sodium phenylbutyrate.
The other ingredients are: sugar spheres (sucrose and maize starch, see section 2 'PHEBURANE contains sucrose), hypromellose, ethylcellulose N7, macrogol 1500, povidone K25.

What PHEBURANE looks like and contents of the pack
PHEBURANE granules are white to off-white.

The granules are packaged in a plastic bottle with child-resistant closure and a dessicant.
Each bottle contains 174g of granules.
Each carton contains 1 bottle.

A calibrating measuring spoon is provided.

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Detailed information on this medicine is available on the European Medicines Agency web site: http://www.ema.europa.eu.

There are also links to other websites about rare diseases and treatments.