



# Sodium phenylbutyrate for urea cycle disorders

This leaflet is about the use of sodium phenylbutyrate for urea cycle disorders.



This leaflet has been written specifically for parents and carers about the use of this medicine in children. The information may differ from that provided by the manufacturer. Please read this leaflet carefully. Keep it somewhere safe so that you can read it again.

## Name of drug

**Sodium phenylbutyrate**

**Brand name:** Ammonaps®, Pheburane®

## Why is it important for my child to take this medicine?

When protein from food is digested, it is broken down into small molecules called amino acids, which are needed by all the cells in the body. If there is too much amino acid in the blood, it is turned to ammonia in the liver, and then into uric acid, which the body gets rid of in urine (wee).

Urea cycle disorders are inherited disorders where ammonia is not turned into uric acid properly and so it builds up in the blood. Your child needs to take sodium phenylbutyrate regularly to help reduce the levels of ammonia in their body.

## What is sodium phenylbutyrate available as?

- **Tablets:** 500 mg
- **Granules:** 483 mg or 940 mg in 1 g
- **Liquid medicine** can be ordered specially from your pharmacist: 1.25 g in 5 mL

## When should I give sodium phenylbutyrate?

Sodium phenylbutyrate is usually given **three or four times** a day.

- **Three times a day:** this should be in the morning, early afternoon and at bedtime. Ideally, these times are at least 6 hours apart, for example 8 am, 2 pm and 8 pm.
- **Four times a day:** this is usually first thing in the morning, at about midday, late in the afternoon and at bedtime. Ideally, these times should be at least 4 hours apart, for example 8 am, midday, 4 pm and 8 pm.

Give the medicine at about the same time(s) each day so that this becomes part of your child's daily routine, which will help you to remember.

## How much should I give?

Your doctor will work out the amount of sodium phenylbutyrate (the dose) that is right for *your* child. The dose will be shown on the medicine label.



**It is important that you follow your doctor's instructions about how much to give.**

## How should I give it?

Ideally, give sodium phenylbutyrate to your child after a meal, some food or milk, as it is less likely to upset their stomach. Avoid giving it on an empty stomach.



**Tablets** should be swallowed with a glass of water, milk or juice. Your child should not chew the tablet.



**Granules:** Sprinkle or stir the granules into a small amount of soft food (e.g. yogurt or feed). Your child should then swallow the food straight away, without chewing. Make sure that they take it all.



**Liquid medicine:** Measure out the right amount using an oral syringe or medicine spoon. You can get these from your pharmacist. Do not use a kitchen teaspoon as it will not give the right amount.

## When should the medicine start working?

This medicine should start to work straight away. You may not see an obvious difference but your child should start to feel better as the amount of ammonia in their body falls.

## What if my child is sick (vomits)?

- If your child is sick less than 30 minutes after having a dose of sodium phenylbutyrate, give them the same dose again.
- If your child is sick more than 30 minutes after having a dose of sodium phenylbutyrate, you **do not** need to give them another dose. Wait until the next normal dose.

If your child is sick again, seek advice from your GP, pharmacist or hospital. They will decide what to do based on your child's condition and the specific medicine involved.

## What if I forget to give it?

Do not give the missed dose. Just give the next dose as usual.

## What if I give too much?

You are unlikely to do harm if you give an extra dose of sodium phenylbutyrate by mistake.

If you are concerned that you may have given your child too much, contact your doctor or local NHS services (111 in England and Scotland; 0845 4647 in Wales). Have the medicine or packaging with you if you telephone for advice.

## Are there any possible side-effects?

We use medicines to make our children better, but sometimes they have other effects that we don't want (side-effects).

- Girls may find their periods become irregular or stop. If this happens, contact your doctor.

The following side-effects should settle down after a few days, as your child gets used to the medicine; if you are worried, or any of them are still a problem after one week, contact your doctor, but continue to give sodium phenylbutyrate:

- Your child may feel less hungry (lose their appetite). Encourage them to eat small meals often
- Your child may say they have a metallic or bitter taste in their mouth. Eating citrus fruits (e.g. oranges) and taking sips of water may help

- your child may have an unpleasant body odour and bad breath. Using breath fresheners and deodorant will help.

There may, sometimes, be other side-effects that are not listed above. If you notice anything unusual and are concerned, contact your doctor. You can report any suspected side-effects to a UK safety scheme at <http://www.mhra.gov.uk/yellowcard>.

### Can other medicines be given at the same time as sodium phenylbutyrate?

- You can give your child medicines that contain paracetamol or ibuprofen, unless your doctor has told you not to.
- Check with your doctor or pharmacist **before** giving any other medicines to your child. This includes herbal or complementary medicines.

### General advice about medicines

- Try to give medicines at about the same times each day, to help you remember.
- If you are not sure a medicine is working, contact your doctor but continue to give the medicine as usual in the meantime. Do not give extra doses, as you may do harm.
- Only give this medicine to your child. Never give it to anyone else, even if their condition appears to be the same, as this could do harm.
- If you think someone else may have taken the medicine by accident, contact your doctor.
- Make sure that you always have enough medicine. It may take up to 3 weeks for your pharmacist to order sodium phenylbutyrate, so make sure you order a new prescription at least 3 weeks before you will run out.
- Make sure that the medicine you have at home has not reached the 'best before' or 'use by' date on the packaging. Give old medicines to your pharmacist to dispose of.

### Where should I keep this medicine?

- Keep the medicine in a cupboard, away from heat and direct sunlight. It does not need to be kept in the fridge.
- Make sure that children cannot see or reach the medicine.
- Keep the medicine in the container it came in.

### Who to contact for more information

Your child's doctor, pharmacist or nurse will be able to give you more information about sodium phenylbutyrate and about other medicines used to treat urea cycle disorders.

You can also get useful information from:

#### England

NHS 111: 111- [www.nhs.uk](http://www.nhs.uk)

#### Scotland - NHS 24

NHS 24: 111 - [www.nhs24.com](http://www.nhs24.com)

#### Wales/Galw Iechyd Cymru - NHS Direct

NHS Direct: 0845 4647 - [www.nhsdirect.wales.nhs.uk](http://www.nhsdirect.wales.nhs.uk)

#### Northern Ireland

NI Direct: [www.nidirect.gov.uk](http://www.nidirect.gov.uk)

#### Climb (Children Living with Inherited Metabolic Diseases): National information Center for Metabolic Diseases

[www.climb.org.uk](http://www.climb.org.uk) - 0800 652 3181

[www.medicinesforchildren.org.uk](http://www.medicinesforchildren.org.uk)



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The primary source for the information in this leaflet is the British National Formulary for Children. For details on any other sources used for this leaflet, please contact us through our website, [www.medicinesforchildren.org.uk](http://www.medicinesforchildren.org.uk)

We take great care to make sure that the information in this leaflet is correct and up-to-date. However, medicines can be used in different ways for different patients. It is important that you ask the advice of your doctor or pharmacist if you are not sure about something. This leaflet is about the use of these medicines in the UK, and may not apply to other countries. The Royal College of Paediatrics and Child Health (RCPCH), the Neonatal and Paediatric Pharmacists Group (NPPG), WellChild and the contributors and editors cannot be held responsible for the accuracy of information, omissions of information, or any actions that may be taken as a consequence of reading this leaflet.