PATHLIGHT

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INNOVATION for an Independent Life

PLUS

'Medical Heroes' Behind the Data

The Future of PH Care

Participating in Research During the Pandemic

30 YEARS
LIGHTING THE WAY FORWARD

PATHLIGHT

2021 // Volume 30 No. 2

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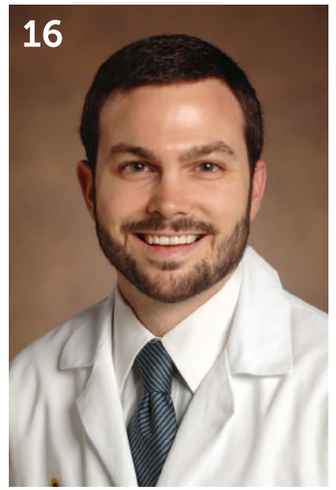

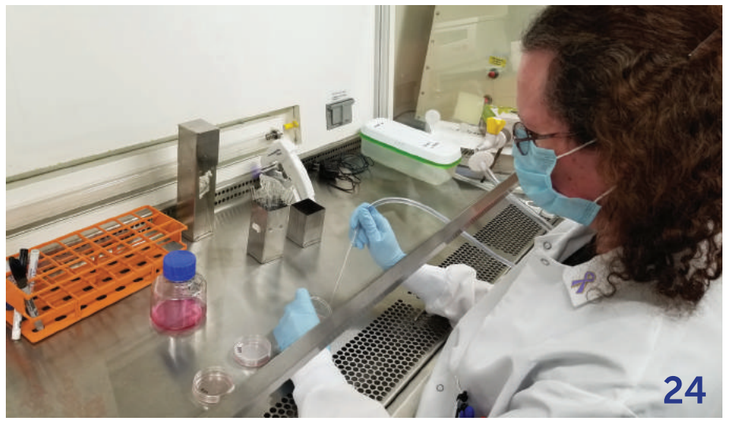

On our cover: Shavini Fernando invents a wearable device to monitor oxygen levels. This month's issue covers advancements in PH research.

IN EVERY ISSUE

PATHLIGHT

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PHA Pulmonary Hypertension Association
Empowered by hope

LETTER FROM THE CHAIR

To me, there are four words that most define the pulmonary hypertension (PH) journey. They are hope, community, advocacy and research. Clinical trials encompass all of them.

Trials give us hope. We're lucky to have the medications we have now, but there is still a way to go as far as managing difficult side effects, slow or maybe someday reverse PH, and of course, get to the actual cure. With each clinical trial, we have hope of getting there sooner.

While clinical trials are conducted on individuals, they are also about community. Clinical trials and participants hold sacred spots in the PH community. When someone mentions they have participated in a trial, others rally around to thank them. We know trials can be challenging and are experimental. We are grateful for those who put themselves forward so we can all have a better tomorrow.

Then there is the advocacy and research part of trials. Funding for trials comes from all different sources. Thus, we also have opportunities to advocate for federal funding and raise money or donate to PHA to fund its research initiatives.

I'm so pleased that this issue of *Pathlight* magnifies the importance of clinical trials and PH research. In this issue, you will find inspiring stories of scientific advancement and hear from medical professionals and patients about their experiences with trials.

Our cover story, "Innovation for an Independent Life," is a first-person story by software developer and entrepreneur Shavini Fernando. Shavini, who has PH, invented a wearable device to continuously monitor oxygen saturation. Shavini, originally from Sri Lanka, created her company, OxiWear, while pursuing a master's degree at Georgetown University.

In "Trial Participant Takes Control of Her Health," Catherine Falardeau shares her story of testing a drug for PH related to interstitial lung disease (ILD). While her trial medication, Tyvaso, has been on the market for over a decade, its success and ultimate approval for ILD patients was established in this important trial. It's very exciting when we can expand the use of a known therapy to help more patients.

In "Medical Heroes Behind the Data," you'll hear from three patients who have participated in clinical trials: Monica Penaranda, Linda Sullivan and Susan Huber. Each brings a unique view to the process, and a voice of hope for the future.

"The Future of PH Care" takes us behind the scenes with Corey Ventetuolo, M.D., and Steven Kawut, M.D., M.S. The doctors piloted a study during the COVID-19 pandemic that could validate at-home six-minute walk tests. Such digital tools expand the possibilities of how patients and doctors can work together to monitor health progress. While the pandemic has brought many challenges, expanding our abilities to connect and communicate by harnessing technology is surely one of the bright spots.

No matter the reason for your participation, financial support or scientific interest in clinical trials, it's clear that research moves the needle forward for the lives of those affected by PH. Every trial brings us a step closer to improved quality of life, better clinical outcomes and perhaps a cure someday. We all have a stake in that!

Warmly,

Colleen Brunetti

Colleen Brunetti, M.Ed., C.H.C.

Board of Trustees Chair,
Pulmonary Hypertension Association

Influencing Change

PH Advocates Urge Health Care Reforms

Pulmonary hypertension (PH) advocates are sharing their stories to urge reforms that address treatment and insurance challenges.

In recent months, advocates have addressed step therapy, copay accumulators and high costs of PH treatment with members of Congress and state legislators.

"We need to take our personal stories to Congress and let them know what our life and experiences are," says Karen Fragale of Johnston, Iowa.

Karen spoke with legislative staff from the offices of U.S. Sens. Joni Ernst and Chuck Grassley, both Iowa Republicans. Karen shared how she was discharged from the hospital without a referral or prescription after a medical professional told her family that she had less than a year to live.

'We need to take our personal stories to Congress and let them know what our life and experiences are.'

– Karen Fragale

Fighting on the federal front

Karen struggles with health insurance plans that routinely refuse to cover her PH medications after the first year or that require her to shift to less expensive medications than those her physician prescribed. Karen asked her senators to help reduce those practices by co-sponsoring the Safe Step Act.

Similarly, Gerry Langan spoke with staff from the offices of Sen. Marco Rubio, R-Florida, and Rep. Charlie Crist, D-Florida.

Until recently, Gerry had minimal out-of-pocket costs for her PH medications because her husband serves in the military and they were covered by Tricare. When Gerry's husband shifted to military reserve to take a federal job, the family had to switch health plans, which dramatically increased Gerry's out-of-pocket costs.

'The only way to make real change it is to put yourself out there.'

– Gerry Langan

Because Rubio is a leader in the health appropriations process for the federal budget, Gerry also encouraged the senator to increase NIH funding for FY 2022.

"I encourage everyone to find their voices and share their story," Gerry says. "The only way to make real change it is to put yourself out there."

Seeking state-level change

On the state level, PHA Chair Colleen Brunetti recently testified before the Connecticut legislature from a patient's perspective. She asked the General Assembly's Insurance and Real Estate Committee to support of a bill to eliminate copay accumulators, which have affected her personally.

Copay accumulators allow health insurance companies to make money from copay cards and grants while keeping patients' out-of-pocket responsibility high. For example, an insurance plan might accept copay cards for PH medications but won't apply them to deductibles or out-of-pocket maximums.

Interested in sharing your story with your lawmakers? The Pulmonary Hypertension Association provides free advocacy training. Contact Advocacy@PHAssociation.org, or call 301-565-3004 x749.

TESTING TYVASO

Trial Participant Takes Control of Her Health

When Catherine Falardeau was diagnosed in 2018 with pulmonary hypertension (PH), her doctor suggested she participate in a clinical trial. Just last month, the treatment Catherine tested received Food and Drug Administration (FDA) approval for people with Group 3 PH associated with interstitial lung disease (ILD).

Catherine, 59, of Kissimmee, Florida, was among the first to test Tyvaso (treprostinil) Inhalation Solution for Group 3 PH. She participated in a clinical trial for Tyvaso from February 2018 to September 2020. The treatment, manufactured by United Therapeutics, is designed to improve patients' exercise ability. The FDA approved Tyvaso in 2009 to treat Group 1 PH.

Catherine's PH journey began in 2015, when she started feeling out of breath and constantly was coughing. She had been a regular runner, but the short walk down her driveway to the mailbox left her breathless. Later that year, Catherine was diagnosed with scleroderma and mixed connective tissue disease.

Three years later, a right heart catherization showed Catherine had pulmonary arterial hypertension (PAH) with ILD, a group of lung diseases characterized by marked scarring or fibrosis within the lungs.

Signing on

When her cardiologist at Advent Health in Orlando recommended she participate in the Tyvaso trial, Catherine gladly signed on. Testing was included in the trial, and she received a United Therapeutics grant to participate.

"I had never done a clinical trial before, so I didn't know what to expect," she says. "Tyvaso was the first course of treatment I went on. My cardiologist felt that because I also had ILD and mixed connective tissue disease, it was the best course of treatment for me."

During Phase One of the trial, Catherine wasn't sure whether she was receiving the drug or a placebo. She believes she received the placebo because she didn't seem to be improving. At the six-month mark, patients learned everyone would receive Tyvaso in Phase Two.

Breathing easier

Catherine felt sick to her stomach the first four months of taking the drug, but luckily those symptoms subsided. She noticed she could breathe better by January 2019. Her coughing greatly

Catherine in 2009 after the 2009 Torch Relay walk for the Children's Miracle Network Hospitals.

Dachshunds Archie and Edith join Catherine on her evening walks.

diminished throughout the trial, and she could resume normal activity without feeling tired.

Every month, Catherine underwent pulmonary function and six-minute walk tests. She also opted in for genomic testing, which examines how genes interact and affect one's health. At first she had to have blood drawn at each visit. Eventually, the blood tests were conducted every three months.

Catherine's six-minute walk tests improved with each visit to the clinic. At the start of the study, Catherine says she walked about 488 meters during her tests. By September 2020, she was up to 530 meters.

"As time went on … I started looking forward to getting the testing done," she says. "I learned a lot about the stats they look at and what the tests actually mean. I was able to take control of my own health."

Health improvements

Now, Catherine takes hour-long walks around her neighborhood with her husband, Wayne, and their two dachshunds, Archie and Edith. She can walk to her mailbox without feeling winded. She says she has tried spurts of running and has made it around her half-mile block. Stairs are still hard, but she tries to take them every time she visits her doctor. Catherine looks forward to her cardiologist appointment to see how her tests improve.

Catherine continues to take Tyvaso, which she receives for free from United Therapeutics. She says her whole attitude has changed for the better. In December 2020, she changed her diet after learning she had a blockage in her carotid artery. She since has lost 20 pounds.

> 'It meant so much to be part of a successful study. The only thing that tops it is the feeling of hope. I feel so much better.'

Catherine connects with other PH patients who have connective tissue disease as a volunteer for the Pulmonary Hypertension Association (PHA) Email PHriends program. Some of her phriends also take Tyvaso for their PH, and Catherine says she encourages them to continue the treatment despite initial side effects.

"It meant so much to be part of a successful study," she says. "The only thing that tops it is the feeling of hope. I feel so much better."

‘MEDICAL HEROES’ BEHIND THE DATA

Study Participants Push Science Toward Better Treatment

Clinical trials and research studies are essential to understanding and treating pulmonary hypertension. And patient participation is critical for both. Three pulmonary hypertension (PH) share their experiences participating in research and how it has changed their lives.

Monica Penaranda

Hacienda Heights, California

I can’t remember how many clinical trials I have participated in, but each one made me feel like I was getting closer to a goal: potentially the perfect treatment for me or even a cure. I didn’t fear going into a study. I had hope.

When I was diagnosed with severe pulmonary arterial hypertension (PAH) 23 years ago, I was offered only one treatment. Anything that came after 1997 was considered a trial.

I started Remodulin under its trial name UT-15 in 2001. I was excited to have a smaller pump and to be on a drug that lasted longer than three minutes in my system.

When I transitioned from Flolan to Remodulin, I was happy that my side effects were about the same, not worse. My breathing remained stable. It was almost as if I had made a lateral change as far as PH symptoms and side effects.

However, the pain from the subcutaneous injection was more than I could handle. I decided to go back to IV, but I stayed on Remodulin. I was a part of that trial for three years.

Newfound freedom

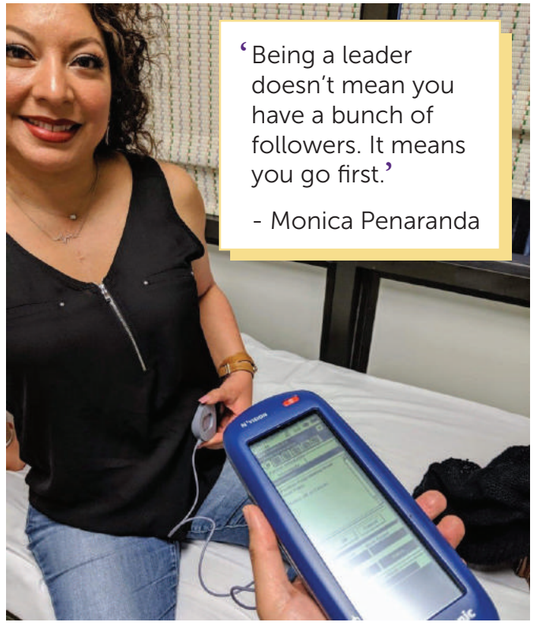
My doctor told me years ago that one day a pump would allow me to swim without worrying about infections or pain. It seemed like a distant dream, but it became a reality when I started a trial for a new pump.

Ten years ago, I was the 15th person to receive an implantable system for Remodulin. It was a leap of faith but a blessing. There was no difference in my breathing or PH symptoms because I received the same amount of medication with the internal pump as I had with the external pump.

No longer having to drag my external pump around was liberating. I can’t tell you how many times I had to take my pump with me before the implant just to grab a pen off the counter. Living with an external pump had limitations. I lived in fear of infections, I avoided pool parties, and showers were a chore.

When I received my implant, I felt free. Showers became my favorite part of the day.

When I attended the Pulmonary Hypertension Association (PHA) 2018 International PH



‘Being a leader doesn’t mean you have a bunch of followers. It means you go first.’

- Monica Penaranda

Monica (right) with husband Brandon (left) and son Bear (center).

Conference and Scientific Sessions in Orlando, I jumped in a pool for the first time in 17 years. I can’t express how much joy that brought me. It is one of the top moments in my life.

I’m still a part of the trial for the pump implant. Every 58 days, I get my pump refilled. As part of the trial, I can’t do my own fills or adjust the rate myself. It has to be done at the hospital.

The biggest benefit is that I haven’t had a hospital stay from a line infection in 10 years. Those infections are no joke.

Sign me up

I credit my long life after diagnosis in part to the clinical trials I participated in. I learned to fight for my life, try new things and keep an open mind. Where would I be if I said no? I fear to answer that question. I treasure every gray hair on my 41-year-old head. I didn’t expect to live long enough to turn gray.

I have an amazing PH team. My doctor, Shelley Shapiro, M.D., of Ronald Reagan UCLA Medical Center, always made sure I knew about trials that seemed promising.

I just concluded my portion of a yearlong stem cell trial, and my doctor recently asked me if I want to start another trial. I said, “Sign me up.” We need people to try new treatments. How else will we know what can make us better?

Being a leader doesn’t mean you have a bunch of followers. It means you go first.

Linda Sullivan

West Islip, New York

Three or four medications were available when I was diagnosed with PAH in 2005. I had a congenital heart defect that caused Eisenmenger’s syndrome. I was lucky to connect with Robyn Barst, M.D., in New York City. She educated me and gave me

Linda (center) with family: (from left) son Matthew, husband Stephen, mother Alice, daughter Catherine.

> ‘Clinical participants aren’t guinea pigs, but medical heroes.’
>
> - Linda Sullivan

peace of mind as I began my new journey.

My first clinical trial was for Adcirca over 10 years ago. I was so nervous and had a million questions. “Will I die from the drug? Will it damage me? Will it make my PH worse than it already is? Should I wait for someone else to be the guinea pig?”

I have since learned that clinical participants aren’t guinea pigs but medical heroes. I believed in science, and I trusted Dr. Barst, so I participated.

Pushing science forward

I was nervous, and I didn’t know if I received the drug or a placebo when I started the trial. It was a 50-50 chance that I would get the drug, but I was willing to take the chance.

After a couple of weeks, I knew I had the drug because I had pain in my back and legs that I didn’t have before. I started to breathe a little better, and I was happy.

Years later, I am still on the drug, and it continues to be part of my PAH cocktail. If I hadn’t participated in the clinical trial, I wouldn’t have known that I could benefit from Adcirca.

I have learned that each person with PH is unique and has a different cocktail of PH medications that works best for them.

In the past 16 years, I have participated in every study I qualified for so I could help scientists and doctors educate themselves and others about this disease. If PAH patients don’t participate in clinical trials, there’s no way to push science forward for better treatments and ultimately the cure that we so yearn for.

No fear

I recently was asked to participate in a research project for cardiac MRIs. The project studies the heart and how the chambers of the heart respond to different pressures. The study uses images taken by cardiac MRI to evaluate pressures in the chambers.

When my doctor asked me about it, I signed on the dotted line. I have learned on my PAH journey not to let fear dictate my actions for a better life.

Clinical trials and research not only help me but others. The more the medical community learns, the more our treatments will evolve. Research projects are important to help learn more about this disease.

When it’s my time to go, I hope I will leave the PH world a better place or with a cure.

Susan T. Huber

Palm Bay, Florida

Over the past five years, I have overcome and adapted to many health challenges, most recently COVID-19.

I was diagnosed in 2015 with PH. I also was diagnosed with left bundle branch block, a condition that delays or blocks electric impulses that make the heart beat, and heart failure with preserved ejection fraction, which is when the left ventricle of the heart doesn’t properly fill with blood.

I found support and education through the Pulmonary Hypertension Association. I also found a few Facebook groups for PH and heart failure. Through these groups, I learned about studies for my conditions.

I thought it would be a privilege to share my thoughts, opinions and experiences for research. The frustrating reality is that there is no hard and true treatment for me because medication that could help my PH might worsen my heart issues.

However, I have participated in non-treatment related studies, including one about support group participation and another about a new app to track PH symptoms.

I also participated in a study where a sensor was placed under my mattress to track my sleep, heart rate, respiration and daily weight. The hope was that the sensor could identify worsening heart failure before hospitalization was needed. In my case, worsening heart failure would negatively affect my PH.

COVID-19 complications

Most recently I participated in a study for my loss of smell from COVID-19. I had COVID in July 2020 and didn’t feel normal again until November. I had never been so sick in my life. The doctor prescribed two rounds of dexamethasone and two rounds of zythromycin and N-acetyl cysteine capsules, a supplement to build antioxidants.

I had a 102-degree fever for 12 days. It would come down when I took Tylenol, but it always went back up. I had chills, extreme body pain and incredible weakness. Twice, I thought I might be dying, and I didn’t know if I was going to survive.

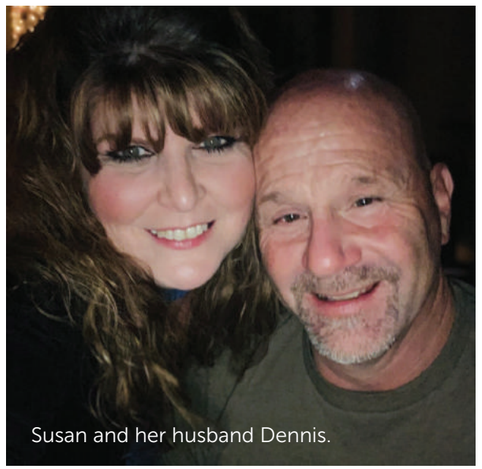
My doctor said I should stay home and rest. If I had started having difficulty breathing, I would have had to go to the hospital. After a televisit with my primary care doctor, another round of dexamethasone relieved some respiratory symptoms.

Although I recovered from COVID-19, I haven’t regained my full sense of smell. Certain things have a distorted smell I don’t like. Soap, detergent, my favorite body spray, olives, white wine and coffee all smell the same.

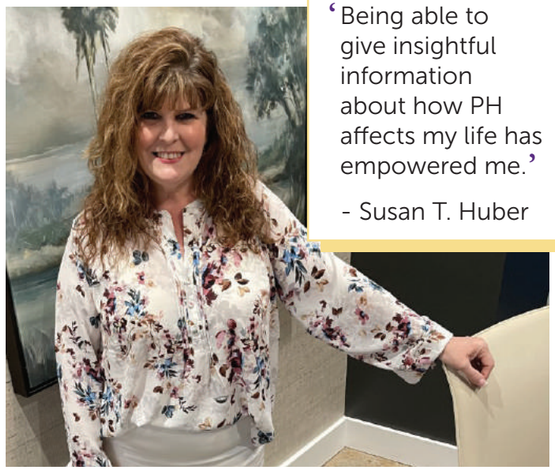
As part of the COVID-19 study, I received three boxes by mail. I had to peel the top off and sniff each one. The first box had the faintest smell, but it reminded me of the distorted odor I don’t like. I smelled nothing in the other boxes. I then visited a website and answered questions about what I smelled.

Why I participate in research

Being able to give insightful information about how PH affects my life has empowered me. I choose to live my best life and not let PH, heart failure or any other illness take away my joy and will to survive.



Susan and her husband Dennis.



> ‘Being able to give insightful information about how PH affects my life has empowered me.’
>
> - Susan T. Huber

PARTICIPATING IN RESEARCH

DURING THE PANDEMIC

By Colleen Brunetti, PHA Board of Trustees Chair

As a longtime beneficiary of medications that began as ideas tested in clinical trials, I've always wanted to give back and participate in a trial. However, the timing and trials available were never quite right.

Last year, I found out about a trial I was really interested in and qualified for. I chased down the participating research centers, and to my initial disappointment, found that none were close to where I live in Connecticut.

I'm not easily deterred, so I decided to extend how far I would travel to make sure I could participate in this thing that was so important to me.

I reached out to Penn Medicine, a Pulmonary Hypertension Association-accredited Pulmonary Hypertension Care Center (PHCC) in Philadelphia, and made my case.

Pre-pandemic logistics

The clinical research coordinator and I made a plan that involved my leaving home in the early morning, traveling by train to Penn for my appointments, and getting home by late evening. Longer appointments would require overnight hotel stays.

We put the plan in action, and I completed two key appointments in February 2020. The researchers switched me from a trusty medication that had supported me for years and started me on a new treatment.

I was among the first PH patients in the world to participate in this trial phase. Testing included a physical exam, exercise tests and extensive blood draws that measured the medication level in my system over the course of a day.

All was going well, and then the pandemic struck.

Managing change

I was on a return train from Philadelphia, going through New York City, when New York's first COVID-19 cases were announced. Suddenly, my carefully laid public transportation plans no longer were an option, and Penn was too far for me to drive round trip by myself in a day.

Thankfully, the trial sponsor and Penn were willing to work with me, first with telehealth visits. As infection rates seemed to improve last summer, I began driving with a friend who was taking the same safety precautions I was.

However, as the third wave hit the East Coast, I couldn't risk even commuting with a friend. I went back to telehealth visits in the fall, which wasn't ideal in the middle of a study.

Weighing risks

Through it all, I worried. I was doing well, but not as well as I wanted to be. I had to increase the dose of the study drug, and I delayed one increase because I worried what would happen if things went wrong.

I didn't want to go to my own beloved PH center because the infection rates were so high in my community. And I didn't want to be on the road, going through rest stops, then into the hospital for the study visits. It was a calculation of risk, and it was

Colleen with Harold Palevsky, MD.

Colleen (left) with Tess LaPatra.

unnerving to be mid-study and mid-pandemic. I stayed in the trial, and I'm really glad I persevered. In March 2021, I mustered my courage and drove back to Penn by myself for a much-needed in-person visit.

At first, I sat in my driveway, ready to leave, almost in tears. I had barely stepped into a grocery store during the pandemic, never mind traveled.

Stashed in my suitcase was a new can of wipes, and I cleaned my own hotel room as soon as I arrived. I double masked. I sanitized everything I touched. And I made it through just fine. Now that I'm fully vaccinated, I feel safer and more confident and look forward to more in-person visits.

Working toward a cure

People often ask what drives me. It's simple. I have hope for a cure, and I'm determined to do whatever I can to help get us there.

While I didn't see improvement, I was starting from a decent baseline. I was happy to maintain and reap the benefit of a medication that was much easier to take than my previous one. To me, trials should be as much about quality of life as they are about controlling disease. I have always sought a balance of the two in my medical care.

I also have experienced severe declines in other medication switches, so I was relieved and happy to stay steady in the trial.

If all goes well, the Food and Drug Administration could approve the treatment. That would mean the PH community would have one more piece of hope to hang on to. And I look forward to that.

Nobody ever said this PH life would be easy, or that clinical trials were a cakewalk. Progress takes work. It takes sacrifice. It takes giving of ourselves and of our resources. Donating to trial research is a wonderful way to participate if you can't be in a study. It's worth it.

THE FUTURE OF PH CARE

The COVID-19 pandemic was the perfect time for Corey Ventetuolo, M.D., M.S., to launch a pilot study to validate an at-home six-minute walk test.

The six-minute walk test is fundamental to pulmonary hypertension (PH) treatment and monitoring and typically an end point for clinical trials, she says. That's why Dr. Ventetuolo and her longtime friend and colleague Steven Kawut, M.D., M.S., wanted to find a way to avoid disrupting ongoing clinical trials during the pandemic while keeping patients safe.

Drs. Ventetuolo and Kawut study wearable technology to improve PH care. Similarly, a team from Vanderbilt University and Johns Hopkins University hospitals are examining how text messages and Fitbits can increase patient step counts.

In winter 2020, Drs. Ventetuolo and Kawut joined their centers, the Rhode Island Hospital Pulmonary Hypertension Center and the University of Pennsylvania Pulmonary Hypertension Program, to determine whether home tests were possible and useful.

People with pulmonary arterial hypertension or chronic thromboembolic PH participated with a friend to record their walks. The friend helped set up markers that adhered to American Thoracic Society standards and reported to the coordinator the number of laps the patient completed and the distance of the final lap. A research coordinator stayed on a call with the patient and friend pairs.

The coordinator calculated the total distance the participant walked and entered the results into a database. The results then were compared to results of six-minute walk tests completed in clinic. Each patient had to complete at least two remote six-minute walk tests, one practice and one official walk.

"If we can prove this test is valid, I think this is one small way to give patients back some control and make them feel empowered about the management of their disease," Dr. Ventetuolo says.

Twenty patients have participated in the study, and the doctors plan to complete the study this summer.

Life outside the clinic

When patients visit their care centers for six-minute walk tests, they might be nervous or uncomfortable, Dr. Ventetuolo says. Since the pandemic, patients have said they felt short of breath while wearing their masks during the test. At-home six-minute walk tests allow patients to do the tests in their everyday environments.

"We owe it to our patients to try to find ways to capture what their lives are like outside of the clinic," Dr. Ventetuolo says. "How they function and feel at home is more important. Part of being a good provider and physician is to address their needs and see how limited they may be outside of the clinic."

'We owe it to our patients to try to find ways to capture what their lives are like outside of the clinic.'

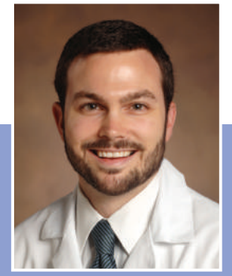
– Dr. Ventetuolo

Develop a wearable

Ultimately, Drs. Ventetuolo and Kawut hope the study will prove the validity of at-home tests so the doctors can develop an app or wearable device that helps patients conduct their own at-home walk tests.

It's important to recognize not all PH patients have access to wearable technology, Dr. Ventetuolo says. If this study proves valid, she wants to see a wearable device accessible to all patients who want one.

Dr. Kawut



Dr. Brittain

Accessible and affordable

In another study using wearable technology, cardiologist Evan Brittain, M.D., MSCI, wanted a practical way to encourage PAH patients to increase physical activity.

With Anna Hemnes, M.D., other colleagues at Vanderbilt University Medical Center, and co-investigators Seth Martin, M.D., and Michael Blaha, M.D., at Johns Hopkins University, Dr. Brittain started a pilot study using text messages and Fitbits to measure patient step counts.

Many insurers don't routinely cover cardiopulmonary rehabilitation, Dr. Brittain says. Even when insurers cover rehab costs, travel to rehab centers can be difficult for patients who live in rural areas. The study offers an accessible, affordable way for patients to stay active.

"Exercise is beneficial for virtually every organ in the body, and it's good for quality of life overall," Dr. Brittain says, noting that exercise has been proven to be beneficial for most people with PAH. "The challenge is getting people to exercise consistently, which is a behavioral change. We wanted to see if we could tip the scale in terms of these behavioral choices."

Personalized messages

The research team gave 42 patients Fitbits to track their step counts. The patients shared information about their lifestyles, including activities they liked, whether they lived near parks, had pets, etc.

The investigators developed personalized messages based on patient step count with a behavioral psychologist. They then randomly separated the patients into two groups. One group received motivational text messages. The other group received no messages. Patients in the text group received three automated texts a day with words of support and encouragement to stay active.

Everyone had individual step goals based on the number of steps they walked during a two-week prep period. Patients wore Fitbits during those two weeks, and researchers collected data from the second week to measure patients' average step counts. The goal increased 20% every four weeks during the 12-week study.

Fitabase, a Fitbit data program designed for health research, allowed Dr. Brittain to see patient's steps, heartrate and their Fitbit charge level in real time. If a patient didn't appear active one day, the program sent an automated text to make sure everything was working properly.

Quality of life

In April 2020, the study's results showed that patients who received personalized text messages increased their daily steps and averaged 1,300 more steps in week 12 than they did the first week. The second group's steps didn't change.

Ultimately, Dr. Brittain would like the study to develop into a service or device patients and physicians can use to improve quality of life.

In the meantime, he is working to expand the study nationwide and increase the length of the study to six months. He received approval to source participants from the Pulmonary Hypertension Association Registry.

"Our main goal is to improve quality of life," Dr. Brittain says. "That's what we really want to change."

INNOVATION FOR AN INDEPENDENT LIFE

Device Detects Low Oxygen

Software developer and entrepreneur Shavini Fernando invented a wearable device to continuously monitor oxygen saturation to help herself and other people with pulmonary hypertension (PH). Shavini, originally from Sri Lanka, created her company, OxiWear, while pursuing a Master of Arts in communication, culture and technology at Georgetown University. In addition to a bachelor's degree in computer science and a Master of Business Administration, Shavini is completing an executive certificate in innovation and entrepreneurship through Stanford University.

By Shavini Fernando

For as long as I can I remember, I was always wheezing. Monthly midnight nebulizer treatments were routine.

I loved playing sports, but I couldn't run beyond 100 meters. Swimming beyond three lengths of the pool caused me to gasp for breath. Despite winning medals, I couldn't compete beyond college-level sports because it was too risky to my health. I wanted to do so much more, but my sickness limited me from going further.

In 2006, after completing my undergraduate degree in the United Kingdom, I left for Sydney, Australia, for my graduate studies. That's when my real problems started.

I had difficulty breathing and found it hard to climb stairs and walk uphill. Running to the bus became a problem; I had to leave home 30 minutes earlier to catch it.

Using my staff ID, I took elevators to reach the upper campus. I couldn't swim even two lengths anymore. My life became more limited. An x-ray and MRI showed scarring of lungs, and I was diagnosed with pneumonia. I believe it was the starting point of my pulmonary arterial hypertension (PAH).

Emergency episode

I went back to Sri Lanka in 2009 to work as a lecturer. My breathing got worse. On flat surfaces, I could walk fast, but uphill and on steps, I was gasping for breath.

In 2010, due to an elevator malfunction, I had to run to the third floor. As I started class, I suddenly couldn't breathe, everything went blurry and I couldn't feel my pulse.

My students started to scream, "Ma'am are you alright? You are going blue." I gulped a bottle of water but still couldn't breathe so I started to hit my chest until I could catch my breath.

At the emergency room, I explained what happened. The doctors attributed the episode to palpitations from gastritis and prescribed antacids.

Similar episodes didn't stop me from living my life. I finished my MBA, traveled to multiple countries and did everything I loved.

Grim prognosis

In 2015, while I was trekking with a friend, I had another episode. My friend, an experienced pilot, recognized my low oxygen and said I should visit a cardiologist.

I consulted a cardiologist, who identified an atrial septal defect. He was surprised that it was never diagnosed and referred me for a right heart catheterization. During the procedure, I suffered a cardiac arrest and had to be revived.

When I woke up in the ICU, I was told that I had severe PAH due to Eisenmenger's syndrome, a blood circulation abnormality caused by my heart defect. No treatments were available, and the doctor said I had two years to live and enjoy my life. But I wasn't ready to accept that prognosis.

> 'Similar episodes didn't stop me from living my life. I finished my MBA, traveled to multiple countries and did everything I loved.'

I flew to the United States for a second opinion from John Hopkins University Medical Center in Baltimore. I ended up in the ICU for four weeks because of a stroke, and I was diagnosed with severe PH, a barely working left heart, a severely enlarged right heart and a shunt.

The doctors said that I might need a transplant but they would decide after checking my progress on vasodilators and 24/7 continuous oxygen.

From left: Charuni (sister), Shavini, Ramani (mother), Sarath (father) and Kushalya Fernando (brother).

Regaining health

Within three months, I started to feel a difference. I changed from continuous oxygen to a pulse concentrator and started more physical activities to increase my lung capacity.

During a six-minute walk test in the ICU, I could walk only 500 feet. But three months later, I could walk 1,300 feet. My results kept improving. Six years later, I go rock climbing with my friends without oxygen, and the doctors believe I have five to 10 years before I'll need a transplant.

With everything improving, I moved to a studio apartment in 2017 and started my master's degree at Georgetown. To help create awareness for PAH, I made a documentary for the Pulmonary Hypertension Association (PHA) and participated in PH fundraisers and events.

But during my summer internship, history repeated. I couldn't breathe, and my heart stopped. The incident raised the question of whether I could safely live alone and study at Georgetown, which has hills and many steps. I didn't want to go back to zero. I just started my life again.

Shavini demonatres her ear-worn oxygen monitor.

Becoming an entrepreneur

I started to think about ways to live independently and came up with an idea for a wearable device. I discussed my OxiWear idea with my physician at Johns Hopkins University Hospital. After realizing the number of people I could help, I developed the device as an independent study.

OxiWear, worn over the ear, continuously monitors my oxygen saturation and warns me when my oxygen levels fall below the safe threshold. It also sends alerts with my location to my emergency contacts to ensure I receive immediate help.

'I started to think about ways to live independently and came up with an idea for a wearable device ... After realizing the number of people I could help, I developed the device as an independent study.'

The device began to receive attention, earning technology and entrepreneurship awards and grants. The awards allowed me to form a company, patent the device and attract more investors.

Next steps

My company is conducting tests to ensure accurate readings for people with dark skin or who have skin conditions. Skin pigmentation is among the factors that can affect the accuracy of pulse oximeter readings.

We plan to release OxiWear in December as a fitness device while we work toward FDA approval as a medical device.

As a PAH patient who's gone multiple life-threatening events, I believe OxiWear can warn patients of dangerously low oxygen levels before they end up in an emergency room. Ultimately, I think it also can reduce medical bills and put a stop to preventable deaths from silent hypoxia.

PHPN SYMPOSIUM

Health Care Professionals

Gear Up for Symposium

Registration is open for the Pulmonary Hypertension Association's biennial event for health care professionals. Set for Sept. 30 through Oct. 3, the Pulmonary Hypertension Professional Network (PHPN) Symposium provides invaluable information about advancements in PH research and care.

This year's PHPN Symposium, "PHiguring Out the Puzzle: Pieces to Innovative, Patient-Centered Care," will be virtual for the first time because of the COVID-19 pandemic. The event is expected to attract about 400 PH-treating health care professionals for continuing education, networking, fundraising and advocacy.

Popular features, including Advocacy Day and Trivia Night, will continue in digital formats. Trivia Night, set for Oct. 1, will raise money for patient scholarships to PHA 2022 International PH Conference and Scientific Sessions.

Keynote sessions

- **PH Through the Decades: Progress and Patient-Centered Care**, presented by Bob Frantz, M.D.
- **State of the Union in PH**, presented by Vallerie McLaughlin, M.D., and Victor Tapson, M.D.
- **Navigating Difficult Discussions in Pulmonary Hypertension**, presented by Rana Awdish, M.D.
- **Risk Assessment and the Importance of Achieving Low Risk Status** by Raymond Benza, M.D.
- **Recent Findings from the PVDOMICS Study** presented by Anna Hemnes, M.D.

Don't miss

- **Pediatric programming.** Pediatric topics will be scheduled for every breakout session during the conference. Programs can be applied to pediatric and adult patients.
- **Accredited education sessions.** Attendees can earn up to 12.5 hours of CE credit for attending sessions and lightning-round poster presentations during the virtual event, as well as visiting abstract authors in the virtual visit hall. Attendees also can earn up to 12 hours afterward through on-demand recordings.

Health care professionals: Take advantage of early-bird registration discounts until July 7 at PHAssociation.org/Symposium. All active members of PHA's medical membership networks – PHPN and PH Clinicians and Researchers – receive discounted registration.

VIRTUAL ADVOCACY DAY VISITS

REMAIN VITAL

Remind your pulmonary hypertension (PH) care team or your health care colleagues that Advocacy Day is a step toward improving life for the entire PH community.

Advocacy Day is a vital, powerful element of the PH Professional Network Symposium.

Symposium, set for Sept. 30 through Oct. 3, shifts to a virtual format this year because of ongoing COVID-19 concerns. Advocacy Day, which will kick off Symposium Thursday, Sept. 30, is free to registered Symposium attendees, including nurses, physicians, social workers and respiratory therapists.

This year, Advocacy Day participants will meet virtually with congressional staffers and/or lawmakers to educate them about PH. They also will urge Congress to take specific actions to improve life expectancy and quality of life for people with PH.

Personal stories from the pulmonary hypertension (PH) community are the most important way you can convince members of Congress to support PH-related legislation. Even in a virtual format, face-to-face visits are the most powerful ways to connect with lawmakers.

No prior advocacy experience is necessary. Advocacy Day provides comprehensive training and opportunities to make virtual visits with other attendees. PHA employees or experienced PH advocates will facilitate the calls.

During previous Advocacy Days, members of Congress pledged support for PHA priority legislation because of stories health care professionals shared.

For example, after Sen. Kyrsten Sinema, D-Arizona, co-sponsored the Safe Step Act of 2019, a member of her staff wrote to specifically thank the PH Professional Network advocates who had visited her office.

"We were happy to [co-sponsor the Safe Step Act], and please know that the group's advocacy for the bill made all the difference," the staffer wrote. "Our internal approval process takes a while – but that discussion with them was SO key."

For questions specifically about Advocacy Day, contact Katie Kroner at Advocacy@PHAssociation.org or 301-565-3004 x749.

Research Room Energy

Fuels Investigator's Work

Micheala Aldred (right), Ph.D., is among the researchers who in 2000 identified a gene for pulmonary arterial hypertension (PAH) that runs in families. At the time, she ran a genetics diagnostic laboratory at the University of Leicester, England. Today, Dr. Aldred works at the Indiana University School of Medicine, where she continues to research pulmonary hypertension (PH).

By Micheala Aldred

I first heard of PH over 20 years ago. A young lady came into the clinic because she had a family history of pulmonary hypertension, including a sister who was recently diagnosed, and she was worried about her own risk. At the time, we knew nothing about the genes that can cause PH, but her visit sparked the start of our research.

Our genetics specialist collected blood samples from members of the woman's family, as well as other families in the United Kingdom. My role was to help analyze the samples. Later, we teamed up with groups from Vanderbilt University, Indiana University and the University of Michigan, who were analyzing samples from families in the United States.

Our collaboration led to the discovery of genetic changes in the gene called bone morphogenetic protein receptor 2, or BMPR2. Since then, many other genes have been associated with PH, but BMPR2 is still the main culprit.

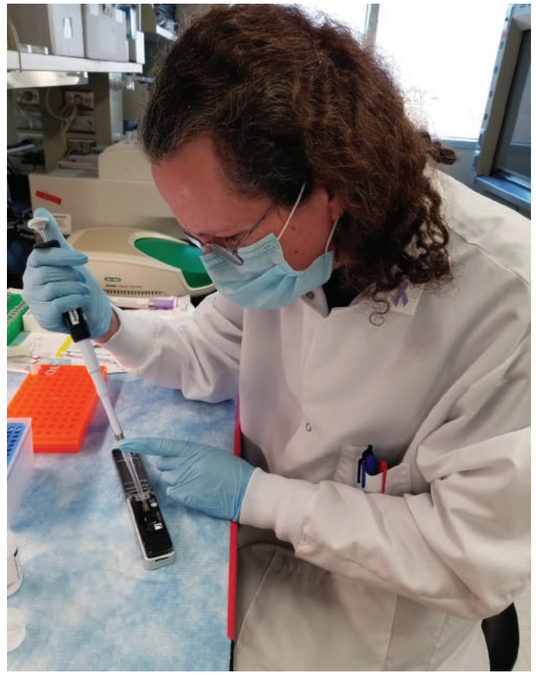
I often joke that I did not choose to specialize in PH research – it chose me. I continued to work on PH for several years before moving to the United States in 2006.

Initially, I focused on other areas of genetic research. However, when I began working at the Cleveland Clinic, the physicians there were keen to incorporate my knowledge of PH genetics into their studies. (Cleveland Clinic is a Pulmonary Hypertension Association-accredited Pulmonary Hypertension Care Center.)

I started to collaborate with them, and eventually PH genetics became my major research focus. Through my work at Indiana University, I am privileged to work with PH physicians and researchers throughout the United States and the world.

I learned of PHA a year or two after moving to Cleveland. I was asked to speak at a Cleveland-area PHA support group meeting about the genetics of

'I've often wished I could take the atmosphere of hope and cooperation from the Research Room, put it in a bottle and take it back to the members of my lab.'



PH. I'd never done anything like that before, so it was a bit daunting. But I really enjoyed meeting everyone, and I guess they liked my talk, because they invited me back for an encore later that year.

After that, I attended PHA's International PH Conference and Scientific Sessions in Orange County, California, in 2010, and I have been a regular participant in the Research Room at every Conference since.

I'm not a physician, so Conference is the only time I get to meet patients and their families. It's one of the drivers that keeps me going when experiments fail, or grants are rejected. I've often wished I could take the atmosphere of hope and cooperation from the Research Room, put it in a bottle and take it back to the members of my lab.

I've also participated in Q&A sessions at Conference. In the past year, I joined a PHA Registry committee, which has given me new insight into clinical studies.

My involvement with PHA led me to become an annual donor. I understand the importance of patient support groups and the services PHA offers.

And I see how PHA directly supports PH research, such as "Redefining Pulmonary Hypertension through Pulmonary Vascular Disease Phenomics." The multi-center study funded by PHA and the National Institutes of Health performs detailed clinical and lab studies to better understand current PH classifications. The PHA Registry, which collects data from patients treated at PHCCs, is equally critical to PH research.

When I look toward the future of PH research and treatment, I am excited about the strides we are making. As we understand more about the genetic and cellular changes that lead to PH, we have a better chance of trying to correct those changes in more specific ways.

This concept, known as precision medicine, has led to significant advances in cancer therapy, where treatment can be targeted to a specific genetic change in a tumor.

We are making great progress and starting to see our PH research translate into new therapies. I believe PHA is instrumental in helping us achieve these goals.

To be honest, working in the PH space is quite humbling, because I'm just a small part of a giant jigsaw puzzle. But it's also exciting. When I started studying genetics at age 19, I never imagined that I would be among those making important discoveries in PH. It's amazing how fast things have changed.

Help continue PHA's critical work to provide patient support and services and fund PH research. Visit PHAssociation.org/donate.

COVID-19 SHAPES PH CARE

HEALTH MATTERS

Long-standing systemic health and social inequities pose greater risks for people with pulmonary arterial hypertension (PAH) to contract COVID-19 complications, new research suggests.

An article in *Advances in Pulmonary Hypertension* addresses how the pandemic has exacerbated health disparities that affect people with PAH, including racism and discrimination; environmental, societal and economic factors; and access to health care.

The article, "Health Disparities in Pulmonary Arterial Hypertension and the Impact of the COVID-19 Pandemic" by Jeanna T. Ryan, M.S., MPAS, MSCIS, PA-C, RDN, of the University of Utah, appeared in the January 2021 issue of *Advances in Pulmonary Hypertension*, PHA's quarterly scientific journal.

The issue addressed "PH in the Time of COVID-19." Other articles in the issue addressed how the pandemic has affected PH research and telehealth.

In the disparities article, Ryan and her co-authors noted that limited access to recreational areas, bike lanes, walkable sidewalks and healthful foods affect patients' ability maintain healthy lifestyles. The pandemic's economic challenges compound the health disparities, the article points out.

Additionally, increasingly complex PAH therapies require more customized treatment plans and frequent interaction with PH health care providers. But the transition to telehealth visits during the pandemic poses new challenges to people with limited technology or internet access.

Those challenges can delay diagnosis and or the introduction of new therapies, creating the potential for further health disparities. Pandemic-related stress also makes it difficult to cope with existing challenges, further increasing risk of poor health, the article notes.

The authors called for more research into how the pandemic has intensified health disparities and urges health professionals from various disciplines work together to find solutions to prevent and end inequities.

"Overcoming environmental and socioeconomic barriers requires creative solutions and shared decision-making to identify realistic personalized interventions that increase the likelihood of patients' ability to adhere to medical recommendations," Jeanna Ryan writes.

Clinical trials

In "Pulmonary Hypertension Clinical Trials and COVID-19," guest editor John J. Ryan, M.D., M.B., B.Ch., B.A.O. of the University of Utah and Roham Zamanian, M.D., of Stanford University discuss how COVID–19 has affected PH research, including clinical trials and data collection.

For example, the pandemic created problems for Stanford's program to collect exhaled breath condensate from people with PH. Concerns about how to safely collect such samples and patient hesitancy limited the program's ability to collect those samples.

Another challenge arose for Stanford's National Institutes of Health-funded trials that depend on six-minute walk tests. Pandemic restrictions prevented patients from coming to the clinic for just a research visit, preventing researchers from collecting data.

The doctors also address how remote meeting software such as Zoom, electronic patient consent forms and remote site initiation for clinical studies helped address pandemic-related challenges such as limited in-person visits.

Other benefits include learning to conduct clinical research in a real-world environment, Dr. Zamanian says.

"Can we take six-minute walk testing to the patient's home, and can we collect registry information using telehealth approaches? I think it's a challenge, but it's rising up to that challenge and overcoming some of the limitations. That's exciting, the opportunities that we can create for the future of research."

Telehealth

Telehealth was an underlying theme in the January issue of *Advances in PH*. In the PH Roundtable article, Dr. John Ryan talks with pulmonologist Mark Avdalovic, M.D., of the University of California-Davis Health and Jennalyn Mayeux, D.N.P., of the University of Utah about how the pandemic has affected patient care.

Before the pandemic, UC-Davis Health routinely offered telehealth visits because it covers a wide geographic area, with some patients coming from as far as Arizona and Oregon. So the institution was prepared early on for additional telehealth visits.

"We knew when the pandemic first hit that we had to ... be careful about who should really come and physically see us," Dr. Avdalovic says in the discussion transcript.

Like many institutions, UC-Davis Health weighed the risks of contracting COVID-19 to determine whether patients should be treated in-person or remotely. Those at high-risk who needed routine follow-ups generally were steered toward telehealth.

Similarly, telehealth has helped alleviate the challenges of treating patients from the five states covered by the institution, Dr. Mayeux says. Early on, her patients had many COVID-19-related questions. Telehealth helped them connect with their care teams, and understand they would still receive care and their concerns would be addressed.

Drs. Avdalovic and Mayeux have faced common challenges related to limits on in-clinic PH diagnostic testing. They discussed how they adapted their approaches so they could evaluate patients. Dr. Mayeux said her team partnered with hospitals and providers in patients' home communities to provide care and keep patients safe from COVID-19.

Pediatric PH

Elizabeth Colglazier, M.S., CPNP-AC, of the University of California-San Francisco and Anna Brown, D.N.P., CPNP, of Vanderbilt University Medical Center also discuss testing limitations and technology in "The Benefits and Challenges in Delivering Telehealth in Pediatric Pulmonary Hypertension."

They review various telehealth platforms, including Zoom for Healthcare, Doximity, GoToMeeting, Cisco Webex Meetings and Amazon Chime. They also outline the process for conducting telehealth visits.

Colglazier and Brown touched on future telehealth opportunities, such as educating patients, families and caregivers to understand and manage PH and transitioning from pediatric to adult care. They also say telehealth provides opportunities for social support.

"Patients, siblings, parents and caregivers all experience the physical and mental challenges from living with or caring for a child with PH," they write. "Telehealth offers a platform to obtain support from providers and peers. For example, support groups may be offered by social workers or clinical psychologists over telehealth for patients, siblings, and caregivers. The barriers that might restrict support group attendance, such as time and distance, may be overcome when telehealth modalities are used."

Going forward, PHA will continue to advocate for the PH community and work towards our mission of improving the lives of people living with PH. PHA recently formed a COVID-19 Taskforce of clinicians, allied health providers, patients and caregivers to share knowledge about COVID-19, and understand and address pandemic-related challenges.

PHA Registry Enrollment **EXPANDS**

Researchers have better access to data on patients with pulmonary hypertension (PH), thanks to growing participation in the Pulmonary Hypertension Association Registry (PHAR).

PHAR enrollment recently surpassed a new milestone with more than 1,600 patients. PHAR collects data from people with pulmonary arterial hypertension (PAH), pediatric PH due to developmental lung disease and chronic thromboembolic pulmonary hypertension (CTEPH). PHAR data includes demographic characteristics, diagnosis and treatment information, and quality-of-care metrics.

Researchers use the PHAR database to learn more about PH. PHAR is the largest, active PH patient registry in the United States, with data collected at 59 PHA-accredited Pulmonary Hypertension Care Centers (PHCC) throughout the country.

Each patient entry in the database potentially could lead to new treatments, earlier diagnosis or ultimately a cure.

"Each patient is unique, and it is important to capture their specific data," says Gloria Hamm, R.N., B.S.N., MSHA, who represents patients on PHA's PHAR steering committee. She was diagnosed with PAH in 2007.

Why participate?

Participation can help health care professionals better understand the factors that affect patient health, such as geography, differences among PH care center practices and socioeconomic limitations, says PHAR steering committee member Elise Whalen, M.S.N., APRN, FNP-C, of Texas Children's Hospital, a PH Care Center.

Patients can help improve care for themselves and others with PH because health care professionals can use the data to learn best care practices from each other. Researchers use the data to study disease patterns, which potentially could lead to funding for new PH studies.

How PHAR helps improve care

PHAR delivers information to PH care teams that can help them learn more about their PHCCs and PH care.

Plus, patient surveys can open conversations between patients and providers about how health-related quality of life and similar topics.

"The PHAR was developed so each PH center can learn more about its strengths and weaknesses, over time leading to quality improvement that delivers better care to each person with PH," says steering committee chair Daniel Grinnan, M.D., of Virginia Commonwealth University School of Medicine. "It was also developed as a research tool, so that questions answered by each participant would help the PH community to learn more about PH and improve care and outcomes."

How researchers benefit

PHAR makes data available to scientists and doctors who have specific research questions about PH. Interested investigators submit their questions to PHCC Inc., the organization that manages PHAR, and explanations of how they plan to use PHAR data to answer their research questions.

When approved, PHA provides identity-protected data limited to the topic, with the expectation that investigators will publish their findings to broaden understanding of the disease.

In the past year, PHAR data has contributed to more than 30 research proposals and several published manuscripts.

"We are seeing results and changing how we perceive PH," Dr. Grinnan says. "[PHAR} is a well-established registry with tremendous potential. Each participant gives a very important boost toward helping the PHCC, medical research, and the care of PH patients."

If you have PAH, CTEPH or pediatric PH due to developmental lung disease and are starting evaluation and/or treatment at a PHCC, ask your care team about participating in PHAR. Your participation can help improve understanding of PH.

TRANSFORMATION in Telehealth

Michael Mathier, M.D., used telehealth to see some of his pulmonary hypertension (PH) patients before the COVID-19 pandemic, but now, even more patients are scheduling telehealth visits.

Expanding options to see patients has been critical for Dr. Mathier and his team throughout the pandemic as they strive to limit gathering in their clinical spaces and promote social distancing. For many people with PH and their health care providers, telehealth services have become more common, and in many cases, the new normal for getting medical care.

At the beginning of the pandemic, telehealth visits rapidly increased at Pulmonary Hypertension Association (PHA)-Accredited PH Care Centers, according to a study published in the July 29, 2020, *Annals of the American Thoracic Society*.

After the study, PHA informally polled PH health care providers about whether they still used telehealth visits six month into the pandemic, as some in-person doctor visits were resuming. More than 90% of survey respondents said they continued to use telehealth visits as a positive way to stay connected with their patients.

Patient benefits

Improved access to care. Telehealth can help people who live far from PH care centers access their doctors. Telehealth expands the geographic reach, offers flexible options for getting care and allows patients to connect with expert care without travel. Those advantages are especially beneficial for people who live in rural or isolated areas where regular trips to the doctor can be challenging and time consuming. Telehealth visits save time as well as travel costs, Dr. Mathier says.

Finding new providers. Telehealth may be an option for initial meetings with new doctors or specialists who might be hundreds of miles away.

Accommodates busy schedules. A telehealth appointment might be easier to schedule than an in-person visit – for patients and providers.

Enhanced communications. Various telehealth platforms allow patients to share information with PH care teams about health changes, ask questions about medications or get support from those who know your PH best.

"They allow for more frequent contact with our patients, which is very beneficial in PH," Dr. Mathier says.

Managing PH during the pandemic. Using telehealth during the pandemic can prevent patients and care teams from contracting or spreading COVID-19.

What's ahead

While the benefits of telehealth are clear, some challenges exist. "We can hear our patients, and we can see our patients, but we can't fully understand their progress without being with them in person," says Deborah Levine, M.D., director of pulmonary hypertension at the University of Texas Health Science Center in San Antonio.

Beyond the pandemic into the new "normal," it will be important for patients to work with their health care team to find a balance that combines telehealth with in-person doctor visits. "These types of visits are here to stay," Dr. Mathier says.

PHA Registry Adapts to Pandemic

Patients who agreed to share data with the Pulmonary Hypertension Association Registry (PHAR) before the pandemic had to sign consent forms and complete study documents in person at clinic visits.

As telehealth visits increased during the pandemic, PHAR adopted remote patient-consent and data-entry procedures. The remote procedures allow people with pulmonary hypertension (PH) to continue participating in the registry, which helps advance science and improve PH care.

"These remote data collection options enable the registry to continue accruing critical data ... to improve quality outcomes for people living with PH," says Kristina Blank, project director for the PHAR Data and Clinical Coordinating Center.

Rare PH and Liver Transplant
Inspire Volunteer Service

About a year and a half after a liver transplant, Kim Crowell wanted to give back after receiving the gift of life. Today, she is a Pulmonary Hypertension Association (PHA) support group leader and moderates Facebook groups related to pulmonary hypertension, liver disease and organ transplant. She and her husband Andy and their three Labrador retrievers live on a 45-acre non-working farm in Dothan, Alabama. Retired from her career in IT consulting, Kim spends her free time doing yoga, tai chi and strength training.

By Kim Crowell

After several years of searching for answers to my health problems, I was diagnosed in 2017 with a rare form of PH called portopulmonary hypertension (PoPH). It involves the heart, lungs, and liver. My specialists believe that a condition related to my liver, portal hypertension, caused me to develop pulmonary arterial hypertension (PAH).

When I was diagnosed, my liver was end-stage, and I needed a liver transplant. However, my mean lung pressure of 55 was too high for me to be placed on the transplant list. My PH specialist recommended that we pursue an aggressive treatment plan to lower my pressure in hopes of getting me listed.

At that point, my specialists thought my odds of making it to transplant were 50/50 at best. After a lot of discussion with my specialists and my husband, I decided to pursue the treatment. I started taking Veletri, an IV medication targeting the prostacyclin pathway, as well as Adcirca/Tadalafil to relax and widen the vessels in my lungs.

In August 2017, a right heart catheterization showed my pressure had dropped to 34. That was just low enough to place me on the liver transplant list. I received my transplant about three months later.

Recovery process

I was very sick going into my transplant, but I made it through the surgery. My hepatologist (liver doctor) had told me it would take me two years to fully recover because of my condition.

After surgery, I continued Veletri for about four months. When a right heart cath showed that my PH pressure had dropped to 21, I was taken off Veletri and transitioned to Uptravi for a few months. After a few months with no issues, I was taken off Uptravi. At the time, I was taking only Adcirca for my PAH.

For the next year and a half, I spent most of my time recovering. With time, I got better with only minor complications related to the anti-rejection medication for my liver. I began exercising and pushing myself physically.

In October 2019, an echocardiogram and six-minute walk test showed that my heart was functioning normally. At that point, I was taken off all PH medication.

> 'I find the most inspiration from the selfless 36-year-old woman who was my organ donor. I try to live my life in a manner that would make her proud of me every day.'

In terms of my liver, all of my labs have been great with no signs of rejection. Now, I take one medication for anti-rejection, so my side effects are minimal. I feel great!

Moving forward

Eighteen-months post-transplant, I began looking for ways to give back for the gift of life I received. I now volunteer for PHA, the American Liver Foundation, Donate Life and Global Liver Institute.

I moderate a large Facebook group for liver patients and two groups for PH. I recently started my own Facebook group for organ transplant recipients because a transplant changes your life in many ways.

I believe transplant patients, much like PH patients, have a common bond that comes with many challenges. I hope my new group will provide a safe and caring environment where transplant patients can post freely and openly about issues they have.

My PH specialist, Dr. Karen Fagan, M.D., told me about PHA at my initial diagnosis. I began using PHAssociation.org to help me understand everything that was happening to me.

One of my goals post-transplant was to learn everything I could about PoPH in hopes that I could help others who have it or might not even know they have it. Eventually, I became more and more involved with PHA because I believe they supply the best information and support for PH patients. I wanted to be part of that.

Providing support

For me, the most rewarding thing about volunteering is helping people who are traveling this journey with PH and/or liver disease. Both are terrible diseases and are difficult roads to travel physically and mentally.

I want to be there for anyone who needs help or support. I also want to provide hope and encouragement for those who have these diseases. I am living proof that you can fight this disease and go on to lead a normal life. I always tell people to find their inner warrior and gear up for battle. It can be done.

I am proud that I took the time to learn about PH and liver disease. I love to share my knowledge with newly diagnosed patients who are struggling to understand it.

I find the most inspiration from the selfless 36-year-old woman who was my organ donor. I try to live my life in a manner that would make her proud of me every day. Without her, I would not be alive today, so I am living my second chance at life with every ounce of my being.

Thanks to Janssen Pharmaceuticals, United Therapeutics Corporation and Bayer Healthcare for sponsoring PHA's peer support program.

QUICKTAKES

Federal Insurance Exchange Still Open

You can still apply for health insurance coverage through the federal marketplace and some state exchanges. A special enrollment period related to the COVID-19 pandemic is open through Aug. 15. Changes and new enrollments are permitted.

Even if you weren't previously eligible for coverage, you might qualify under pandemic-related rules.

Visit healthcare.gov to enroll, change coverage or learn if you qualify for Medicaid or the Children's Health Insurance Program. Questions? Contact PHA's Treatment Access Program at Insurance@PHAssociation.org or 301-565-3004 x749.

Ask your members of Congress to increase National Institutes of Health (NIH) funding in the federal budget. NIH provides grants to advance researchers' work to understand and treat pulmonary hypertension (PH).

Visit the Pulmonary Hypertension Association's Advocacy Action Center to keep legislators informed about the challenges of living with PH and ask them to help.

Here's how it works:

- Enter your street address and zip code to route your message to your members of Congress.
- Personalize your message.
- Hit send.

To get involved in PHA's grassroots advocacy network, contact Advocacy@PHAssociation.org, or call 301-565-3004 x749.

Insurance Find: Navigate the SSDI Application Process

Learn more about Social Security Disability Insurance (SSDI). The cash benefit helps cover expenses for people who no longer can work because of a medical disability.

The application process is lengthy, so start sooner than later. The Pulmonary Hypertension Association's online insurance guide provides an overview of the SSDI application process, tips for building your case and information about the appeals process.

Visit PHAssociation.org/patients/insurance-and-treatment-access/social-security-disability or call us at 301-565-3004 x749.

Study: PH Support Groups Are Good for Your Health

Your support group participation might improve your life more than you realize.

A recently published study shows that support groups improve meaningful health-related outcomes, such as self-care, pulmonary hypertension (PH) symptom management, medication adherence and understanding related medical procedures.

The study, published in the April 2021 issue of the journal Pulmonary Circulation, examined the effects of support group participation on people with PH and their caregivers. The study is thought to be the first to show that PH support group participation is associated with improved outcomes.

Learn more at PHAssociation.org/phassociation.org/pha-news-home.

Help Support Our Mission Through Fundraising Events

Support Pulmonary Hypertension (PHA) fundraising events, which help us provide critical support and services for the pulmonary hypertension community. Get information about upcoming events at PHAssociation.org/pha-fundraising-events. Find out how to host an event at Events@PHAssociation.org.

PHA thanks its national fundraising event sponsors for their generous support:

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FDA Clears Generic Subcutaneous PH Treatment

A generic form of Remodulin is available for to treat pulmonary arterial hypertension (PAH) subcutaneously, thanks to a new medication cartridge.

The new cartridge became available May 21. The U.S. Food and Drug Administration approved generic treprostinil injection for intravenous and subcutaneous PAH treatment in 2019. However, manufacturer restrictions on medication pump cartridges limited the generic's use to only intravenous delivery.

The injection contains the same active ingredient, strength, dosage form and inactive ingredients as Remodulin, as required by the FDA. Both the intravenous and subcutaneous versions are available from the same specialty pharmacy that dispenses the brand name medicine.

Learn about financial assistance for PAH therapies visit at PHAssociation.org/HELP or call 301-565-3004 x758.

PHA's Legacy of Hope
To honor those who have included PHA in their estate plans or whose legacies have been realized, PHA created the *Legacy of Hope Society*. PHA is pleased to recognize the following members:

Laura and Rino Aldrighetti*
*Alice A. Arnott**
*Sandra A. Awood**
*Dauna L. Bauer**
*Sylvia M. Becherer**
*Joan F. Bennett-Schenecker**
Kris L. Best
*Gloria G. Blodgett**
*Dorothy E. Bradley**
*Mary M. Brady**
R. J. Braun
Roberta F. Browning and Lee A. Broadbent*
Rita and Bruce Brundage
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James Kenney
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*Terri L. Kopp**
Debbie L. and Mitchell Koppelman
Dee and Walter Kruger**
*Gloria J. Lang**
Marie and Ronald J. Levendoski
Mary Jo and Thomas Linnen**
*Sally Maddox**
Bonnie and Michael D. McGoon
*Joseph W. Mihuc**
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Larry D. Moody
Pamela R. Morris
*Marjorie D. Mott**
*Joyce L. Mowrer**
Dorothy M. and Harry J. Olson**
*Theresa "Terry" E. (Cavanaugh) O'Reilly**
Rita and Guy Orth
Patricia R. and Gerald D. Paton**
Cynthia and John R. Pickles
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Judith and Edwin L. Simpson
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*Helena M. Strauch**
Douglas R. Taylor
*Frank A. Tobac**
Martha and Carlos Torres
Deborah J. and Roger K. Towle
Carol B. Ungar
*Daniel R. Walsh**
Andrea and Stephen L. White

** deceased members*

Passages

Since the Pulmonary Hypertension Association (PHA) began publishing *Pathlight*, "Passages" has provided a place to memorialize people with pulmonary hypertension who pass away. PHA extends its sympathy to the families and friends of these individuals and rededicates itself to its mission in their memory.

Patricia A. Adcock
Sandra L. Affholter
Ada M. Baranello Gonzalez
Joy A. Barker
Susan I. Benjamin
Audrey Besserman
Debra Boyd-Moore
Helen Bradford
Esther M. Brown
Linda Chambers
Michael R. Chase
Ray Churchill
Sue V. Cohen
Ardith Coleman
Lucinda C. Conley
Jean M. Cook
Margaret P. Corrigan
Mary A. Cremin
Paula Cull
Diane Davis
Kevin Deeb
Marpha Ditilato
Thomas Eames
Renee S. Elder
Paul Elkins
Charles W. Fall
Byron L. Farmer
Helen Frazier
Anne Freeland
Gary L. Freeze
Carmen S. Froseth
Susan Garner
Jeffrey J. Glen
Cheryl Gonzalez
Elsie Gray
Heidi L. Griffin
Richard S. Guinan
Helena Gupton
Donna Gyde
Kelly Hawley
Ruth A. Heineman
Reid D. Hodges
Susan Hoffmann
Debbi Howell
Eileen Hunter
Barbara L. Jacquette
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Marlene J. Johnson
Belinda Jones
Grace I. Leonard
Kelly Like
Ashley J. Macaluso
Helen V. McCarney
Janice C. McGuigan
Claire P. Montminy
Wayne Moreland
Robert E. Morrill
Kathleen Novak
Stephanie Oliveira-Mello
Nancy B. O'Neal
Ramona Overton
Michelle Patrick
Kent F. Petersen
Angie Petro
Peter G. Pierce
Elizabeth Quetschenbach
Jenny Reinertson
Patricia Rider
Nicole Ridgeway
Victoria L. Roark
Juliette Robinson
Sherry Rouse
Carole Sadowsky
Ronald Santti
Diane R. Schwartz
Darlene M. Sebastian
Arlene Stuart
Barbara Tanen
Susan V. Temple
Sandra Tramer
Jerrelyn W. Verrett
Rebecca D. Watson
Jo Ann Wells

The accuracy of this list is important to us. Please contact the PHA office at 301-565-3004 x746 or Passages@PHAssociation.org to share the name of your recently deceased loved one or report an error or omission.

Your donations in memory of others, in honor of others and in support of our mission mean so much to the entire PH community, and we thank you.

Pulmonary Hypertension Association
8401 Colesville Road, Suite 200
Silver Spring, MD 20910

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