Navigating a pulmonary arterial hypertension (PAH) diagnosis can be challenging. That's why we partnered with Janssen Pulmonary Hypertension to share treatment journey stories from two patient advocates and some tips from a healthcare provider who specializes in PAH.

transcript:

Emily: My name is Emily. I work in healthcare here in Nashville, but I'm sharing my story with you as a person with pulmonary arterial hypertension (PAH). Thirteen years ago, I married Brad, and now we're raising two little girls, three cats, and 12 chickens.

(cacophony of animal sounds)

Our dream moving out here four years ago, was to have some land, to have a garden, and to have chickens. Just a few months later, I was diagnosed with PAH.

I'd just had my youngest child, and just kind of knew that things were not right, but couldn't quite put my finger on it. Doctors would say, "You just had a baby so you're just tired," or, "You can't climb that flight of stairs because you haven't lost your baby weight." We could attribute my symptoms to other things, but my gut said something's not right. But I didn't give up. I found a doctor who really listened.

My doctor got the feeling that something was not right, too. My EKG wasn't normal. She sent me to a cardiologist who performed my echocardiogram. Those results sent me to a pulmonologist. I have a fantastic relationship with my pulmonologist. We work TOGETHER in managing my PAH. Brad and I keep life moving together.

Brad: When Emily was first diagnosed, it was scary, and I was uncertain of what that diagnosis meant. We went to all of her appointments together. She included me in everything.

You know, there's not a day that goes by that I don't think about how she's doing at work or at home if I'm not around. Your mind can wander and sometimes it can wander too far, and you can scare yourself. She means everything to me.

Not knowing what the future holds is hard, but taking it day by day helps us… But I see her struggle on her worst days. She's always there beside me and tells me she's okay, even though sometimes she's not. When we go out on a short walk with the kids or I watch Emily play with them or garden, I make sure to pay attention, and sometimes I see that she stopped walking. It's just noticing the little things and saying, "Okay, this is making her tired," and to slow down.

Emily: I couldn't manage all of this without Brad. We had to figure out how to make our dreams still work in the day to day, while still managing my PAH. Working, being a mom—all of that is a struggle. But we found a really nice balance dividing and conquering and making life work. When my four-year-old wants me to pick her up, Brad will swoop in and pick her up. I don't

always have a lot of energy to give to them. Having PAH is hard raising two kids; just getting them ready and out the door is tough.

Many times, I talk about what my dreams are, and Brad helps me make them happen. And it's a success because we do it, and that's what I want life to look like. That means being really open with my doctor. She knows I'll call her if I have any concerns. She wants to help, but she can't do that if she doesn't know what's happening. We talk about how things are going, what life looks like. Life doesn't stop when you have PAH.

I'll tell her how long I was able to work that week, and how I have been feeling, how many steps I tracked each day, whether I'm able to garden, if I can care for my chickens, and most importantly, if I can take care of my girls. The ability to do these things, even if it’s challenging, means a lot to me.

The day that I can't do some of these things will be a different conversation. It is important to me to do what I can, and recognize it may be at a different pace.I see my doctor regularly, and we evaluate how I am doing at that point and what life looks like. The goal is always to take care of and monitor my health.

I started UPTRAVI® (selexipag) a few years ago. My doctor said UPTRAVI has been shown to help delay or slow down the progression of PAH and to help lower the risk of PAH-related hospitalizations.

It’s important to me to delay disease progression, and I really appreciate the convenience of an oral medication. My doctor reviewed the important safety information with me and we discussed that I should tell him if I’m experiencing any side effects, such as reddening of the skin, headache, nausea, or vomiting. Also if I am pregnant or planning on getting pregnant.

My hope is that I am able to continue to work and to care for my children. And sometimes that means making compromises in life. Sometimes when friends want to go walk and get together, I just can't quite do what everybody else does. And that's okay. Life looks different for me, I move at a different pace. But I'm still moving and I'm still walking.

And I'm still having life look like what I want it to look like. My kids want to play in the yard, they want to go and do activities like going to the park or beach. My kids know that my lungs aren't great. They know that I have to take medicine. They know that I have to wear oxygen at night. They see that their mom has to have those things. It takes a while to find a new normal, you miss what you thought life would look like.

A big part of life with PAH is the learning curve. It can be a hard learning curve to be a good patient and follow the treatment regimen that my doctor and I developed. There's so much to know, there's so much new terminology. The tests are new, the choices are new, the medications are new. It all is so very much to take in at one time.

It's almost like you need an interpreter to help you walk through and break things down for you. It's a lot to process. It's almost like its own language, its own world. I would say there's a large communication gap between patients and physicians or providers. It's a new world for patients …you're unsure of the new normal, you're unsure of what life might look like, what things are expected, what things are worth reporting.

I know someone else who was diagnosed with PAH recently. She says she knows that things aren't feeling right, that things are off for her. As someone with PAH, I'm encouraging her to be honest with her healthcare team about how she feels, and let her know they can best help her if she is open and tells them how she's feeling.

You learn certain things are normal for you and other things are abnormal for you. And they're all worth noting and reporting. You've got to communicate all that to your doctor. They can't help you if they don't know.

Brad: The advice that I would give anyone who has a loved one that has PAH is … to have a 'stop and smell the roses' mentality. Slow down the pace of the family so someone who isn't feeling well still feels included and is able to participate. Our focus has shifted to what is important—being able to do things together and enjoy time together. Because the most important thing is to be with the ones you love.

Emily: Part of living with PAH is learning a rhythm. You learn what your new normal is going to look like. And so I'm an advocate for education. I think the more information you have, the more peace it brings. There's a lot to know, and this is definitely a journey, but I think my family and I are on the right path.

Video 3 transcript:

Dr. Oudiz: It surprises a lot of people—but, if I were to put a couple of people living with pulmonary arterial hypertension, or PAH for short, at a table with other people, and start chatting with them all, you may be hard pressed to know which ones are affected by PAH.

Although some patients with PAH are on oxygen, some have swelling in their feet, and some have shortness of breath just talking to you— many are unidentifiable at first glance. Similar to patients with other conditions that don't always have external manifestation. My name is Dr. Ron Oudiz. I've been a cardiologist and specialist in treating pulmonary arterial hypertension for more than 20 years.

PAH has a significant effect on patients. PAH is an invisible disease. There's no cast or crutches, but it can be really difficult for some patients with PAH to walk across a parking lot, even though they may look fine when they are just sitting in a car. Their own family members

might judge them, because even though they appear okay, they might not be up to doing the dishes, or cleaning, or going out.

Patients have told me they feel guilty for not being able to do more, and it breaks my heart to hear that. I understand the burden of the disease. We try to explain patient limitations to their friends and family without affecting the morale of the patient. Being newly diagnosed is a very difficult experience. Every patient is different—what they want to hear, how they feel in that moment, what they already know.

As a doctor, I know a lot about how to talk to PAH patients about their diagnosis. I try not to overload them on the first visit. My team sees a lot of PAH patients in various stages of their condition. We specialize in the diagnosis and treatment of PAH. We follow the patients closely, and we follow patients throughout their journey. I tell them the underlying problem is that the blood vessels of the lungs are narrow, and because of this, the heart has difficulty pumping blood through the lungs.Although we often do not know what causes the PAH, there are many treatment options for PAH.

A lot of patients are afraid to ask questions. So, I tell them to write down their questions at home before coming to my office, because sometimes it's too much for them to remember when they come in. And I tell them I'm ready to listen. I sit down and I say, fire away. Building an open relationship with the patient is critical.

In each visit, I set aside a lot of time for each PAH patient, not only for my evaluation, but for discussion. My team and I spend much more than my usual 10-15 minute time slot for a typical follow up patient without PAH. I tell them, “We'll answer whatever you need answered” and I think that that sets them at ease and helps us build a relationship based on trust.There's incredible variability to how patients prepare for office visits and keep records.

Some patients bring in binders with records of their weight, their vital signs, their test results. They measure their own exercise performance, like a six minute-walk test. Patient symptoms are also highly variable: Most of the time, the manifestation is shortness of breath, getting really dizzy, but it could also be retaining water, or even passing out.

If a patient comes in with that binder of recordkeeping, it’s very helpful. But it’s also valuable to get the perspective of someone who lives with the patient. Often, a family member or friend can remind a patient to tell us about symptoms they’re having that they might not have mentioned. It really helps because some patients are not really aware of certain behaviors or feelings they’re expressing. Some become stoic in the doctor’s office, and some are afraid because they think their complaint might lead to potential problems and they are often afraid of what they don't know. They’ve already been through a lot.

If it's a patient who has advanced PAH or has progressed a lot, I get them prepared to deal with the onslaught of information and the challenges they're about to face. If it’s a patient who is going to be on oral therapies, I talk about side effects and what to expect, to help them

understand how the therapy works and what they might experience.And I try to get them to understand the importance of taking their medications, of following directions, and of close follow-up.

I tell them what I see on physical exam, some of the test results, like the echocardiogram, the right heart catheterization, and blood tests. I tell them there's so much that we can do, and our goal is to get them to respond well to therapies. Then we can give them a better idea of how they're doing after some time on treatment. We talk about treating the different key pathways and about medications that either block something that’s closing the arteries, or helps to open the arteries. I tell them how, when, and what to expect and be prepared for when they are told of their diagnosis and throughout their care. A two-way dialogue is so important. And to ensure we’re treating the whole person, not just the disease.

We talk about personal goals, and how these goals relate to living with the disease, and what their daily life might look like with the goals they’ve set for themselves. It all starts with setting short term goals at first. It’s about benchmarks. Can you walk up the stairs? Do you have to stop along the way?

We celebrate and give them encouragement when there is a clinical improvement. Or we’ll give them some perspective, if they're not improving. And we can modify the treatment plan if needed. It’s about making decisions together.PAH is a progressive disease. We often have goals of delaying disease progression and reducing risk of PAH related hospitalization.

UPTRAVI is approved to help delay disease progression and to reduce the risk of PAH-related hospitalizations. UPTRAVI was studied in the largest randomized, multicenter, double-blind, placebo-controlled outcomes trial ever conducted in patients with PAH, WHO Group I that included 574 patients receiving UPTRAVI and 582 receiving placebo (sugar pill). Nearly all patients were WHO functional class II or III at the beginning of the trial and received UPTRAVI for up to 4.2 years, with an average of 1.4 years. This clinical trial evaluated the efficacy and safety of UPTRAVI in patients with PAH, and showed that UPTRAVI demonstrated a 40% risk reduction in disease progression versus placebo. Doctors prescribing UPTRAVI are often specialists in PAH.

I start with the lowest recommended dose of UPTRAVI twice a day and slowly increase the dose based on how patients adjust to treatment. If they experience side effects, such as headache, nausea, muscle pain, or other side effects they are not able to tolerate, then physicians can lower the dose until they find a dose that the patient can tolerate or that they're comfortable with. Patients may think more is better, so I try to make the point as often as possible: It should be the dose that is right for them.

When prescribing UPTRAVI to my patients, I go over the side effects they may experience such as headache, diarrhea, jaw pain, nausea, muscle pain, vomiting, pain in arms or legs, and flushing and I encourage them to tell me about all the medicines they are taking since this may

cause side effects and if they have liver problems, are pregnant or planning to become pregnant, or have any other medical conditions.

While physicians are the experts in treating PAH, it's important that we recognize that patients and their families are the experts on their needs and their personal goals. That's why our focus is always on creating a strong patient/physician relationship, built on trust, to ensure we identify a treatment approach that helps them reach their goals.

Emily is partnering with Actelion Pharmaceuticals, US, Inc. to share her story. She has been paid an honorarium for her time. Individual results may vary. Please consult with your healthcare team for treatment and medical advice.