Breathe In, Speak Out

Your Toolkit for Pulmonary Arterial Hypertension (PAH)

breathe in
SPEAK OUT

Living with PAH: How to make this toolkit work for you.

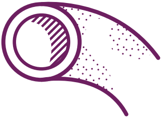
Living with PAH affects all of you. Learning more about PAH, and listening to your own emotions, needs, and goals can give you the power to be more active in your care, and to help find the path that works best for you. Each set of cards in this toolkit adds a new dimension to your understanding of PAH 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	Review & learn	Talk about the facts	Bring the cards to each appointment
EMOTION CARDS	Reflect	Share	Tell your team how you’re feeling
CONFIDENCE CARDS	Recharge	Enlist support	Carry the cards to boost your confidence
REFLECTION CARDS	Make it your own	Build your plan	Enlist your team to achieve your goals

How PAH Affects Your Body

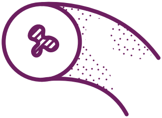
KNOWLEDGE

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.



Healthy artery

Narrowed artery

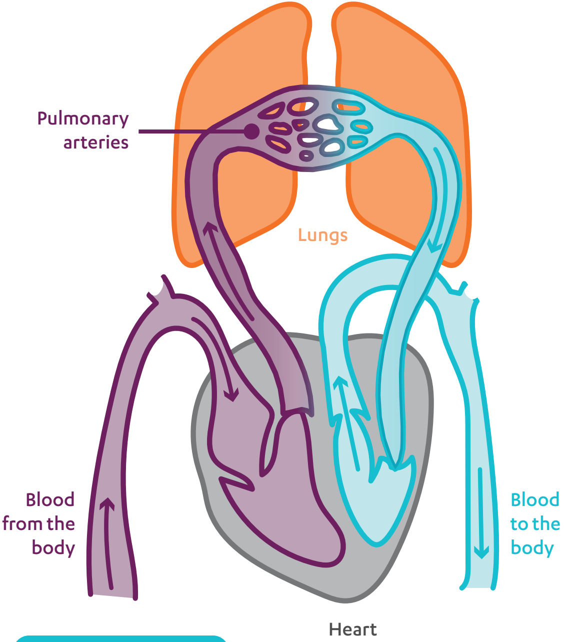


Severely narrowed artery

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.

PAH is a serious condition that gets worse over time or 'progresses'. While there's no cure for

PAH yet, there are medicines that can help slow it down (your healthcare provider may call this, ‘delaying progression’).



Treating PAH

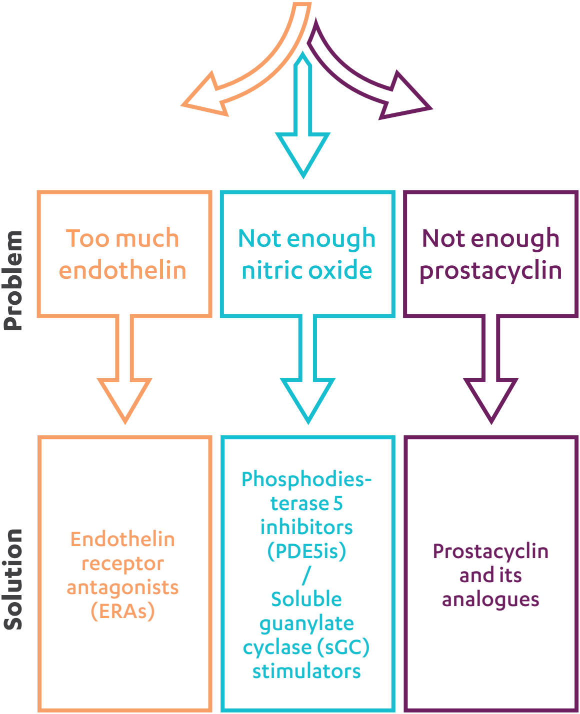
KNOWLEDGE

When you have PAH, there are 3 different ‘pathways’ in your body that may not be working their best for you. Each of these pathways has its own treatment.

Experts who know about PAH believe that the most important goal for treatment is to slow down the speed at which PAH gets worse.

Your healthcare provider may prescribe one or more medicines to treat as many pathways, as early as possible, to improve your chances of feeling better, longer.

Speak to your doctor about your medicine choices as they know 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. You and your doctor can work together to try the combinations that work best for you.



Problem	Too much endothelin	Not enough nitric oxide	Not enough prostacyclin
Solution	Endothelin receptor antagonists (ERAs)	Phosphodiesterase 5 inhibitors (PDE5is) / Soluble guanylate cyclase (sGC) stimulators	Prostacyclin and its analogues

Managing Risk

KNOWLEDGE

PAH affects you in ways that you can't always see or feel. Your doctor may call this "silent progression." Your doctor will always be working to lower your risks from these unseen changes.

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:

- **Blood tests**
- **ECG (electrocardiogram)**
  to record the electrical activity of your heart
- **Echocardiogram**
  an ultrasound scan of your heart
- **6-minute walk test (6MWT)**
  to measure how far you can walk in 6 minutes
- **Right heart catheterization**
  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

Your doctor should talk to you about your test results and treatment options after each risk assessment. Be sure to ask your doctor about any changes since your last assessment. You should also talk to your doctor about treatment choices.

KNOWLEDGE

Gathering People to Support You

KNOWLEDGE

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

KNOWLEDGE

When you have PAH, your healthcare team is there to find the best treatments for you. You can make their job easier by making sure your needs are understood. Be open with the team by 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.

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 new prescribed medication or treatment. 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. Your healthcare team can't improve your treatment plan, or be better partners with you, if they don't know there's a problem. When they know your honest opinions, you can work together to adjust your plan.

- **Come prepared**
  Reflect on what's important to you before each appointment. Bring any lists, logs, and questions with you—including cards in your *Breathe In, Speak Out Toolkit*. During your appointment, keep your lists and questions in front of you.

- **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

To help you name and share your feelings

Joyful

What’s bringing me hope and energy?

EMOTION

Excited

What's most exciting in my life right now?

EMOTION

Thankful

What am I most thankful for?

EMOTION

Optimistic

What am I looking forward to?

EMOTION

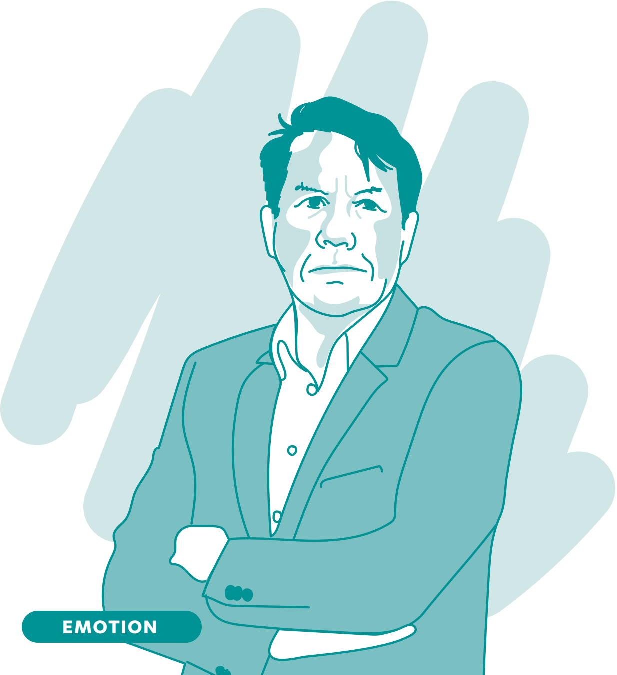
Encouraged

What recent accomplishments make me feel proud?

EMOTION

Eager

What do I want to accomplish?



EMOTION

Relieved

What do I look forward to returning to?

EMOTION

Lonely

What do I want to say to others?

EMOTION

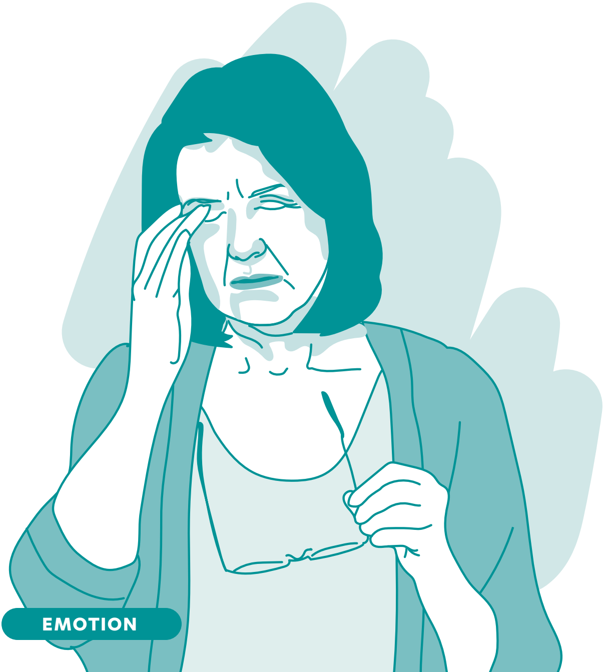
Anxious

What's one small step I can take to feel better ?

EMOTION

Frustrated

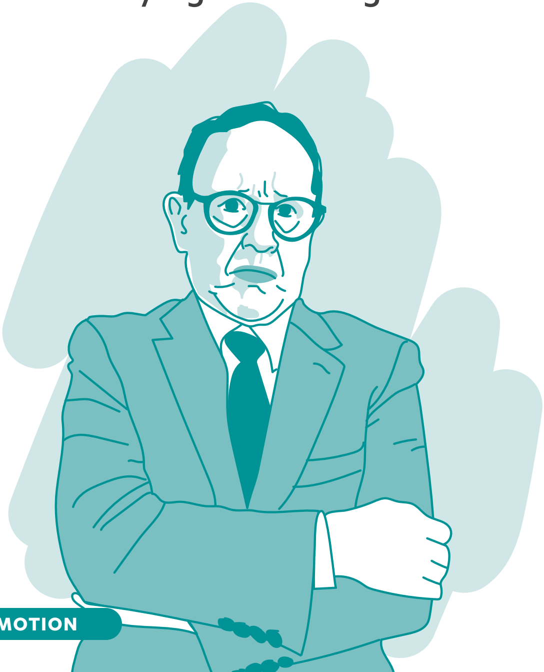
What would bring me some relief?



EMOTION

Afraid

Is fear preventing me from doing or saying something?



EMOTION

Sad

What do I need to feel supported?

EMOTION

Overwhelmed

What are things that are in my control?

EMOTION

Scared

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

EMOTION

Stressed

How am I coping with the stress?

EMOTION

Confidence Cards

With tips and quotes from people living with PAH

“It’s about mentality not physicality. Don’t let PAH change the course that you want to take.”

—Person living with PAH

CONFIDENCE

> “Start accepting that the disease is not you, just a curve in the road. There are good moments and bad, but take 1 day at a time.”
>
> —Person living with PAH

> “Keep your mind set on your goals, as a glimmer of positivity can improve your day, month, or even your year.”
>
> —Person living with PAH

CONFIDENCE

> “I don’t let the disease define who I am. I’m going to keep on being me.”
>
> —Person living with PAH

CONFIDENCE

> “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

CONFIDENCE

“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

CONFIDENCE

> “I’ve found it’s 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 vocalize any concerns I have. After all, I am the only one inside my body.”
>
> —Person living with PAH

CONFIDENCE

> “With PAH, it’s important to be proactive.”
>
> —Person living with PAH

“Be honest with your doctor. Don’t say it’s fine when it isn’t.”

—Person living with PAH

CONFIDENCE

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 organize your thoughts and create a plan that works for you.

REFLECTION

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.

On my own...

With friends and family...

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.

On my own...

With friends and family...

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.

REFLECTION

My goals for my health and managing PAH…

I prefer a treatment that will allow me to…

My long-term goals and milestones…

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...

Supports my treatment decisions

Supports my emotional needs

Supports my social needs and hobbies

Supports me around the house

Supports me during an emergency

Supports me in celebrating milestones

janssen
PHARMACEUTICAL COMPANIES OF
Johnson & Johnson