

This guide is for adult patients who have been prescribed **ADEMPAS (riociguat) living with pulmonary arterial hypertension (PAH)**

Important information about the disease and treatment with Adempas



Reporting 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 www.mhra.gov.uk/yellowcard.

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

This material has been developed and produced by MSD



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

You will find helpful information in this booklet about the disease and Adempas – including how it works and how to take it. This booklet is not supposed to take the place of your healthcare team, so if you have any questions about PAH, 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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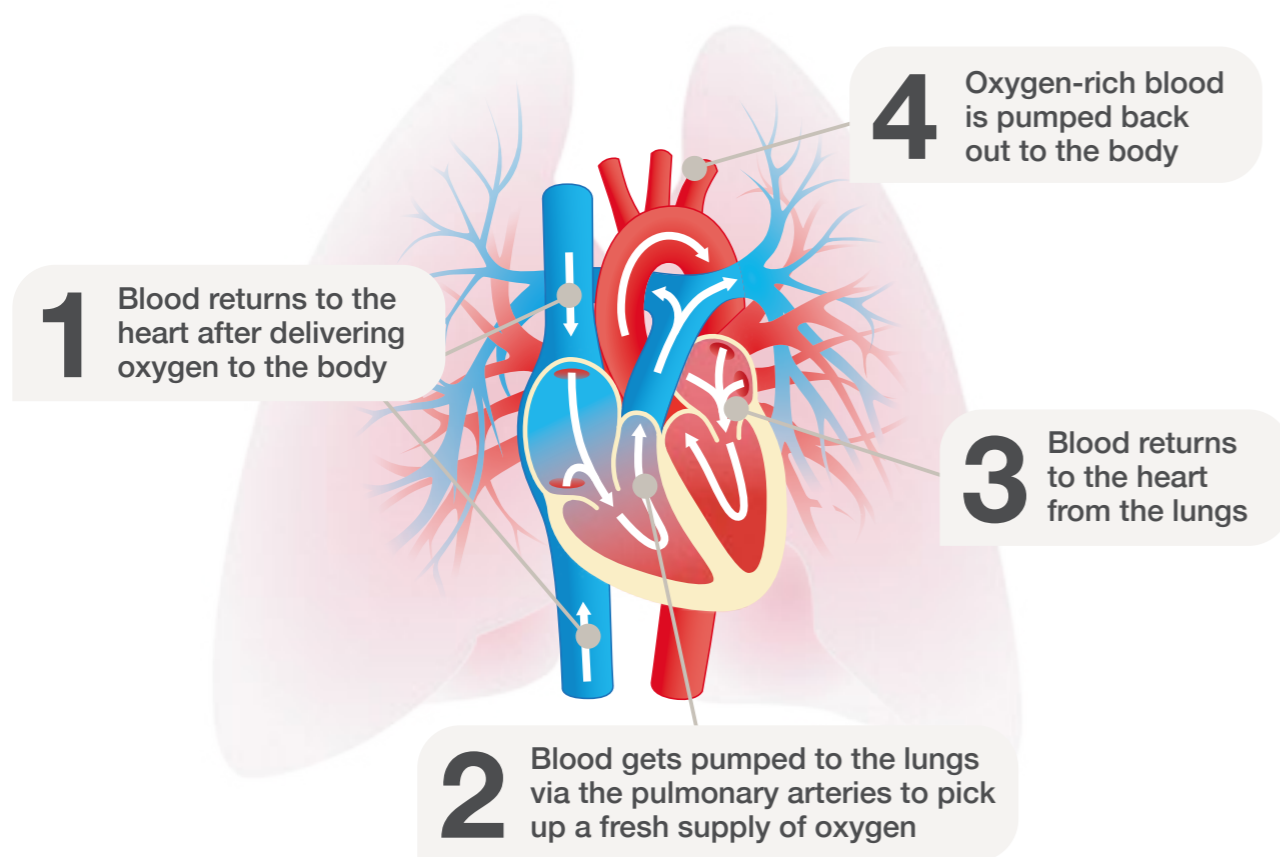
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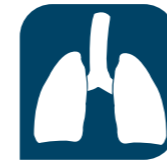
What is pulmonary hypertension (PH)?

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.

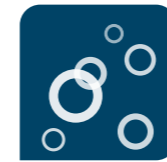
The pulmonary arteries move blood from your heart to your lungs to pick up oxygen. But when they are blocked or narrowed, the blood has to move through a smaller space. In the same way that water pressure increases if you squeeze a garden hose, this blockage causes the blood pressure to rise in your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



Key Facts



When the blood pressure in your lung 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.

What does pulmonary hypertension feel like?

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 PAH?

PAH is a form of pulmonary hypertension which is defined as:

Pulmonary

means relating to the lungs

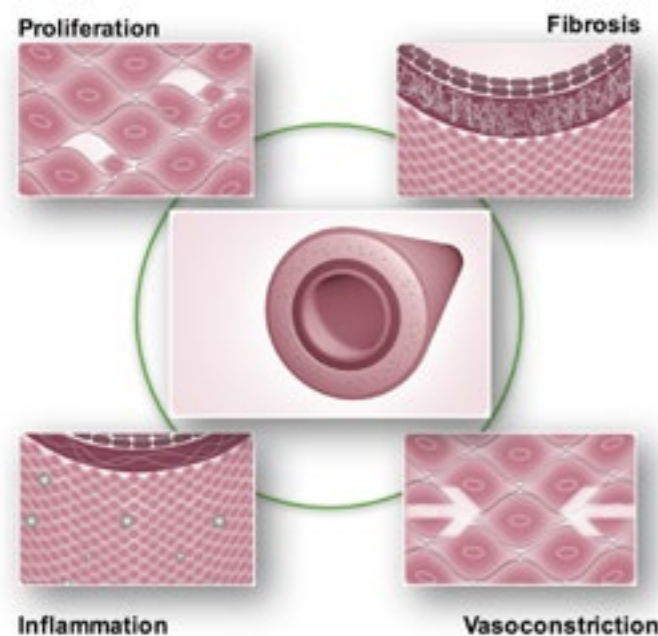
Arterial

means the condition affects the arteries supplying blood to the lungs

Hypertension

is the medical term for high blood pressure

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



Blood vessels in the lung lose some of their ability to relax, and underlying processes cause the walls of the arteries to tighten and thicken, further increasing pressure.

Because of this the right side of the heart has to work harder to pump blood, and eventually becomes enlarged and less able to function properly.

Changes within the artery walls cause the observed effects of PAH

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

Ask your healthcare team if you would like to find out more about what to expect from the treatment options for PAH. There are specific medicines that have been designed to help with your condition, and your health care team will be able to prescribe these for you.

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

Your WHO FC helps your healthcare team figure out how PAH 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 PAH. It is licenced for adult patients with pulmonary arterial hypertension (PAH) with WHO Functional Class (FC) II to III to improve exercise capacity.

Adempas contains the active substance riociguat. Adempas is a type of medicine called a soluble guanylate cyclase (sGC)-stimulator. It works by widening 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 leads to an improvement in your ability to carry out physical activity.



Widens narrowed arteries



Increases ability to do physical activity



Aims to decrease WHO FC

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 center before you start treatment.

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

Very common $\geq 1/10$:

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

Common $\geq 1/100$ 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

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 one of your healthcare 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 healthcare team.

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)

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 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.

Breast Feeding

If you are breast-feeding or planning to breast-feed, consult your doctor or pharmacist before taking this medicine. Stop breast-feeding 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 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

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, 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.

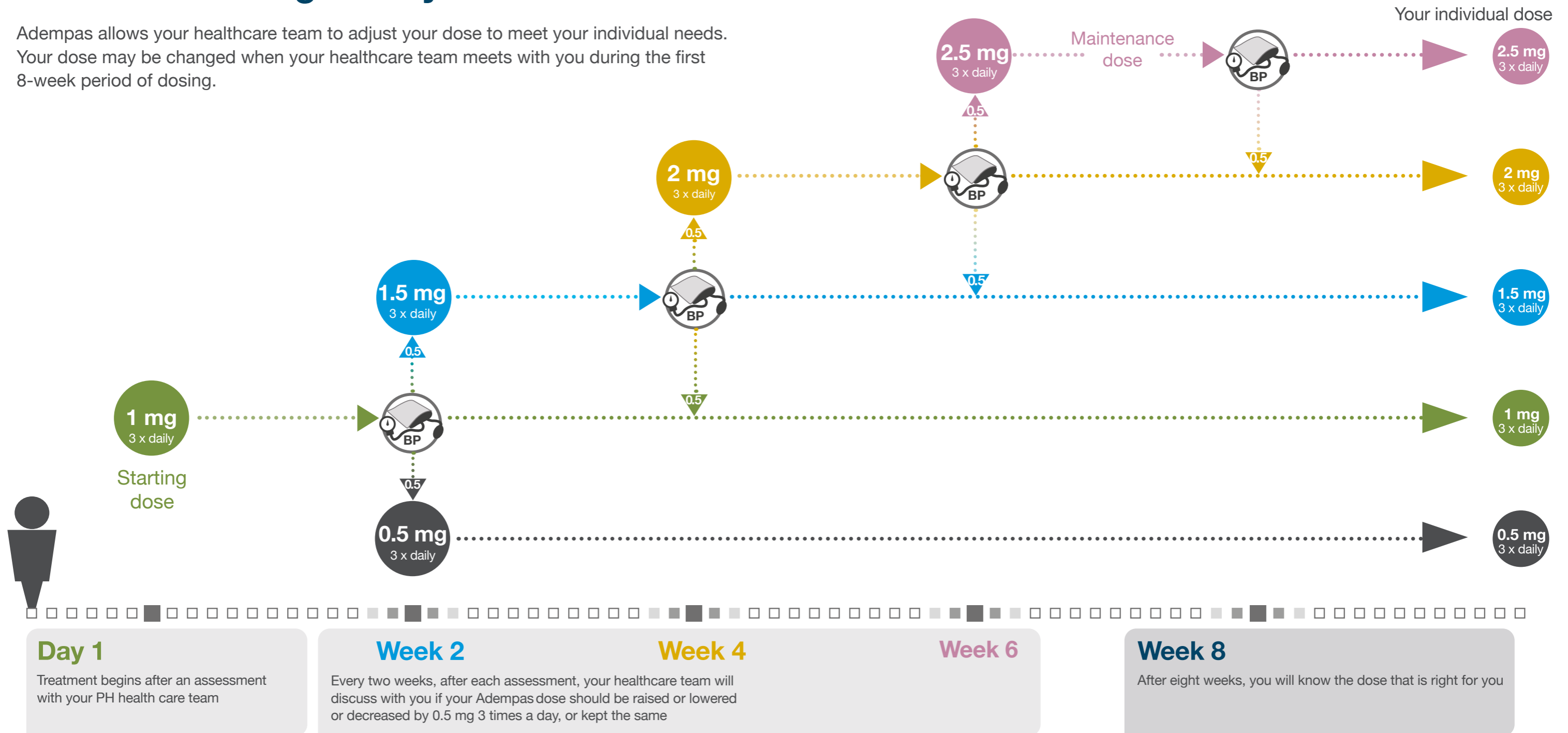
- Adempas is taken orally as a tablet three times a day, with each dose 6-8 hours apart.
- The Adempas starting dose is 1 mg three times a day, taken with or without food.
- Adempas is available in the following doses:



- 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 one day, do not take 2 tablets at the same time – just take the next tablet as planned.
- In case treatment has to be interrupted for 3 days or more, treatment should be restarted with 1 mg three times daily for 2 weeks, and continued with the dose titration regimen as described above.

How your doctor decides which dose is right for you

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



Guide to the symbols



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



Your dose stays the same if your systolic blood pressure is less than 95 mm Hg and you have no 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 healthcare team

Where can I get more information?

The following Web sites and resources offer additional support for people diagnosed with PAH:

PHA UK

www.phauk.org

PHA Europe

www.phaeurope.org



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