

# PAH TODAY

## ARTIST, LEADER, PATIENT

Karen's battle  
with PAH leads to  
renewed creativity

## PAH 101:

A beginner's  
guide to PAH

## PAH AND EXERCISE

Ways to work  
exercise into  
your PAH  
treatment plan



**PAH Initiative**

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# WELCOME TO PAH TODAY!

## From the PAH Initiative

Welcome to the PAH Initiative and the first issue of our magazine, *PAH Today!*

The PAH Initiative is sponsored by United Therapeutics as part of an ongoing commitment to improving the lives of patients and supporting all those who care for them. United Therapeutics was founded by the parents of a child living with pulmonary arterial hypertension (PAH), so we understand the challenges presented by this rare and complex disease.

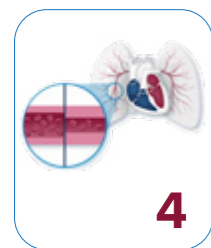
Created as a resource for the PAH community, the PAH Initiative reflects United Therapeutics' commitment to providing knowledge and inspiration based on science and research in PAH today. We are here for you, providing information and inspiration for your journey with PAH. That's what this magazine is all about.

Sincerely,  
The PAH Initiative

Visit [PAHInitiative.com](http://PAHInitiative.com) and like the PAH Initiative on Facebook and Instagram



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# PAH 101:

## A Beginner's Guide to PAH

PAH is a specific type of high blood pressure that affects your heart and lungs.

### P-A-H

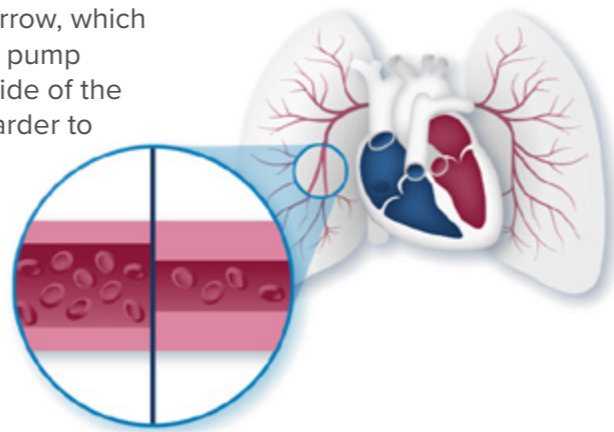
**Pulmonary** adj. [ puh-l-muh-ner-ee ]  
having to do with the lungs

**Arterial** adj. [ ahr-teer-ee-uhl ]  
related to blood vessels that get or carry oxygen to the rest of the body

**Hypertension** n. [ ha-hy-per-ten-shuhn ]  
high blood pressure

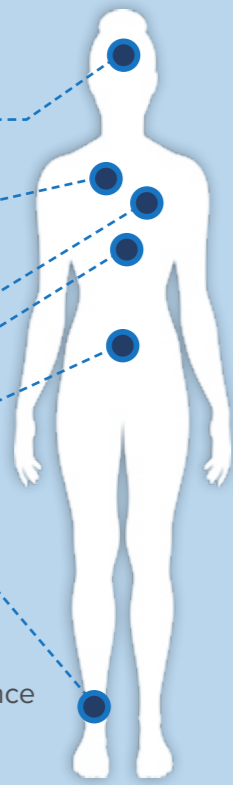
When you have PAH, the blood vessels in your lungs narrow, which means the right side of your heart has to work harder to pump blood to your lungs. This creates pressure on the right side of the heart. The heart then tries to compensate by working harder to pump blood through the narrowed vessels in the lungs.

As blood vessels remain narrow and the heart continues to work overtime, the heart struggles to maintain this level of intensity, and you begin to experience PAH symptoms. You may feel symptoms because less blood and oxygen reach parts of your body where they're needed.



### Common PAH Symptoms

- Dizziness/fainting
- Shortness of breath/difficulty breathing
- Rapid, irregular heartbeat
- Chest pain
- Swollen abdomen
- Swollen ankles



**Note:** These are not all of the symptoms of PAH. You may experience other symptoms.

Over time, as PAH progresses and blood vessels remain narrowed, the symptoms you feel may get worse and you may find it harder to do everyday activities, like yardwork, doing the laundry, walking the dog, or preparing a meal.

It's important to note that symptoms may not always reflect whether PAH is progressing or how it is affecting your body. Even if your symptoms remain the same, there still may be more you can do to improve how you feel. If you have specific questions about your PAH, your symptoms, or how you are being monitored, talk to your healthcare provider as everyone's situation is different. 🏠

**PAH is a serious, progressive disease—meaning it gets worse over time. However, there are PAH medications that can help people living with PAH improve their symptoms. Visit [PAHInitiative.com](http://PAHInitiative.com) to learn more about how PAH is managed today.**



# PAH AND EXERCISE

## Ways to work exercise into your PAH treatment plan

*Please consult with your physician or healthcare provider before starting an exercise routine. Do not start an exercise routine if your physician or healthcare provider advises against it. This article is offered for educational purposes only and is not a substitute for nor replaces professional medical advice, diagnosis, or treatment. If you act on information contained in this article, you agree that you are doing so at your own risk. **IN THE EVENT OF A MEDICAL EMERGENCY, PLEASE CALL EMERGENCY SERVICES IMMEDIATELY.***

When living with pulmonary arterial hypertension (PAH), the thought of exercising can be intimidating. You might even be wondering: *Can I exercise? Where would I even start?* In the past, people with PAH were often advised against exercising because, during exercise, the heart has to work harder to pump blood to the body, especially the arms, legs, hands, and feet.

Today, researchers and doctors increasingly see the benefits of exercise for people with PAH when done safely and in close coordination with a healthcare team. In fact, along with PAH medications, exercise is now considered by many as an integral part of a comprehensive PAH treatment plan.

**Continued on next page...**

**The Evidence for Exercise**

So, what are the benefits of incorporating exercise into your treatment plan? Let's look at the evidence. Studies show that a 12-week exercise training program for people with PAH increased endurance and six-minute walk distance, which is used to assess exercise capacity. Some of the other physical benefits shown in these studies include improved WHO functional class and lowered blood pressure.

Beyond the evidence for physical improvement, studies also show that an exercise program may improve quality of life for people living with PAH, including reduced depression and fatigue.

**Getting Started with an Exercise Program**

If you're wondering how to get started or if an exercise program is right for you, talk to your healthcare team. Recommendations for exercise vary from patient to patient, so it's important to work with a PAH specialist to develop a program that's uniquely designed for you. If available, your doctor may recommend a cardiac or pulmonary rehabilitation center as part of your program.

Regardless of the setting, exercising under supervision is key—a specialist will be able to assess your needs and abilities while monitoring how exercise is impacting you.

Every exercise program will be different, but examples of programs studied in clinical trials of patients with PAH include bicycle training, walking, dumbbell training, respiratory training, endurance training, treadmill walking for 30–45 minutes, aerobic interval training, step climbing, and resistance training. The regimens studied in clinical trials generally range from 10–15 weeks.

**Remember:** an exercise program that is right for some patients may not be right for others. Talk with your doctor about what is right for you.

Always consult with your healthcare provider before starting an exercise routine.



To learn more about exercise, visit the PAH website at [LifewithPAH.com](http://LifewithPAH.com).

**Tips for Exercising Safely**

Once you've worked with your healthcare team to develop the right plan for you, consider the following tips from the Pulmonary Hypertension Association (PHA) for exercising safely:



**Never exercise alone**  
Always exercise under supervision or with a partner.



**Always warm up and cool down**  
Remember that exercising with PAH may mean you need a longer, more gradual warm-up.



**Check in with yourself**  
Keep track of how you're feeling and avoid exercising to the point of light-headedness or dizziness.



**Watch your breath**  
Some shortness of breath is acceptable but make sure you can still control your breathing and talk. Contact your doctor or call 911 if you're unable to regain control of your breathing after five minutes.



**Start small**  
Don't push yourself—gradually build up your endurance.

**Remember:** Always consult your healthcare team before starting a new exercise plan.

**HOPE IS IN OUR HANDS**

Visit [phpal-app.com](http://phpal-app.com)

United Therapeutics has developed a patient and care partner PH app called PHpal™.

This exciting new app provides opportunities for you to be better informed and feel more in control of your disease.



# FROM MOM TO CAREGIVER AND EVERYTHING IN BETWEEN

A chat about balancing self-care and caregiving



**Jane is a caregiver to her 25-year-old daughter, Nicole, who was diagnosed with pulmonary arterial hypertension (PAH) when she was 15. For a time, Jane “lost herself” in the roles of caregiver and mother. But over the past decade, Jane has learned the important lesson of caring for herself, too, and what it means to be a mother, a caregiver, and an individual.**

**Q: Jane, can you tell us a little about how your daughter was diagnosed with PAH?**

A: Nicole was born with a congenital heart defect, but we didn't know that at the time. For most of her life, she had issues catching her breath when she was running, and I noticed that her nails would turn blue sometimes. When she was younger, I didn't think too much of it. Looking back now, it's my biggest regret that I didn't advocate for my daughter's health sooner. When she was 15, it got to the point where she couldn't ride her bike without having to stop because she was having trouble breathing, or if she dropped something, she had trouble picking it up. So, I took her to the doctor, and they did some diagnostic testing that showed she had PAH.

**Q: In caring for Nicole, how do you find the time to take care of yourself, and why do you think self-care is important?**

A: It's changed a bit over the years. When Nicole was born, I lost who I was a little bit. My sole focus became raising her and that was enhanced when

she was diagnosed. But I have learned that I need to take care of myself too. So, what I do is wake up extremely early, before anyone else is awake, and I'll use that as my quiet time. I use my mornings to read the Bible, maybe do a class, or just sit quietly. If I get stressed throughout the day, I look back on that quiet time and it helps me. Sure, it's not as much time as I would like to have, but it gets me through the day. I also think healthy eating is important, so I grow a lot of herbs and incorporate them into meals for Nicole and me. I don't exercise as much as I probably should, but I go for regular walks with Nicole, which is relaxing to me.

**Q: You and your daughter have been through a lot together over the past decade. How have you both adjusted to your changing roles as Nicole became an adult?**

A: As Nicole has gotten older, my involvement in her care has changed a little bit. I try to help Nicole only when she specifically asks for help, and we discuss what she expects of me as her caregiver. I still manage all the administrative aspects of her care. I schedule the appointments and talk to the doctors with her. For the most part, I encourage her to make the decisions about her treatments and to monitor her health because she's an adult now. Of course, if she asks for help, I'll be there for her.

**Q: It sounds like you all function well as a team. How do you and Nicole work together to monitor her health?**

A: Typically, Nicole has an echo done at least once a year or her PAH specialist will do a right heart catheterization when warranted. Nicole is actually very good about tracking her symptoms and we use a shared online document, where she writes down everything from her weight, her oxygen saturation levels, and her blood pressure. She also keeps a food diary so she can better understand whether changes in her vitals are triggered by something she's eaten or if it's something more serious. I let her monitor all of that herself and I just keep an eye on the numbers so I can know what needs to be mentioned to her doctors in case she forgets or feels uncomfortable discussing it.

**Q: You've been Nicole's biggest supporter over the years, but where have you found support as a caregiver?**

A: About six months after Nicole was diagnosed, I found the Pulmonary Hypertension Association and that changed everything for me. That's how I got connected with a support group, and they became like a family to me. It made me feel less alone and eventually I became a co-leader of the support group. Over the years, it's become

important to me to help those who are newly diagnosed with PAH because I know what they're going through. Having that connection helps me realize how important it is to show kindness to people because they might have an invisible illness. Now, I look around and I know that just because someone doesn't look sick, it doesn't mean they aren't. Everyone has their own story.

**Q: Reflecting on your experience as a mother and a caregiver, what advice do you have for others caring for a loved one with PAH?**

A: I think it's important to set boundaries and expectations with the person you're caring for. Each relationship is different, but for example, with Nicole, I only help her when she asks for it. This is hard because I'm her mom and I always want to help. But I know what she needs and expects of me and I have to give her that space to be as independent as possible.

The most important thing I can say is that a caregiver needs to take care of themselves, whatever that means for each person. If you find joy in exercising five days a week, go for it! Finding something that's just for you is necessary because it reminds you that you're not just a caregiver. Your needs are important too. 🍷

## STAY UP TO DATE WITH PAH TODAY

Looking to learn more about PAH? Check out the PAH Today National Broadcast series to hear from nationally recognized PAH experts on topics including:

**Beyond the Basics:** How PAH affects your heart and lungs to cause symptoms

**How Decisions Today Can Shape the Future:** How risk status helps your doctor assess your progress

**The Pathways to Treatment:** How medications work to treat PAH and why the right combination matters



Scan here to watch the videos today or visit [PAHevents.com](http://PAHevents.com).

# ARTIST, LEADER, PATIENT

Karen's battle with PAH leads to renewed creativity

## Everything, all at once

Karen barely remembers that day. And yet, it's one she'll never forget.

She rose from her hospital bed and made her way to the bathroom, her hands clutching a pillow. She remembers *SNL* playing quietly on the TV in her dark room.

After closing the bathroom door, she rested her face against the sink. She breathed in. Then, she collapsed.

"I'll never forget it," she says. "That's when everything went black."

Karen was experiencing right heart failure. She then spent the next eight days in the ICU. Of that, she says she remembers very little. Many of the IVs, tubes, tests, procedures, and specialists were a blur.

She had initially landed in the emergency room

after receiving an abnormal EKG result from an outpatient urgent care center. For years, she'd been told that it was only high blood pressure that was causing her symptoms. The nurses at the free clinic she'd gone to just the week prior didn't have an EKG and Karen didn't have health insurance.

Yet, despite the false alarms and financial roadblocks, when the time came, Karen knew she needed help. She trusted her gut.

And she was right. Just two days after fainting in the hospital, a right heart catheterization revealed that something really was wrong.

Karen was diagnosed with pulmonary arterial hypertension, or PAH.

Then, the real work began.

## Making a change

In 2013, Karen was thrust into an unfamiliar world. At just 48, she was told that she had a very rare,

life-threatening disease.

"Hearing that I was going to need an infusion pump and be on oxygen 24/7 made the seriousness of my condition set in. I started bawling," she says.

She was told she could no longer work at the salon as a hairstylist and decided to move in with her parents. She spent every hour of every day on oxygen.

"I lost 60 pounds in just two months. I felt like death."

She says the first year after her diagnosis was nearly impossible at times. Karen struggled with her weight, her oxygen tanks, and walking.

On top of her PAH diagnosis, she needed hip surgery. If she chose not to have it, she'd have to use a wheelchair, though surgery carried significant risks also.

**"Hearing that I was going to need an infusion pump and be on oxygen 24/7 made the seriousness of my condition set in. I started bawling."**

Continued on next page...



Karen decided to have surgery. She weighed the pros and cons with her healthcare team. She knew that if she was no longer able to walk, it might be harder for her to lose weight and stay healthy. She knew what she wanted to do. So, she gritted her teeth and took the plunge.

After she woke up from surgery, the recovery was exhausting. But she felt as though she could handle it, no matter how hard. More than anything, she was just grateful that she woke up, and after, she vowed to overcome any challenge she faced.

She spent the next few months working with her healthcare team to reclaim control of her body. She lost more weight, no longer required supplemental oxygen, and started participating in PAH events. She made drastic changes to her lifestyle and was determined to do hair in some capacity. She made close connections with PAH support groups and other people living with PAH that helped her on her journey. She persevered.

#### Learning through leading

In 2017, Karen took another big leap. She became a PAH support group leader through the Pulmonary Hypertension Association. Before, she had to travel over an hour to attend the group meetings. Then one day, the idea came to her.

“I figured, well, patients have to start the groups... So why don't I just do it?”

Since then, Karen has run a well-attended support group in her town. She loves connecting with her friends and sharing key insights, experiences, and information.

Karen often attended PAH-related events and met with fellow PAH patients in person. She took every opportunity to connect with the community. That is, until March 2020, when much like the rest of the world, she suddenly found herself cut off.

“I was terrified. I thought we were all going to get COVID and that would be it,” she says, with a sigh.

The pressures of social isolation and the fear of contracting the novel coronavirus overwhelmed her. That spring, she left the house only a handful of times, never straying beyond her mailbox.

But Karen waded through her anxiety and tried to be there for everyone, though it was hard. She found ways to cope, like journaling and learning about COVID-19 through the Pulmonary Hypertension Association website and social media channels. But at the first virtual sessions, she saw less attendance, and the other group members reported feeling pressure from living through a pandemic too.

Eventually, though, after months of wiping down packages and staying home, Karen was relieved as information about COVID-19 and PAH patients became available.

“Together, the group learned more about what to expect with COVID and PAH. As time went on and we gathered more information, we all worried only as much as we needed to,” she says.

Now, Karen has become more comfortable with her new “normal” and maintains her relationships with the PAH community. When it comes to members who no longer attend meetings because of technical issues or lack of access, she calls once a month to check in.

“It's fun now, even virtually! We had a big virtual Christmas party, scavenger hunts, and costume parties,” Karen says.

#### Care with a little creativity

Though she stays busy with her group meetings and keeping up with her care, Karen still finds time to explore her interests. She cooks, pet sits, participates in PAH Initiative activities as a patient Ambassador, and does hair as a hobby at friends' homes, but spends the larger part of her time creating watercolor paintings.

“I had never done anything like it before,” she says.

What started on a whim developed into her new, unrivaled passion. A few years ago, Karen graduated from coloring books—which she'd often fill up completely—to full-blown paintings featuring florals and bright hues.

“At a PAH fundraising auction, we auctioned off one of my paintings. We raised a lot! I thought it was amazing that I could do that with my art. It was the first painting I ever sold,” she says.

Karen uses painting to unwind and believes that creative outlets can be calming for people living with PAH. Now, as an artist, leader, and patient, she advocates for PAH in her local community as well as across the country.

She's proud of the friendships she's made, even if they started from an unlikely place, or even a rare, unwelcome diagnosis.

“What can I say?” she says with a laugh. “I have a big, messed-up heart.” 🧡

**“At a PAH fundraising auction, we auctioned off one of my paintings. We raised a lot! I thought it was amazing that I could do that with my art. It was the first painting I ever sold.”**



# PAH Life Hacks

## Tips from people who've been there

Life with PAH can come with a lot of unknowns. Below, PAH Initiative Ambassadors share their resourceful tricks for tackling life's everyday obstacles.

**Q:** What techniques or tools do you use to make showering less tiring and more comfortable?



**Nicole**, living with PAH: I take cool or lukewarm showers because heat makes me tired. I find that a detachable showerhead can help you move less, control where the water is going, and keep the catheter site dry for people using infused pump therapy.



**Jane**, caregiver to daughter, Nicole: When Nicole was first diagnosed, we got her a shower chair. That has been great, and she can wear her oxygen in the shower. On the days she needs to wash her hair, I help her out.

**Q:** What kind of recipes are easy to prepare, and what should those with PAH aim for when it comes to their diet?



**Denise**, living with PAH: Meal prep and planning is such a big help. That and a low-sodium diet. Prepping is key to sticking to that diet. Even cooking ahead when you have energy can help you out later when you're too tired. Slow cookers can also make things easier. It also helps to have an easy go-to food that you like and can eat anytime so that on bad days, you can just grab it and go. For me, that's things like hard-boiled eggs, yogurt, or fruit!

**Q:** When it comes to household chores like laundry, cleaning, or cooking, are there any shortcuts that can save time and energy?



**Lauren**, living with PAH: Set goals for yourself, especially in terms of physical activity (under the instruction of your doctor, of course). Stay focused and challenge yourself to function the best you can physically, mentally, and emotionally.



**Gwen**, living with PAH: I know I can't do everything in one day, so I make a to-do list of what I will accomplish each day; I always give myself one day of rest—Monday.

**Q:** What advice or inspiration would you like to share with other PAH patients?



**Karen**, living with PAH: Plan your life. Spend time doing something that you love, something that gives you joy, whether painting, reading a good book, or spending time on the phone with a friend.



**Mike**, living with PAH: The best advice I can give to someone living with PAH is to be your own advocate. No one knows better than you what you need. Speak up and get the help you need. Seek out a PAH specialist to ensure you receive proper care. 🗨️

To hear more from our PAH Initiative Ambassadors, visit [LifewithPAH.com](https://lifewithPAH.com).

# PAH Resources

## Helpful support for people with PAH

No matter where you are in your PAH journey, know this: You're not alone. The PAH Initiative is proud to join the ranks of multiple organizations committed to supporting, educating, and inspiring the PAH community. Get the support you need—check out this list of PAH resources.



### Grow Your PAH Knowledge

**PAH Initiative:** Stay informed about PAH through a variety of educational tools, resources, and information, including the PAH Initiative Learning Library. Visit [pahinitiative.com](https://pahinitiative.com).

**PAH Initiative Newsletter:** Sign up to receive helpful information and updates from the PAH Initiative. Visit [pahnewsletter.com](https://pahnewsletter.com).

**Pulmonary Hypertension News:** Get the latest news coverage of PH science, medicine, and human-interest stories. Visit [pulmonaryhypertensionnews.com](https://pulmonaryhypertensionnews.com).

**phaware® Global Association:** Stay #phaware by accessing digital content focused on PH education, resources, and knowledge, including the "Aware That I'm Rare" Podcast. Visit [phaware.global](https://phaware.global).

**PHA Classroom:** Browse the Pulmonary Hypertension Association's (PHA) online learning center for a variety of educational videos and resources. Visit [phassociation.org/classroom](https://phassociation.org/classroom).



### Connect with Community

**PAH Initiative Facebook and Instagram:** Connect with the PAH Initiative on Facebook and Instagram for more stories, information, and inspiration.



**PHA Support Groups and Events:** Learn more and find others in the PAH community by joining one of hundreds of virtual and in-person support groups or attending an event near you. Check out the PHA network at [phassociation.org](https://phassociation.org).



### Help Manage Your PAH

**PHpal App:** Explore an exciting new app developed by United Therapeutics that provides opportunities to be better informed and feel more in control of your PAH. Download the free PHpal App from the App Store or Google Play. 📱







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**knowledge**  
meets  
**inspiration**

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