



Living with pulmonary arterial hypertension: Supporting you to manage and achieve your goals

early diagnosis • best treatment • better quality of life • finding a cure

PHA EUROPE *for the patients*
European pulmonary hypertension association

The information within this brochure has been endorsed by PHA Europe. PHA Europe aims to improve the standard of care for anyone living with pulmonary hypertension, and to support their friends and family.



About this brochure

- This brochure is for anyone who has pulmonary arterial hypertension (PAH)
- You will find lots of useful information in this brochure about PAH, including what it is, and practical advice to help you adjust to living with PAH
- This brochure is not intended to replace disease guidance by your physicians and healthcare providers. If you do have any questions about PAH or your general health, make sure you ask your physician
- This brochure will also be useful to those around you, such as your family, caregiver or friends, who wish to understand more about PAH so they can support you better



Patricia
Age 33,
newly
diagnosed
with PAH

“When I was told I had PAH, I felt overwhelmed by my concerns about the future. I wasn’t really clear on what would happen, and had many questions.”

All the quotes within this brochure are fictitious accounts based on real-life experiences of people living with PAH.



Contents

- 4 What is PH?
- 5 What is PAH, and how does this differ from other types of PH?



- 6 Experiences of people living with PAH



- 7 What does PAH feel like?
- 8 Keeping a note of how you feel



- 10 How will I and my healthcare team decide which treatment is best for me?



- 12 Personalizing your treatment plan to achieve your goals



- 14 Practical advice for living with PAH



- 16 As a caregiver, how can I support someone living with PAH?



- 18 Notes

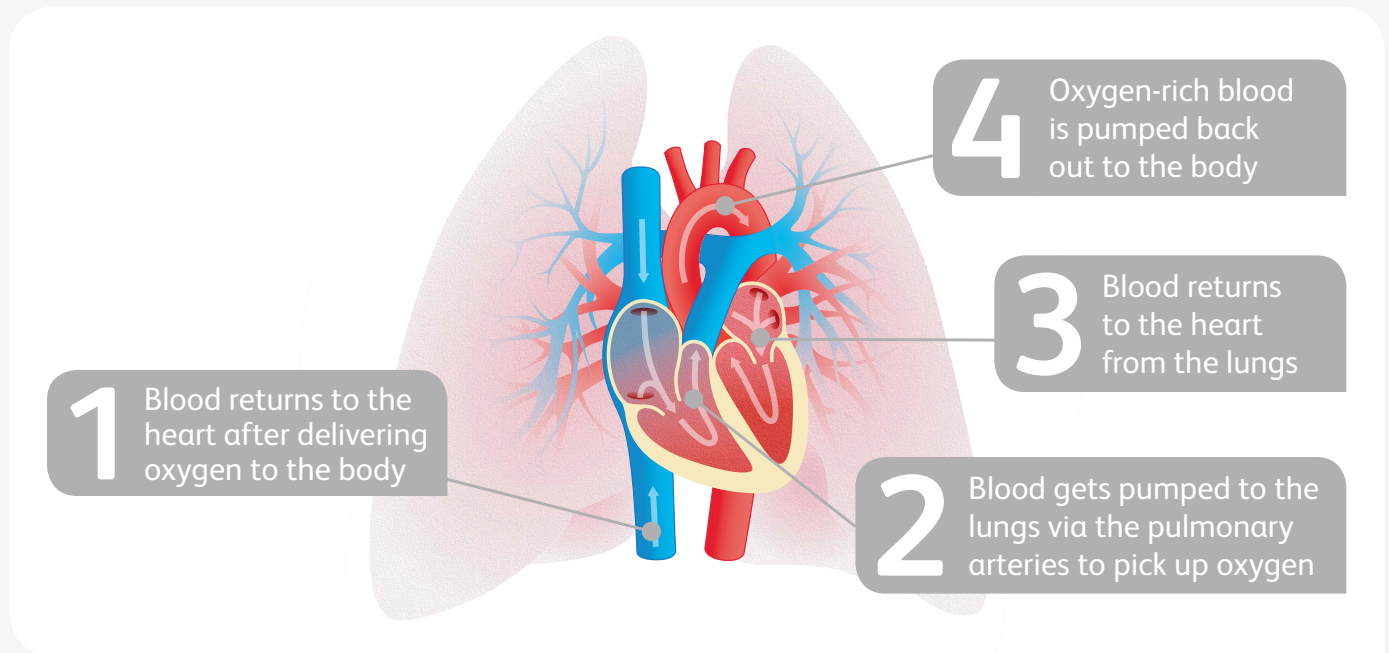


- 20 Further information
- 21 References



What is PH?

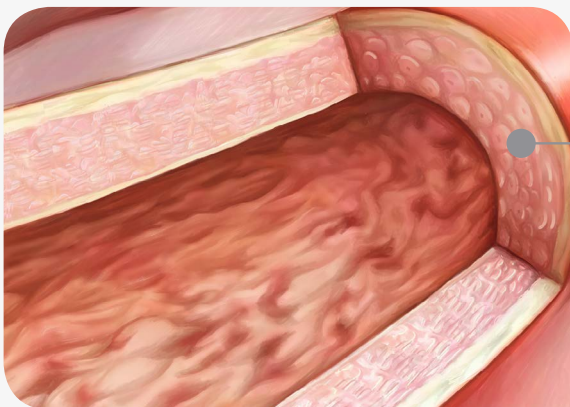
The lungs and heart work together to supply the body with oxygen, as described below:



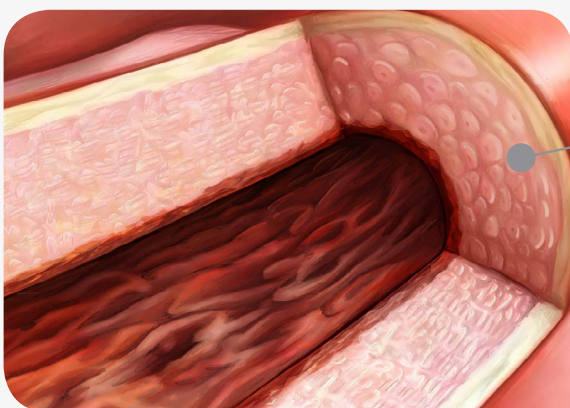
- PH (pulmonary hypertension) is a disease of the lungs and heart, in which the blood pressure in a person's lungs is higher than the normal range¹
- PH may be caused by narrowing or blockage of the pulmonary arteries.¹ This reduces the space in which blood travelling to the lungs can flow, causing the blood pressure in the pulmonary arteries to increase, just like the rise in water pressure that happens when you squeeze a garden hose
- These changes put a strain on the right side of the heart,¹ which means that it needs to work harder than normal to pump blood to the lungs
 - If your lungs receive less blood, less oxygen will be picked up and circulated around your body. This might leave you feeling breathless

What is PAH, and how does this differ from other types of PH?

- There are several forms of PH, which differ in terms of how they develop, and how they are diagnosed and treated^{1,2}
- PAH is one form of PH, caused by changes in the smaller branches of the pulmonary arteries¹
- In people with PAH, the wall of the pulmonary arteries is thicker and stiffer^{1,3}



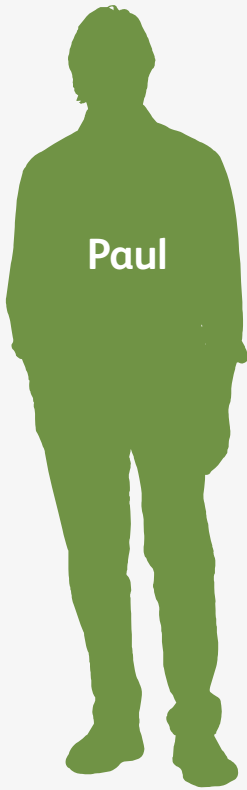
Healthy blood vessel



Blood vessel of a person with PAH

In PAH, the vessel wall in the pulmonary arteries thickens. This makes the opening of the arteries narrower, which in turn restricts blood flow.

Experiences of people living with PAH



Aged 61 and diagnosed with PAH several years previously, Paul had severe symptoms and found it difficult to do many household activities.

“Day-to-day life was a struggle. I often felt out of breath while doing simple activities, like cooking dinner and tidying the house.”

Aged 33 and newly diagnosed with PAH, Patricia went to her doctor because of difficulty keeping up with her children.

“I am worried that I can't keep up with my children without feeling breathless and having to rest often to catch my breath.”



What does PAH feel like?

As someone living with PAH, you may be experiencing symptoms such as shortness of breath during exercise, feeling tired, weak or dizzy, fainting, chest pain, and swelling of ankles, arms or stomach.⁴



Shortness of
breath with
exercise



Feeling tired,
weak or
dizzy



Swelling in the
ankles, arms or
stomach area



Fainting



Chest pain

- These symptoms may become more noticeable over time, making it more difficult for you to do normal daily activities or chores
- Talk to your doctor if you notice that your symptoms are getting worse



Patricia

“I’m finding it more difficult to keep up with my children. I often have to stop to catch my breath. Our walks aren’t as much fun as they used to be.”



Keeping a note of how you feel

- It is important that you see your doctor on a regular basis to review and discuss your health
- Your doctor wants to know how you are feeling now, as well as how you have been feeling since your last visit. So, before you see your doctor, think about how you have been since your last visit
- Your doctor may talk about your 'functional class'. This is one way of describing how severe your symptoms are and whether they affect your daily life (for example, your ability to do exercise or regular chores)
 - Classifying the severity of your PAH will help your healthcare team understand better how it is affecting you

Functional class⁴

I	Symptom-free when physically active or resting	<ul style="list-style-type: none">• Make sure you tell your doctor if you think your symptoms have improved!• If you feel your symptoms are worse, or are not improving, it is important to tell your doctor, so they know that your treatment perhaps isn't working as well as it should and you can work together to find the best way forward
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	
III	Resting may be symptom-free, but normal chores around the house are greatly limited because of shortness of breath or feeling tired	
IV	Symptoms at rest and severe symptoms with an activity	



Keeping a note of how you feel

Here is a list of some of the things you might wish to talk to your doctor about at your next visit. Some people find it easier to write things down, so there is space for you here.

Since your last visit:



How have your general energy levels been?

.....
.....



Have you felt normal daily activities (like washing, dressing, or moving around the house) are noticeably more (or less) challenging?

.....
.....



Have you experienced any new symptoms?

.....
.....



Have you experienced dizziness, weakness or feeling faint?

.....
.....



How many minutes can you walk for before you need to take a break?

.....
.....



How many flights of stairs can you walk up before you feel breathless?

.....
.....



To what extent do you feel that your PAH is affecting your overall quality of life?

.....
.....



How will I and my healthcare team decide which treatment is best for me?

PAH may be treated using drugs that improve blood flow through your lungs.

- These drugs generally fall into four types, and may be taken as a tablet, infused or inhaled²

ERAs

Endothelin receptor antagonists

sGC stimulators

Soluble guanylate cyclase stimulators

PDE5 inhibitors

Phosphodiesterase type 5 inhibitors

PCAs

Prostacyclin analogs

Although there is no cure for PAH, there are several specialist treatments available that can help to relieve symptoms and make day-to-day life better.

What is the best treatment for you?

- Your treatment should **help you to achieve your personal treatment goals**, such as walking with your children, being able to perform routine chores without feeling breathless, or continuing to enjoy your leisure activities such as dancing
- Treatment should help to **improve your day-to-day symptoms** and ultimately **reduce the progression of disease and improve survival**
- **Your doctor will work with you** to find out which treatment(s) will best improve your symptoms, and help you to achieve a low-risk status and your own personal treatment goals
 - From a doctor's perspective, the overall treatment goal is for you to have non-progressive disease (described as achieving, and maintaining, a 'low-risk status')²
- Both doctors and patients have a role in making decisions about treatment, and some patients may wish to be more involved than others. Don't hesitate to discuss treatment options proactively with your doctor and ask about any side effects that might result from taking a particular drug(s)



How will I and my healthcare team decide which treatment is best for me?

- Your doctor might recommend that you take several drugs together (combination therapy) in the first instance, or, depending on your needs and/or availability, your doctor might prescribe a single drug (monotherapy)
- Because not all drugs can be prescribed together, your doctor will advise you on which drugs can be taken together
- You should see your doctor on a regular basis to review and discuss your symptoms, and how you feel you are getting along with your treatment (including whether you have felt any side effects, for example, dizziness)
- If you feel that your symptoms have changed, or have not improved as much as you would have liked, please discuss this with your doctor



Patricia's doctor:

“Patricia might start on a single drug or a combination of two drugs to help improve her symptoms and achieve a low-risk status.”

Patricia's doctor:

“By low-risk, we mean meeting several criteria for stable disease, including achieving a functional class of I or II to decrease the risk of severe problems.”



Personalizing your treatment plan to achieve your goals

'Making do' with how you are feeling is not good enough; you should hope to be symptom-free when resting or while doing physical activities, or you should feel that your symptoms are improving.

- If you feel any symptoms during daily activities or if your symptoms have not improved as much as you would have liked, ask your doctor whether a change in treatment could be helpful for you
- You might also need to change your treatment if your symptoms have become worse or if you experience any side effects on your current treatment
- Your doctor may recommend that you take additional treatments to your existing one(s). Alternatively, they may suggest you stop one treatment and start a new one (i.e. switch treatments)

Your PH specialist will assess whether it is best for you to:

Stay on the same dose of your current treatment

Adjust the dose of your current treatment

Switch to a different PAH treatment

Add another PAH treatment



Personalizing your treatment plan to achieve your goals



Paul's doctor:

“Regular follow-up with Paul is an important part of his care. Because of these visits, we found that Paul's symptoms were not improving so I suggested adding on another PAH medication.”

Paul:

“I felt that my symptoms were not getting better on the treatment that I had been taking for several years. After talking to my doctor about my concerns, I was given an additional PAH medication to try and I have started feeling better. Cooking and tidying do not leave me as breathless or uncomfortable.”

Paul's doctor:

“Now that Paul has started taking another PAH medication, I have seen improvements in his symptoms. If Paul's symptoms do get worse again, I will consider other options such as switching to a different PAH treatment or starting PCA therapy.”



Practical advice for living with PAH



Emotional well-being

Your emotional, spiritual and psychological well-being is as important as your general health. Your doctor can recommend support groups and other experts who can support your emotional well-being.



Social support

In addition to support from family and friends, your doctor can provide you with details of local support groups and patient organizations where you can talk to other people with PAH. It is important to talk about how you are feeling, to reduce stress and to make sure that you do not feel alone.



Work

You might need to think about changes to your day-to-day activities at work, especially if they are leaving you breathless. Once your symptoms start to improve, you could increase your activity level at work.



Education

If you or someone you are caring for is at school, college or university, you may wish to talk to someone at the institution to let them know how PAH might affect studies or extracurricular activities. Changes to the timetable, extensions on assignments or examinations, and fewer or lighter activities outside of study might be helpful.



Reporting side effects

Before starting treatment, it is important to speak to your doctor about any side effects that you may experience. If you think you may have experienced a side effect after using a medicine, contact your doctor who will be able to tell you if you need any alternative care.

In the European Union you can also report side effects or adverse reactions directly through various methods (e.g. online patient reporting forms) without going through your physician.



Practical advice for living with PAH



Travel

Take care not to over-exert yourself while travelling or on holiday. Find out about any PAH clinics where you are going, and take information about your PAH and your medication with you in case you need medical support. High altitudes (for example, >1500 meters) should also be avoided, unless you can take supplemental oxygen. Some people might also need supplemental oxygen to maintain their oxygen levels during long flights.²



Diet and exercise

It is important to eat a balanced diet, and to maintain a healthy weight. You can stay active whilst living with PAH, but it is important to seek advice on exercise from your healthcare team. Carefully supervised physical activity can form part of an effective treatment strategy for PAH. For some patients with PAH, even the mildest forms of activity might be too exhausting. For others, moderate exercise might be beneficial.



Sexual health and family planning

You may experience a decline in sexual intimacy, perhaps because of concerns relating to over-exerting yourself. Talk openly with your partner about your feelings. Pregnancy poses a substantial risk for women with PAH, because it will put extra demands on the heart and lungs. Therefore, women with PAH are recommended to use reliable contraceptive methods to avoid becoming pregnant.²

For further information and support, please refer to the websites listed on page 20 at the end of this brochure.



As a caregiver, how can I support someone living with PAH?

Being emotionally supportive is probably the most important thing you can do as a caregiver. But there are also some practical things that you may be able to do to help the person you care for.



Keep an eye on symptoms

Keep note of any changes you have noticed since the last check-up. It is very important that the healthcare team knows what the person you care for is going through daily and how they are progressing with their treatment goals.



Help prepare for and attend doctors' appointments

To get the most out of doctors' appointments within the often-limited time available, it is important to prepare beforehand. Have a chat with the person you care for and ask them to:

- **think carefully about what they want to get out of the appointment (see page 9)**
- **write down any concerns or questions they (or you) might have**



Make sure the person you care for feels valued and included

PAH is likely to have an impact on a person's social life. Encouraging other friends and family members to visit can stop people living with PAH feeling isolated, as can including them in activities of normal day-to-day life where possible (with consideration of the limitations of their disease).



As a caregiver, how can I support someone living with PAH?



Patricia's mother:
“Patricia and I sit down together the day before each appointment, and write down the questions we want to ask. This way we won't forget anything important, and can make the most of our time with the doctor.”



Paul's caregiver:
“Paul sometimes isn't aware of how his symptoms are gradually changing. It is sometimes easier for me to spot things, so I make a record of the changes I notice.”

As a caregiver, it is vital to take steps to preserve your own health and well-being, because when your needs are taken care of, the person you care for will benefit too. For further information on caring for someone with PAH, please refer to the caregiver brochure, which is free to download at the following link:

http://www.phaeurope.org/wp-content/uploads/14290_Bayer_Caregiver_brochure.pdf



Further information

The following websites offer additional information and support for people living with PAH and their carers:

Pulmonary Hypertension Association Europe

www.phaeurope.org

Pulmonary Hypertension Association

www.phassociation.org

Bayer is not responsible for the content of external websites.



References

The following publications support the guidance and advice within this brochure.

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2. Galiè N *et al.* 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). *Eur Heart J* 2016;37:67–119.
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G.MKT.GM.04.2018.0702
Date of preparation: May 2018