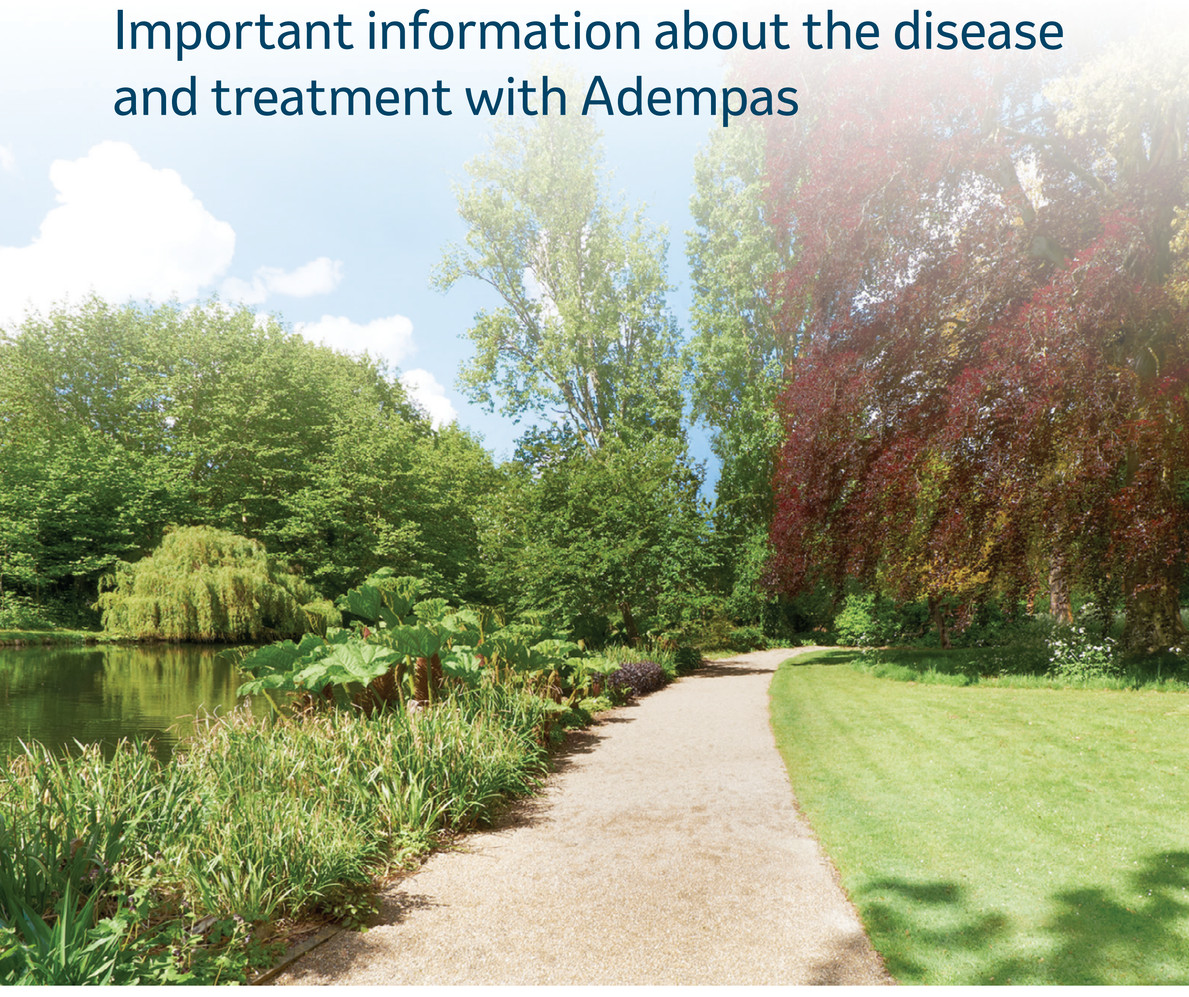
This guide is for adult patients who have been prescribed ADEMPAS® (riociguat) living with chronic thromboembolic pulmonary hypertension (CTEPH)

Important information about the disease and treatment with Adempas



Reporting of side effects
If you get any side effects, talk to your doctor, pharmacist or nurse.
This includes any possible side effects not listed in the package leaflet.
You can also report side effects directly via the Yellow Card Scheme at **yellowcard.mhra.gov.uk**
By reporting side effects, you can help provide more information on the safety of this medicine.

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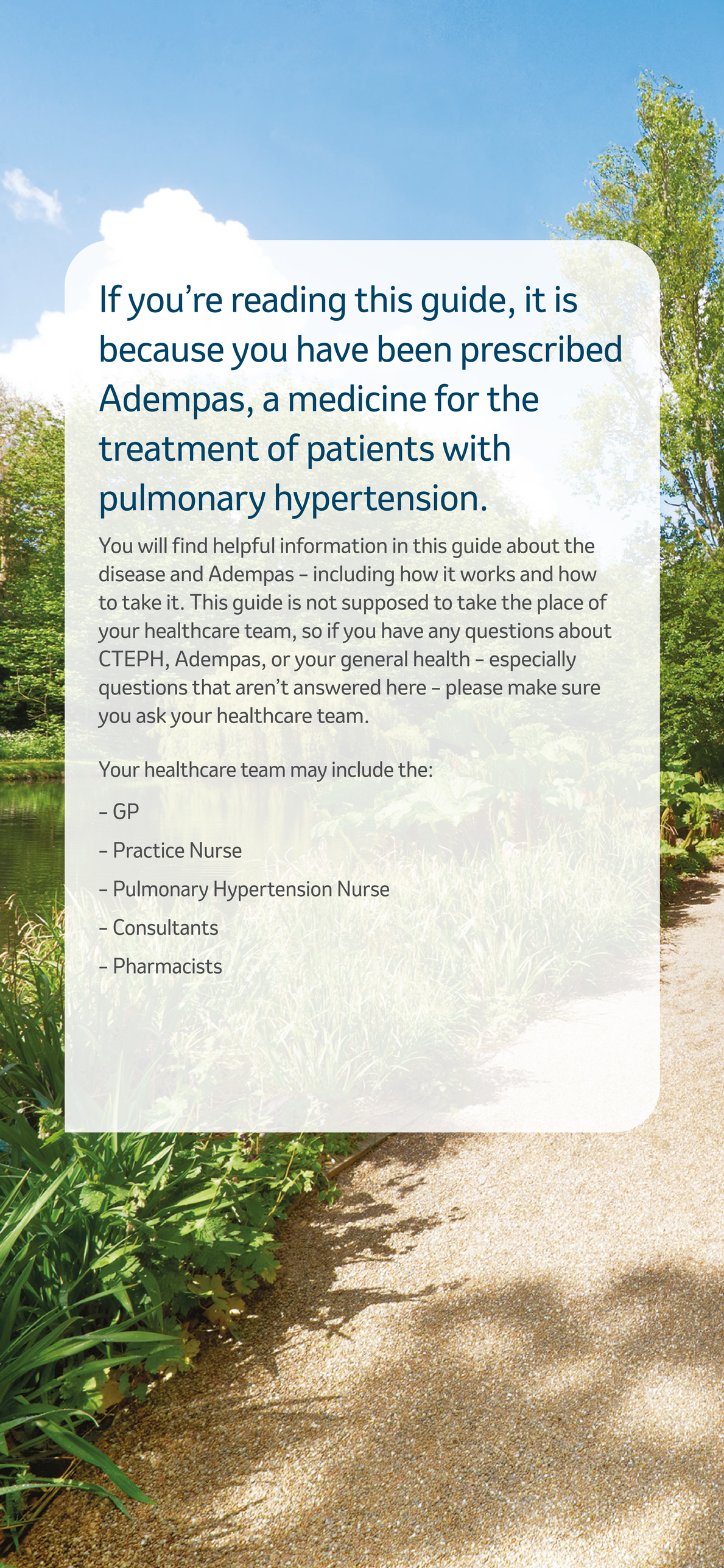

If you’re reading this guide, it is because you have been prescribed Adempas, a medicine for the treatment of patients with pulmonary hypertension.

You will find helpful information in this guide about the disease and Adempas – including how it works and how to take it. This guide is not supposed to take the place of your healthcare team, so if you have any questions about CTEPH, Adempas, or your general health – especially questions that aren’t answered here – please make sure you ask your healthcare team.

Your healthcare team may include the:

- GP
- Practice Nurse
- Pulmonary Hypertension Nurse
- Consultants
- Pharmacists

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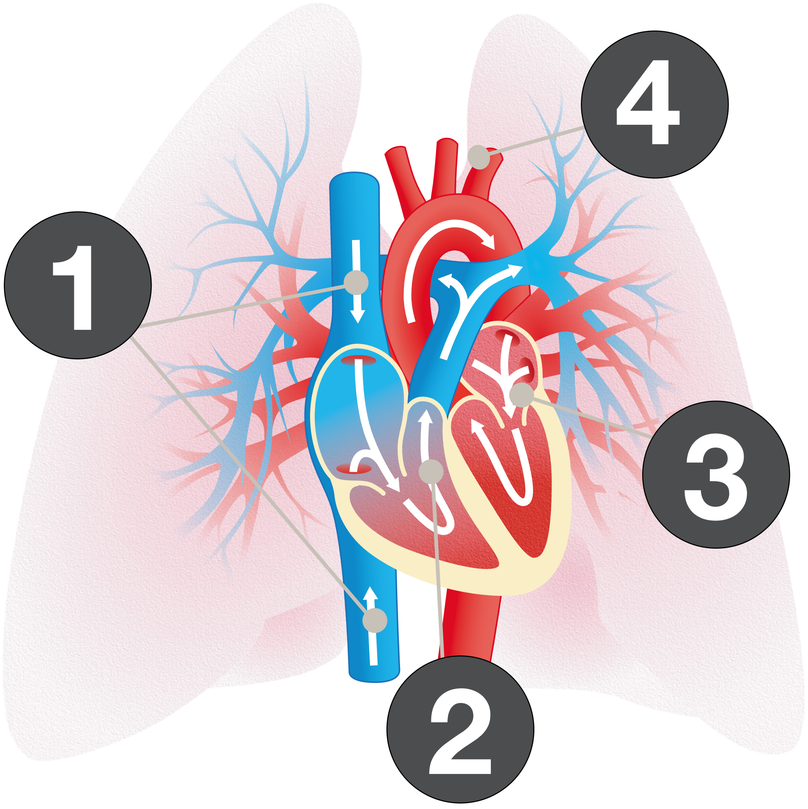
What is pulmonary hypertension (PH)?[1]

PH is the term for a group of diseases that cause the arteries in your lungs (pulmonary arteries) to tighten, which makes it harder for the heart to pump blood to the lungs. It is a condition that can be hard to diagnose, and it usually gets worse without treatment. But there are a number of treatment options available that may help to improve symptoms. Ask your PH team for more information.

The pulmonary arteries move blood from your heart to your lungs to pick up oxygen. But when they are blocked or narrowed, the blood has to move through a smaller space. In the same way that water pressure increases if you squeeze a garden hose, this blockage causes the blood pressure to rise in your pulmonary arteries.

Click each step to view the flow of blood



Key Facts

When the blood pressure in your pulmonary arteries goes up, your heart's right chamber has to work harder than normal to pump blood to the lungs.

If your lungs and right heart chamber are not working properly, you may have symptoms, including shortness of breath.

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What does pulmonary hypertension feel like?[1]

During the early stages of PH, you might not feel like anything is wrong. It is also common for patients to blame their symptoms on getting older or on a lack of exercise. But, as the heart gets weaker you might have any of the symptoms below:

Shortness of breath with exercise

Feeling tired or dizzy

Swelling in the legs or stomach area

Fainting

Chest pain

Over time, your symptoms may become more noticeable, making it harder to do normal daily activities and chores.

What happens in CTEPH?[2]

CTEPH is a form of pulmonary hypertension defined as:

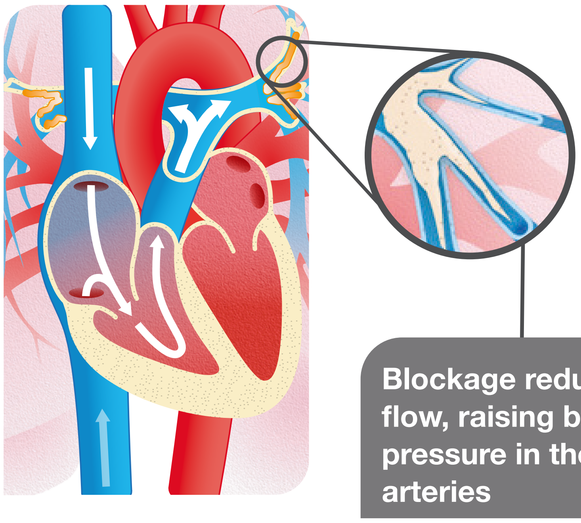
Chronic
refers to an illness that lasts a long time

Thromboembolic
is defined as a blood clot traveling to other parts of the body, which in CTEPH involves the blood clot traveling to and blocking the arteries of the lungs

Pulmonary
means relating to the lungs

Hypertension
is the medical term for high blood pressure[1]

Healthy blood vessels tighten and relax according to the blood flow needs of the body.[1] In people with CTEPH, however, blockages within the pulmonary arteries (arteries in the lungs) reduce the blood flow, raising blood pressure in these arteries.



Blockage reduces blood flow, raising blood pressure in the pulmonary arteries

In some cases, it is possible the blockage may be removed with an operation called a pulmonary endarterectomy (PEA). For other patients, the pulmonary arteries may be widened in a procedure called balloon pulmonary angioplasty (BPA) or using medical therapy.

There are a number of things your PH team will need to consider in order to appropriately diagnose your condition and evaluate what needs to be done to help you.

Ask your PH team if you would like to find out more about what to expect from the treatment options for CTEPH.

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What does World Health Organization functional class (WHO FC) mean?[3]

Your WHO FC helps your PH team figure out how CTEPH is affecting you.

Telling your doctor about your symptoms will help them decide which WHO FC you are in.

The aim of treatment is to improve symptoms. One of the goals is getting you to, or keeping you in, WHO FC I or II.

Functional class description

Class: I	Symptom-free when physically active or resting
Class: II	No symptoms at rest, but normal activities, such as climbing the stairs, grocery shopping, or making the bed cause some discomfort and shortness of breath
Class: III	Resting may be symptom-free, but less than normal activities cause significant symptoms, such as shortness of breath or feeling tired
Class: IV	Symptoms at rest and severe symptoms with any activity

What is Adempas?

Adempas is a medicine for the treatment of chronic thromboembolic pulmonary hypertension (CTEPH). Adempas can be used for adult patients with CTEPH who cannot be operated on, or after surgery for patients in whom increased blood pressure in the lungs remains or returns.

Adempas contains the active substance riociguat. Adempas is a type of medicine called a soluble guanylate cyclase (sGC)-stimulator. It is designed to widen the pulmonary arteries (the blood vessels that connect the heart to the lungs), making it easier for the heart to pump blood through the lungs.

Adempas can be used to treat adults with certain forms of pulmonary hypertension. Because the heart must work harder than normal, people with pulmonary hypertension feel tired, dizzy and short of breath. By widening the narrowed arteries, Adempas may lead to an improvement in your ability to carry out physical activity.

Designed to widen arteries

Aims to increase ability to do physical activity

Aims to decrease WHO FC

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What are the possible side effects of Adempas?

When you take Adempas, you do not need to make any changes to your diet. But, like all medicines, it can cause side effects. Because everybody is different, it is not possible to say which, if any, side effects you will have, or how serious they might be. You should discuss the possible benefits and risks of Adempas with the team at your specialist centre before you start treatment.

The following side effects may occur in people taking Adempas, although these will vary from person to person:

Very common ≥1/10:

- headache
- dizziness
- indigestion
- swelling of the limbs
- nausea
- diarrhoea
- vomiting

Common ≥1/100 to <1/10:

- inflammation in the digestive system (gastroenteritis)
- reduction of red blood cells (anaemia), seen as pale skin, weakness or breathlessness
- awareness of an irregular, hard, or rapid heartbeat
- feeling dizzy or faint when standing up (caused by low blood pressure)
- coughing up blood
- nose bleed
- difficulty breathing through your nose (nasal congestion)
- pain in the stomach, intestine or abdomen
- heartburn
- difficulty in swallowing
- constipation
- bloating
- reflux

Serious possible side effects:

- Coughing-up blood (common)
- Acute bleeding from the lungs (uncommon)

  If this happens contact your doctor immediately as you may need urgent medical treatment

If you experience any of these side effects, or any that are not listed here, please let your PH team know. Reducing the Adempas dose - in consultation with your doctor - may improve any treatment-related side effects.

Consult the patient information leaflet that came with your medication for further information. If you have any further questions on the use of this medicine, ask your PH team.

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What do I need to consider before taking Adempas?

Warnings and precautions

Your doctor will check if Adempas is suitable for you.
However do talk to your doctor or pharmacist before taking Adempas if:

- you have recently experienced serious bleeding from the lung, or if you have undergone treatment to stop coughing up blood (bronchial arterial embolisation)
- you take blood-thinning medicines (anticoagulants)
- you feel short of breath during treatment with this medicine
- you have problems with your heart or circulation
- you are older than 65 years
- your kidneys do not work properly or if you are on dialysis
- you have moderate liver problems (hepatic impairment)
- you have been prescribed medicines outside of your PH centre, to check for interactions with Adempas
- you have pulmonary veno-occlusive disease (PVOD)

Special considerations

Other medicines and Adempas

Tell your doctor or pharmacist if you are taking, have recently taken or might take any other medicines. Consult the patient information leaflet accompanying your pack of Adempas tablets for a list of potential interacting medicines.

Smoking

Smoking may reduce the effectiveness of Adempas. If you smoke, it is recommended that you stop. Inform your doctor if you smoke or stop smoking during your treatment.

Birth control

Women and female adolescents of childbearing potential must use effective contraception during treatment with Adempas.

Pregnancy

Do not take Adempas during pregnancy. You are also advised to take monthly pregnancy tests. If you are pregnant, think you may be pregnant or are planning to have a baby, ask your doctor or pharmacist for advice before taking this medicine.

Breastfeeding

If you are breastfeeding or planning to breastfeed, consult your doctor or pharmacist before taking this medicine. Stop breastfeeding whilst taking Adempas.

Driving and operating machinery

Adempas moderately influences the ability to cycle, drive and use machines. It may cause side effects such as dizziness. You should be aware of the side effects of this medicine before cycling, driving or using machines.

Adempas contains lactose and sodium

If you have been told by a doctor that you have an intolerance to some sugars, tell your treating doctor before taking these tablets. Adempas is essentially sodium free.

Do NOT take Adempas:

- if you are taking certain medicines called PDE-5 inhibitors (e.g. sildenafil, tadalafil, vardenafil). These are medicines used for the treatment of high blood pressure in the arteries of the lungs (PAH) or erectile dysfunction
- if you have severe liver problems (severe hepatic impairment, Child Pugh C)
- if you are allergic to Adempas or any of the other ingredients of this medicine
- if you are pregnant
- if you take other medicines, similar to Adempas (soluble guanylate cyclase stimulators, such as vericiguat)
- if you are taking nitrates or nitric oxide donors (such as amyl nitrite) in any form, medicines often used to treat high blood pressure, chest pain or heart disease.
- if you have low blood pressure (systolic blood pressure less than 95 mmHg) before starting first treatment with this medicine
- if you have increased blood pressure in your lungs associated with scarring of the lungs, of unknown cause (idiopathic pulmonary pneumonia)

If any of these applies to you, talk to your doctor first and do not take Adempas

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How do I take Adempas?

Adempas therapy is tailored so that your doctor can change the dose to meet your needs. By checking your blood pressure and any treatment-related side effects, your PH team can see how you are responding to your treatment.

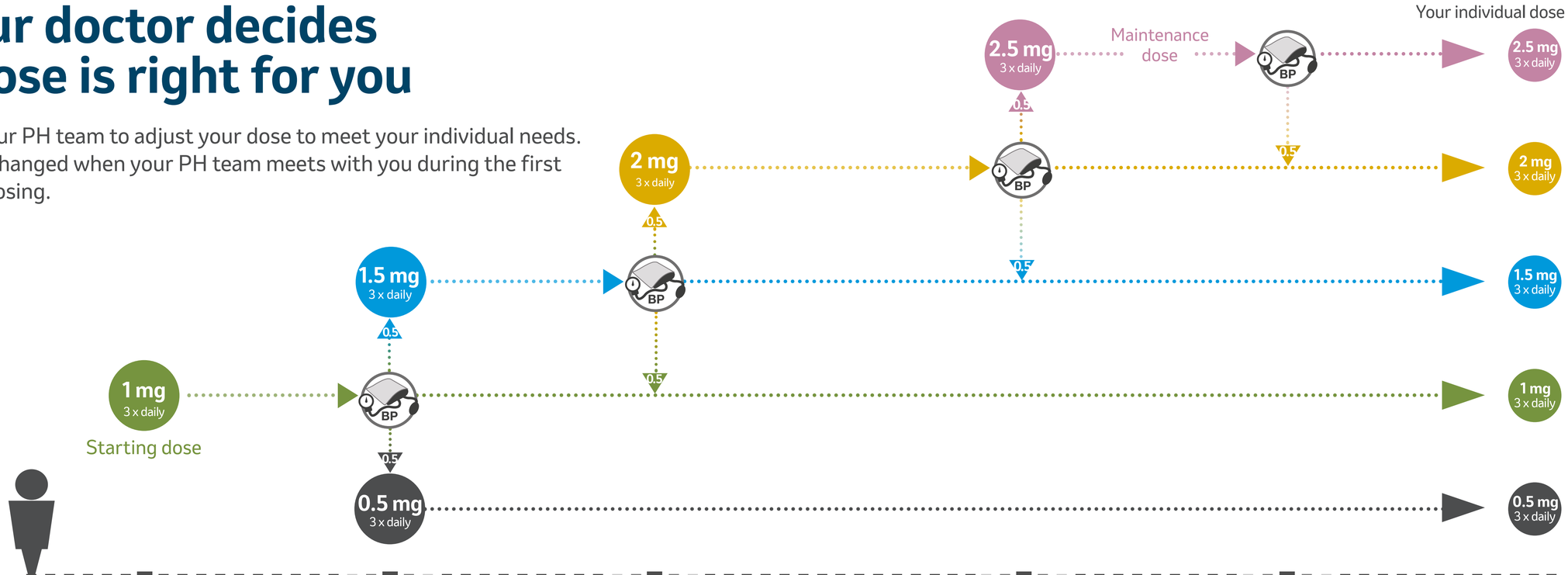
Every two weeks during initiation, you will be checked to see if it is best to increase, decrease, or maintain your dose. Your doctor will continue to monitor you until they have found the correct Adempas dose for you.

- Adempas is taken orally as a tablet three times a day, with each dose 6-8 hours apart.
- The Adempas starting dose is 1 mg three times a day, taken with or without food.
- Adempas is available in the following doses:

- To get the best results from Adempas, it is very important that you take it every day as prescribed. You should not stop taking Adempas without talking to your doctor first.
- If you forget a tablet, do not take two tablets together – just take the next single tablet as planned.
- In case treatment has to be interrupted for three days or more, treatment should be restarted with 1 mg three times daily for two weeks, and continued with the dose titration regimen as described above.

How your doctor decides which dose is right for you

Adempas allows your PH team to adjust your dose to meet your individual needs. Your dose may be changed when your PH team meets with you during the first 8-week period of dosing.



Day 1

Treatment begins after an assessment with your PH health care team

Week 2 Week 4 Week 6

Every two weeks, after each assessment, your PH team will discuss with you if your Adempas dose should be raised or decreased by 0.5 mg 3 times a day, or kept the same

Week 8

After eight weeks, you will know the dose that is right for you

Guide to the symbols

BP

Treatment assessment during initial period

- Every two weeks you will be checked for side effects and to see if your blood pressure is too low
- At each assessment your Adempas dose can be raised or lowered by 0.5 mg 3 times a day or kept the same

Your dose is raised by 0.5 mg 3 times a day if your systolic blood pressure (the top number) is 95 mm Hg or higher and you have no symptoms of low blood pressure

Your dose stays the same if your systolic blood pressure is less than 95 mm Hg and you have no signs or symptoms of low blood pressure

- Your dose is lowered by 0.5 mg 3 times a day if your systolic blood pressure is less than 95 mm Hg and you do have symptoms of low blood pressure
- If your dose is lowered, it can still be raised by 0.5 mg 3 times a day when you next meet with your PH team

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Where can I get more information?

The following websites and resources offer additional support for people diagnosed with CTEPH:

PHA UK | **www.phauk.org**

PHA Europe | **www.phaeurope.org**

References

1. NHS. Pulmonary hypertension. Available at: *www.nhs.uk/conditions/pulmonary-hypertension/* Accessed March 2026.

2. PHA UK. Chronic Thromboembolic Pulmonary Hypertension (CTEPH). Available at: *www.phauk.org/about-ph-2/chronic-thromboembolic-pulmonary-hypertension-cteph/* Accessed March 2026.

3. PHA UK. The WHO functional class. Available at: *www.phauk.org/financial-support-benefits/the-who-functional-class/* Accessed June 2026.

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