

# *BREATHE IN, SPEAK OUT*

## CONVERSATION KIT

**PHuman™**

# LIVING WITH PAH: HOW TO MAKE THIS TOOLKIT WORK FOR YOU

## Quickstart guide

Living with pulmonary arterial hypertension (PAH) can affect several aspects of your life. Learning more about PAH, and listening to your own emotions, needs, and goals can give you the power to be more active in your care, and to help find the path that works best for you.

Each set of cards in this toolkit adds a new dimension to your understanding of PAH and how to manage PAH in your day-to-day life.

**Choose what works for you, based on where you are in your PAH journey.**

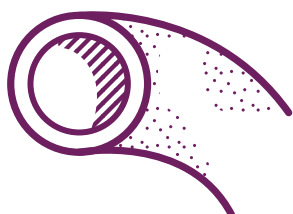
	By yourself	With family and friends	With your healthcare team
Knowledge cards	Understand and learn more about PAH	Talk about the facts to increase awareness	Feel more confident in having informed discussions with your healthcare team
Emotion cards	Reflect on how you feel living with PAH	Share how you're feeling to help loved ones understand	Explain to your healthcare team how you're feeling
Confidence cards	Restore and nurture your mindset on living with PAH	Enlist support from loved ones to help you maintain focus	Carry the cards to boost your confidence
Personal PAH motivation worksheet	Tailor your plan to your own priorities of living with PAH	Build your plan to keep you motivated	Enlist your healthcare team to achieve your goals

# KNOWLEDGE CARDS

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# HOW PAH AFFECTS YOUR BODY

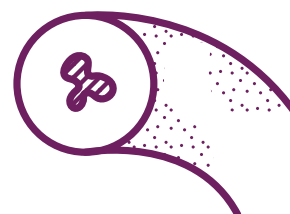
Pulmonary arterial hypertension (PAH) is a rare condition that narrows the walls of the blood vessels that carry blood to your lungs. With PAH, the blood has less room to flow, so your heart has to work harder than normal to pump blood to your lungs.<sup>1,2</sup>



Healthy  
artery

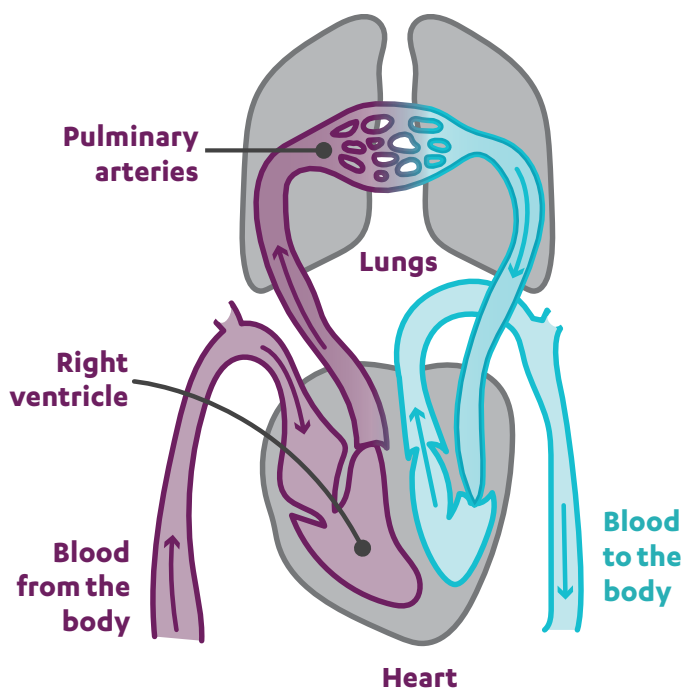


Narrowed  
artery



Severely  
narrowed artery

Over time, the right side of your heart will get larger because it's working too hard. Then, when your heart isn't working properly, you may feel breathless and tired. That's because less blood is flowing to your lungs, and less oxygen is picked up by the blood to take around your body.<sup>1,2</sup>



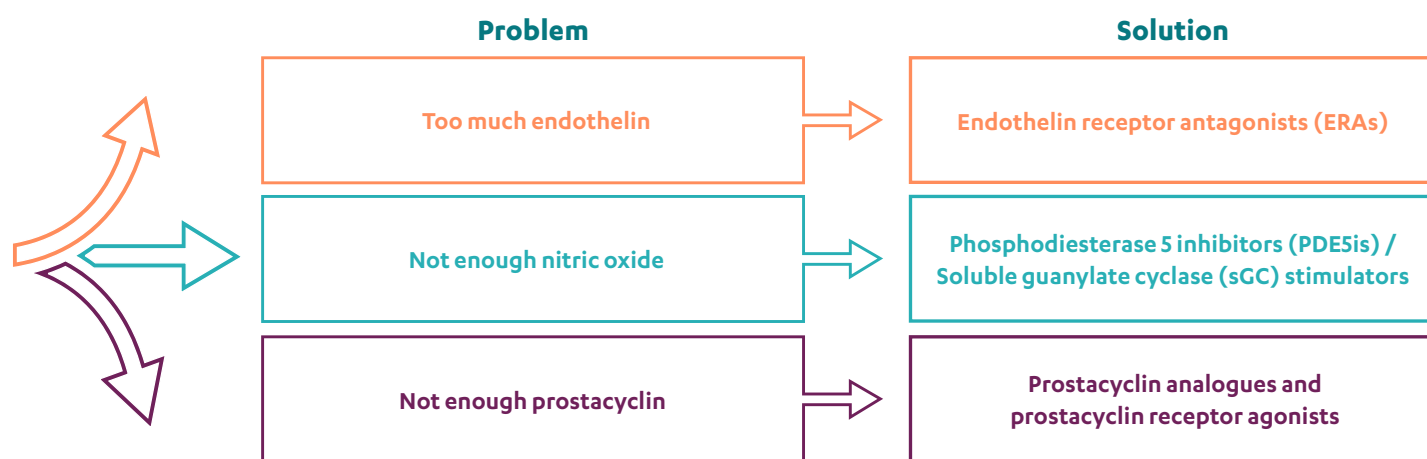
PAH is a serious condition that gets worse over time or 'progresses'.<sup>1</sup> While there's no cure for PAH yet, there are medicines that can help slow it down<sup>2</sup> (your healthcare professional calls this, 'delaying progression').

## References

1. NHS. Pulmonary hypertension. Available at: <https://www.nhs.uk/conditions/pulmonary-hypertension/> Last accessed October 2021.
2. American Lung Association. Learn about pulmonary arterial hypertension. Available at: <https://www.lung.org/lung-health-diseases/lung-disease-lookup/pulmonary-arterial-hypertension/learn-about-pulmonary-arterial-hypertension> Last accessed October 2021.

# TREATING PAH

There are three main cell-signalling pathways in your blood vessels that help ensure the right balance of blood flow through the lungs.<sup>1</sup> In PAH, there is an imbalance in these pathways. There are several different treatments available, including oral, inhaled and intravenous medications,<sup>2</sup> which act on these different pathways to help restore balance and ensure blood can flow freely through the lungs, carrying oxygen to the rest of the body.



Slowing down the speed at which PAH gets worse (also called ‘delaying disease progression’) is an important treatment goal in PAH.<sup>3</sup>

Your healthcare professional may prescribe one or more medicines to treat as many pathways as early as possible, to improve your chances of feeling better for longer. Other supportive treatments that may be recommended for PAH include blood thinners to prevent clots, diuretics (also called water tablets) to reduce swelling, and supplementary oxygen to ensure the body has enough.<sup>4</sup>

Speak to your doctor about your medicine choices. Your doctor knows how to get the best results and make changes in your PAH treatment along the way. Changes don’t always mean that something is wrong. While there is no cure for PAH yet,<sup>3</sup> treatments can help people manage and slow progression of the condition.<sup>1</sup>

## References

1. Sitbon O, Gaine S. Beyond a single pathway: combination therapy in pulmonary arterial hypertension. *Eur Respir Rev* 2016;25:408-417.
2. Pulmonary Hypertension Association UK. Treatment for pulmonary hypertension. Available at: <https://www.phauk.org/treatment-for-pulmonary-hypertension/> Last accessed October 2021.
3. Vachiéry JL, Gaine S. Challenges in the diagnosis and treatment of pulmonary arterial hypertension. *Eur Respir Rev* 2012; 21:313-20.
4. British Lung Foundation. Treatment for pulmonary hypertension. Available at: <https://www.blf.org.uk/support-for-you/pulmonary-hypertension/treatment>.



# MANAGING RISK

PAH affects you in ways that you can't always see or feel. Your doctor may call this 'silent progression'.

You'll probably hear your healthcare team talk about 'risk assessment'. That means they'll run a series of tests at every visit, which may include:<sup>1,2</sup>

- **Blood tests:** to test for potential diseases that are associated with PAH, as well as standard tests like a complete blood count (CBC) and chemistry panels.
- **ECG (electrocardiogram):** to record the electrical activity in the heart and detect abnormal heart rhythms.
- **Echocardiogram:** an ultrasound scan that looks at the chambers and valves of your heart and surrounding areas to see what they look like and how they function. This can detect signs of PH and how severe it is.
- **6-minute walk test (6MWT):** to measure how far you can walk in 6 minutes.
- **Right heart catheterisation:** a procedure that shows how well your heart is pumping, and measures the blood pressure in your heart and the blood vessels leading to your lungs.

There are a number of different treatments for PAH – you can talk to your doctor about treatment choices if you have any questions.



## References

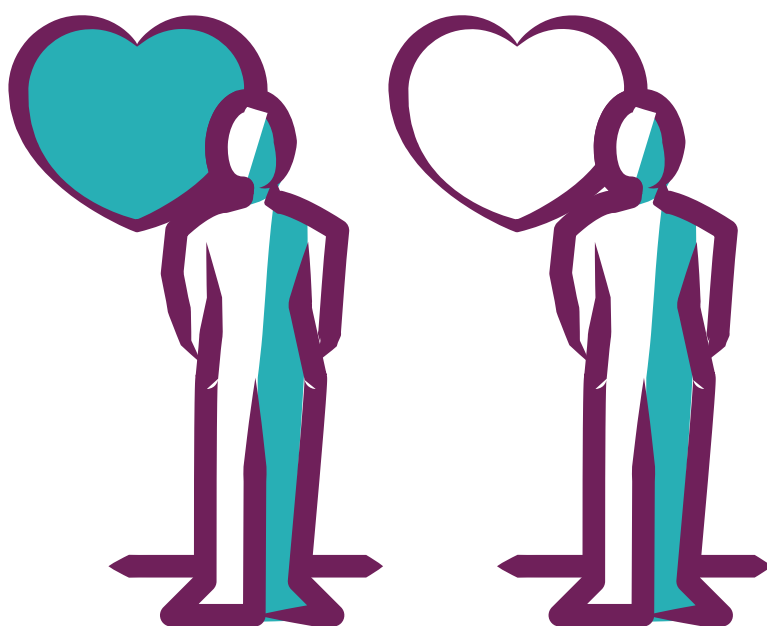
1. American Lung Association. Pulmonary arterial hypertension symptoms and diagnosis. Available at: <https://www.lung.org/lung-health-diseases/lung-disease-lookup/pulmonary-arterial-hypertension/symptoms-diagnosis> Last accessed October 2021.
2. British Lung Foundation. How is pulmonary hypertension diagnosed? Available at: <https://www.blf.org.uk/support-for-you/pulmonary-hypertension/diagnosis> Last accessed October 2021.

# GATHERING PEOPLE TO SUPPORT YOU

You are not alone. Your healthcare team, other professionals, family and friends can help you handle the challenges of living with PAH.

Build a network of people to give you the different kinds of support you may need. Here are some tips to keep in mind when you're thinking about who should be in your network:

- **It's okay to ask for help.** Sometimes the hardest part is letting yourself ask. Remember, people are usually glad to feel needed. You just have to let them know how to support you.
- **Help comes in many forms.** There are different kinds of support you can ask for. Some people may be better at giving one thing you need, while others bring something else to the table.
- **Help them help you.** Remember that only you fully understand what you're going through. If you share what's happening with the people in your life, you may help them do a better job supporting you.
- **Plan for changes to come.** PAH can affect your ability to do everyday tasks, which can be frustrating. Let your supporters know what changes you may need for a plan that puts everyone more at ease.
- **Take time out.** Make sure that you and the people you rely on take time to recharge.
- **Talk about how you're feeling.** Talking about how you're feeling is an important way of making sure you get the support you need as early as possible. Don't be shy! Talk to your support group, your healthcare team, or a therapist. Let your voice be heard.

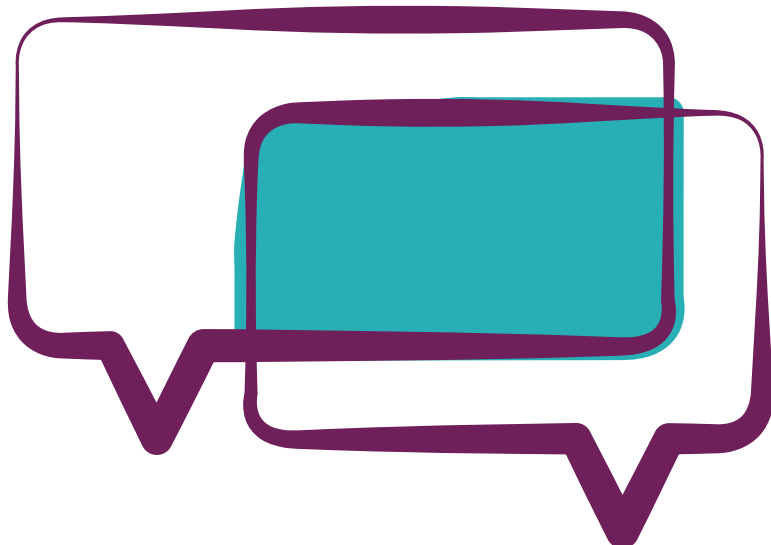


# PARTNERING WITH YOUR HEALTHCARE TEAM

When you have PAH, your healthcare team is there to find the best treatments for you. Make sure your needs are understood by being open with the team and sharing your thoughts and feelings. You can help them make sense of what they're seeing, and even see things more clearly by what you share. Even if they seem busy, you have a right to be heard.

When talking with your healthcare team, keep the following tips in mind:

- **Be open.** Your doctor will monitor your test results, but you're the only one who really knows how you're doing in your daily life. Talk honestly and include everything: thoughts, feelings, goals, and concerns.
- **Keep up to date on treatments.** It's important to learn all you can about any current and newly prescribed medications or treatments. Each medication may have some benefits, but also have side effects. Knowing what to expect can help you prepare for what's ahead.
- **Speak up.** If something doesn't feel right, or if you aren't feeling satisfied with your care, let your doctor know. When they know your honest opinions your healthcare team can work together to adjust your plan if necessary and appropriate.
- **Come prepared.** Reflect on what's important to you before each appointment. Bring any lists, logs, and questions with you. You can also use the cards in your *Breathe In, Speak Out* toolkit to prepare.
- **Ask until you get an answer.** If you don't feel like a question was answered or your concern was heard, ask again.





# EMOTION CARDS

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# Joyful

What is bringing me hope  
and energy?



# Excited

What is most exciting in  
my life right now?



# Thankful

What am I most  
thankful for?



# Optimistic

What am I looking  
forward to?





# Encouraged

What [recent accomplishments that]  
makes me feel proud?



# Eager

What do I want  
to accomplish?



# Relieved

What do I look forward  
to returning to?



# Lonely

What do I want to  
say to others?



# Anxious

What is one small step I can take to feel better?





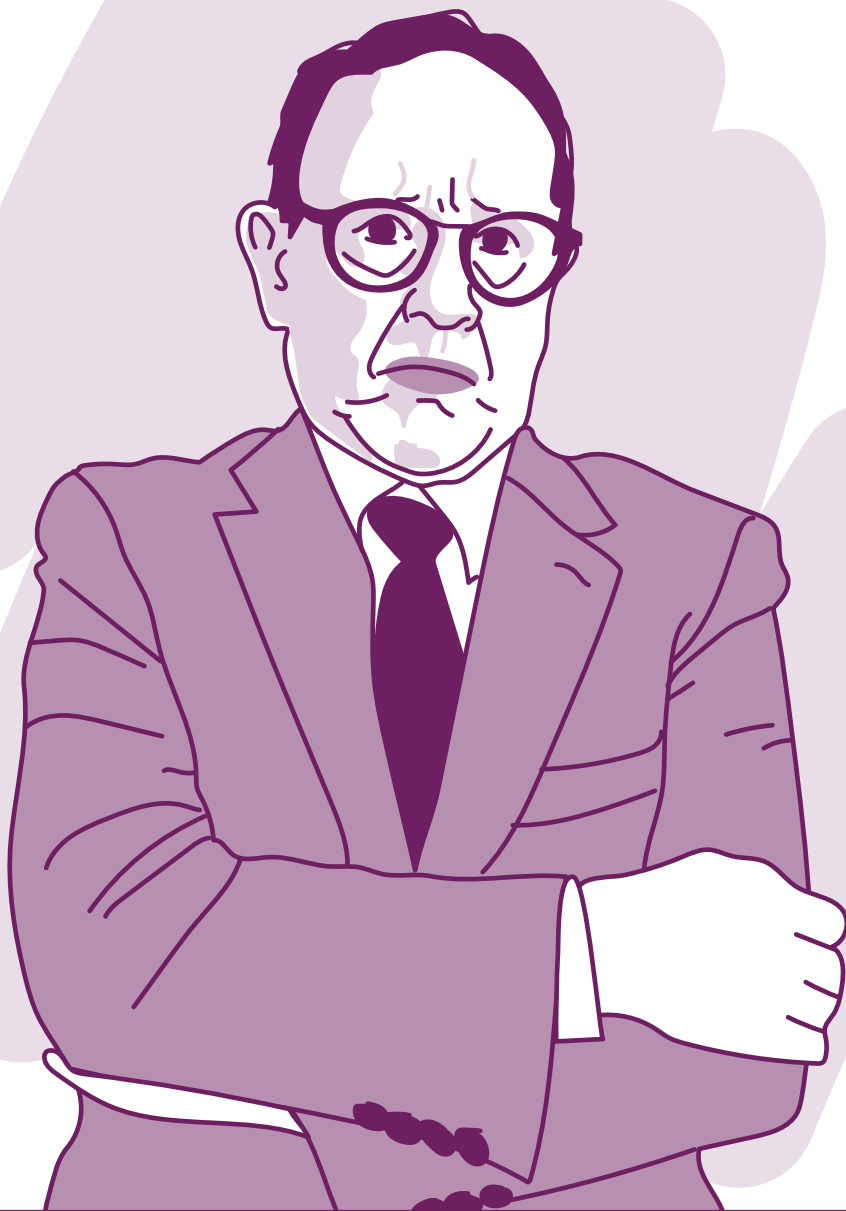
# Frustrated

What would bring  
me some relief?



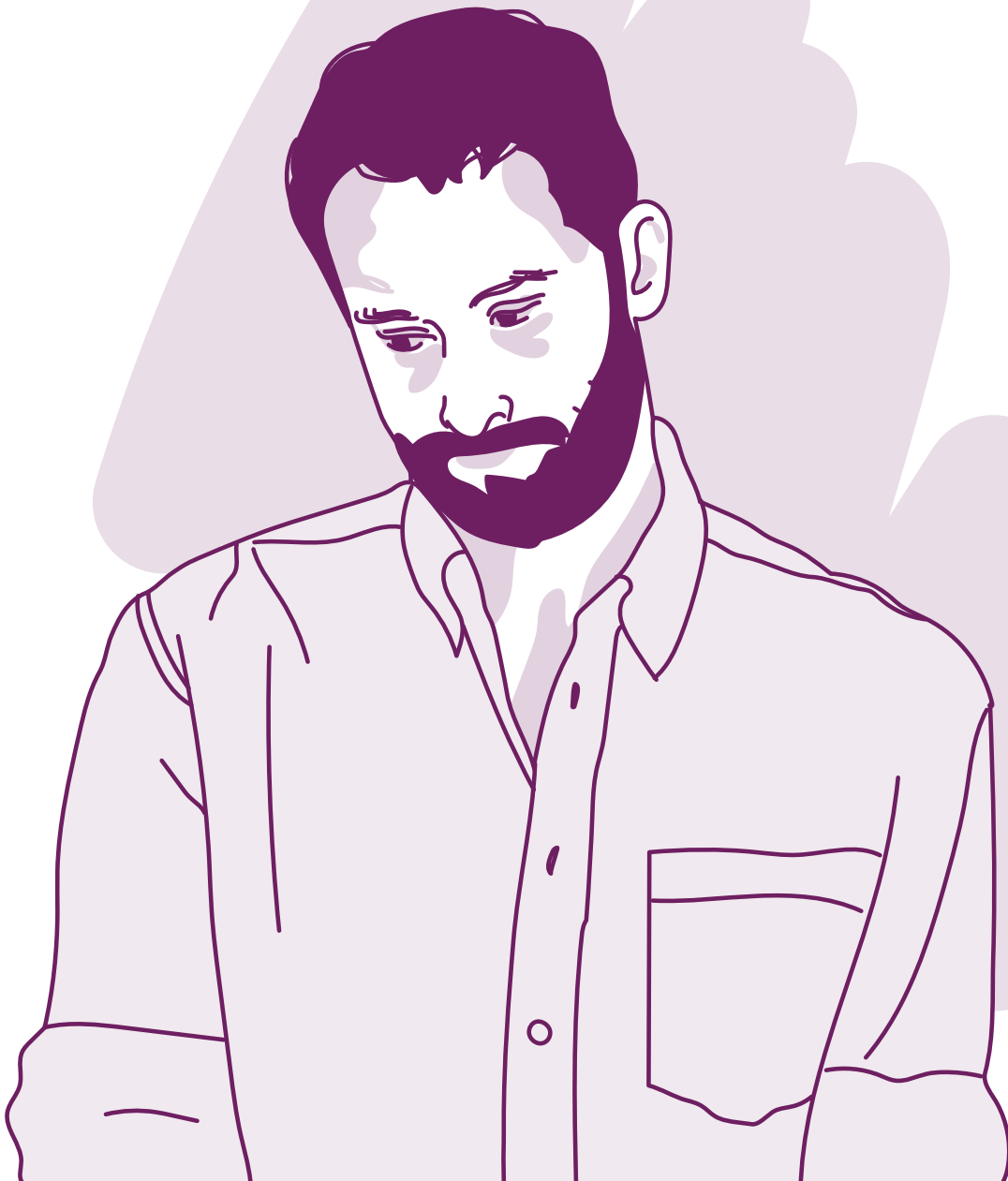
# Worried

Is fear preventing me from doing  
or saying something?



# Sad

What do I need to make  
me feel supported?



# Overwhelmed

What things are  
in my control?



# Scared

What would I say or  
do if I wasn't afraid?





# Stressed

How am I coping  
with the stress?



# CONFIDENCE CARDS

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*'It's about mentality not physicality.  
Don't let PAH change the course that  
you want to take.'*

Person living with PAH



*'Start accepting that the disease is not you, just a curve in the road. There are good moments and bad, but one day at a time.'*

Person living with PAH



*‘Keep your mind set on your goals,  
as a glimmer of positivity can  
improve your day, your month,  
even your year.’*

Person living with PAH





*'I don't let the disease define who I am. I'm going to keep on being me.'*

Person living with PAH



*‘Having hobbies is important for your mental health whether living with PAH or not. Let go of social expectations and pursue something that makes you happy.’*

Person living with PAH



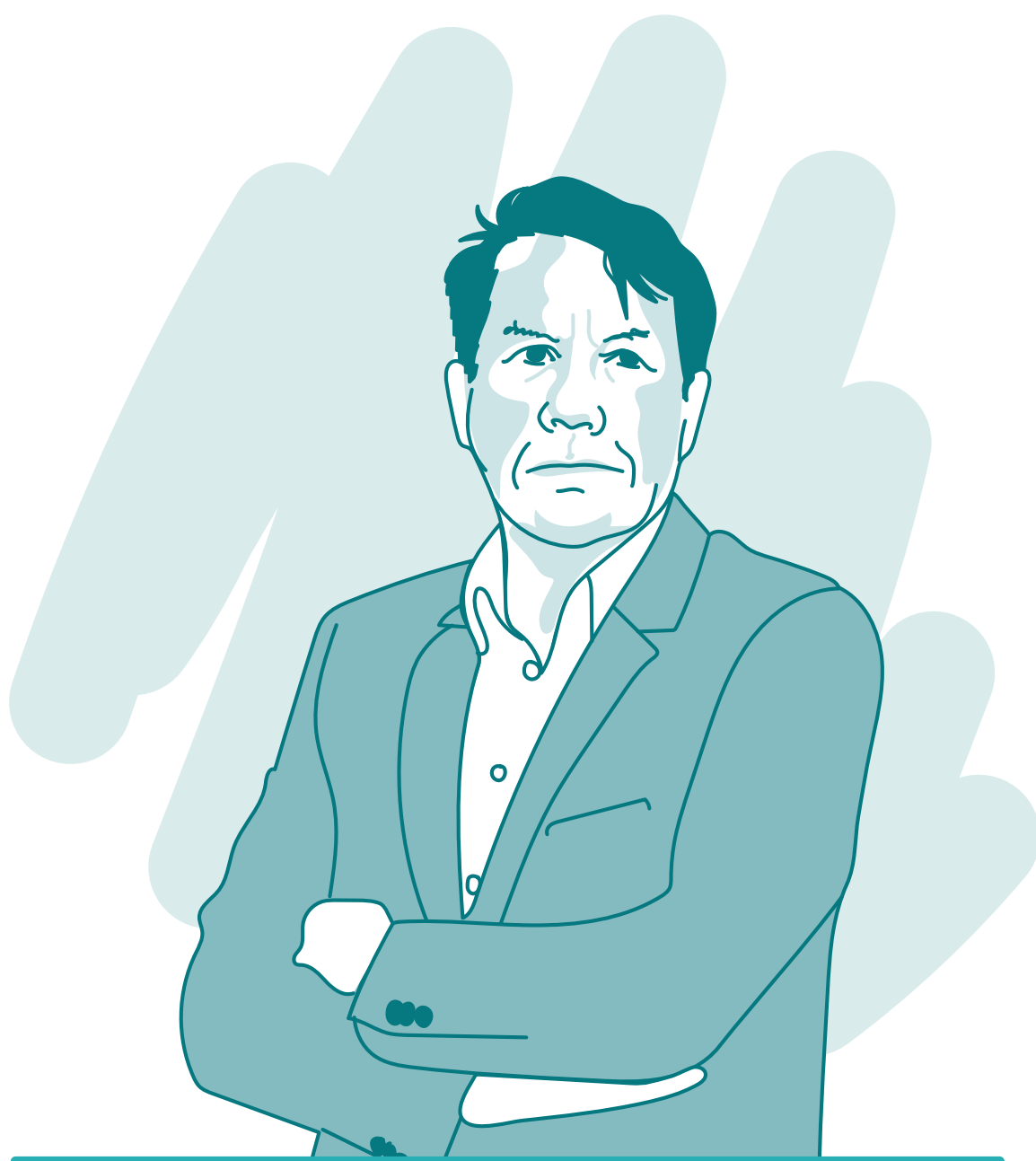
*‘Talk to your peers, learn from them and prepare for what to expect. Social media offers a vibrant network of people, information and resources that can be harnessed in the best ways possible to help you find support.’*

Person living with PAH



*‘Talk to your doctor about your personal goals. Having an open dialogue about your goals will help your doctor plan the treatment strategy that’s best for you.’*

Person living with PAH



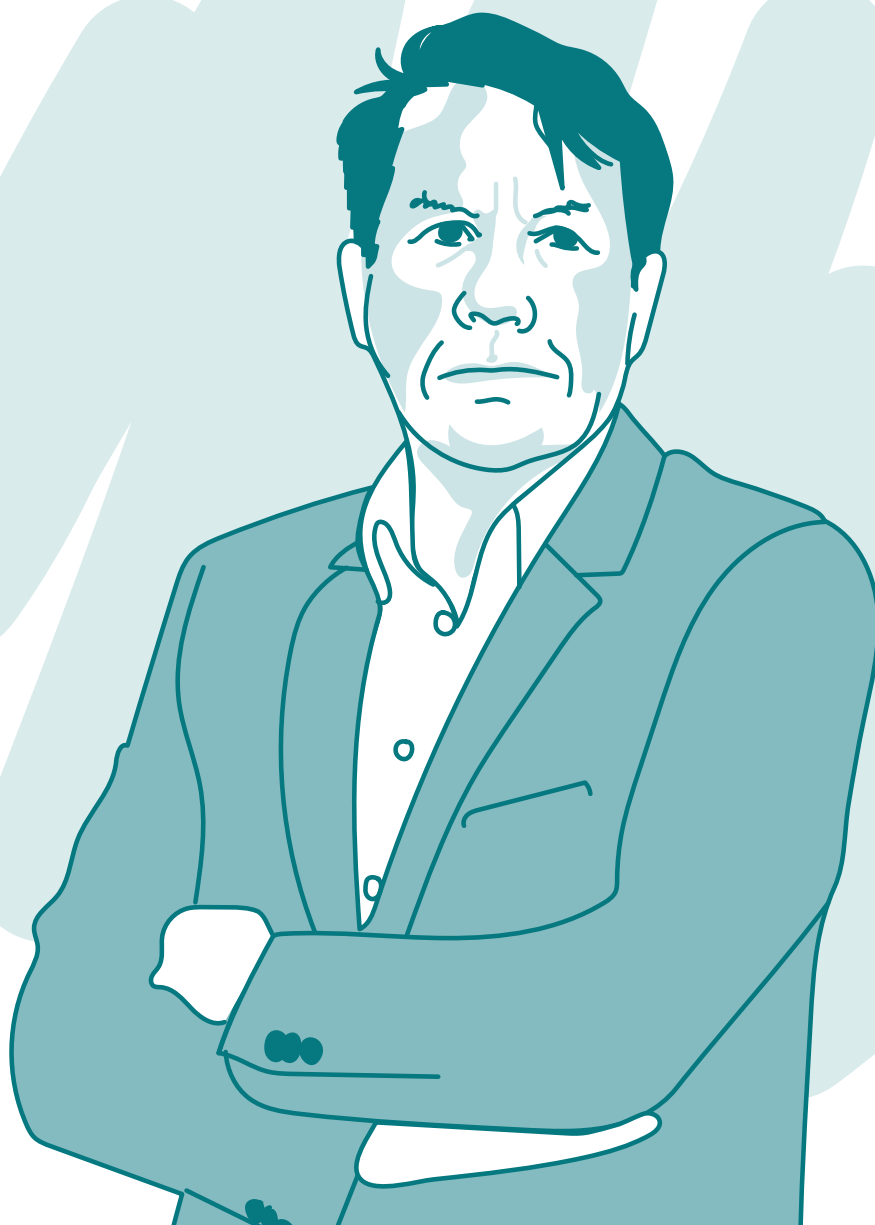
*'I've found it is important to be my own health advocate. This means not taking a passive backseat in my health journey. I try to be as actively involved in my health journey as possible and vocalise any concerns I have. After all, I am the only one inside my body.'*

Person living with PAH



*'Be honest with your doctor. Don't say it's fine when it isn't.'*

Person living with PAH



*‘With PAH, it’s important  
to be proactive.’*

Person living with PAH



# PERSONAL PAH MOTIVATION WORKSHEET

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## REFLECTION

# You are the expert of you.

Your PAH doctor and healthcare team rely on you to speak up about what you want to get out of treatment and what matters to you. Use this worksheet to organise your thoughts and create a plan that works for you.

# WHAT MATTERS MOST TO ME

**What motivates me?** Think about your ‘why’ and what motivates you to fully engage in the management of your PAH.

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**What do I enjoy?** Specific things that you do alone or with friends and family during a typical week. Write down what you like to do so your healthcare team can work with you to find solutions that fit your life.

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On my own...

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With friends and family...

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## WHAT KEEPS ME GOING

**What am I looking forward to?** Think about the things you're looking forward to so you can share them with your healthcare team. Together, you can keep these things in mind as you make your care plan.

[illegible]

**Things I am looking forward to in the next few months:**

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**My goals for my health and managing PAH:**

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**I prefer a treatment plan that will allow me to:**

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**My long-term goals and milestones:**

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# WHO SUPPORTS MY JOURNEY

Think about ways that others can help you stay motivated and achieve your goals. Use this list to help you build a support network of people who can be there for you in different ways.

## People who support me with:

### Treatment decisions

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### My emotional needs

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### My social needs and hobbies

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# My hard days

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# Things around the house

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# An emergency

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# Celebrating milestones

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