

NARRATOR: This is Jennifer. She's a speech pathologist and the mother of two young daughters

JENNIFER: I live every day for my girls. I push and push, because being there for them is so important to me and it's so important to them.

But I also have pulmonary arterial hypertension (PAH). And that changes everything.

When my husband has to travel for work, it's just the girls and me.

We live in New York City where I have to walk a lot. I have to get the housework done. I have to get the kids to school. I have to go to work. I spend all my energy doing whatever my kids are doing. Like every other mom I know.

Before PAH, I was very active. I used to run 30 miles every week and before PAH, I ran six marathons all over the country.

But then things started to change—even when I wasn't running. I was short of breath. I felt like I had a lot of pressure in my chest. Dizzy when I stood up. My legs were so swollen, my ankles were bigger than my thighs. It wasn't clear how sick I really was. Something was wrong, but I didn't know what.

I finally discovered what was happening to me.

I was diagnosed with PAH four years ago. Two days after Christmas, actually.

The doctors weren't sure what was wrong when they met me. I was a young runner who prior to experiencing these symptoms, did not have a history of health issues.

NARRATOR: When a physician suspects PAH, they may run a variety of tests, such as an echocardiogram, pulmonary function tests, and/or a lung scan. Often, patients will be referred to a pulmonologist or cardiologist and a right heart catheterization procedure is done to measure pressure in the heart and arteries of the lungs to confirm a diagnosis of PAH.

JENNIFER: I always tell the doctors, the numbers we have to remember are 8 and 6. The current ages of my little girls.

My doctors have gotten to know me and my daughters ever since that first time I walked into their office with my one daughter in one hand and my other daughter in the stroller.

My daughters have never been away from me a day in their lives. The doctors could see how my treatment protocol had to work with my life.

At the beginning, I wasn't always committed to my health. When I was feeling well, I didn't always make time for follow up appointments.

But there were times when I was struggling to just follow my regular routine. When my walk home became more challenging, or if I couldn't carry my backpack, I knew it was time to see my doctor.

And, in the past, I might not have said anything unless the healthcare team asked. Now, I want them to understand my goals and needs, and I recognize that I need to speak up and advocate for myself.

Now, I go in to see the healthcare team regularly. They'll ask questions specific to my life, my needs and my treatment. They're like family. I've found the right team.

My first piece of advice to anyone with PAH is find a healthcare team that you trust so that you feel comfortable sharing your personal thoughts, concerns and ideas. You should make living with PAH be about successes as much as it's about challenges.

My other advice is to educate yourself. Find reliable resources. Then you can make a list of questions, so when you see a doctor, you have a starting point. You'll know what to share, and what to ask.

I like to talk about my test results with my doctor—things like echocardiograms, pulmonary function tests, lung scans, and right heart catheterization results. I'm really involved in my care.

I know people are sometimes afraid to voice their thoughts to their healthcare teams, because they're the medical experts.

But I'm the expert on me. My healthcare team are experts in the medical field of PAH, and I'm the expert on how I want to live my life with PAH.

**NARRATOR:** Understanding her own needs and ensuring that her doctors do as well helps Jennifer to make important decisions about her health with her doctors.

**JENNIFER:** My medical team helped me set goals in the beginning, as they were getting to know me.

**NARRATOR:** In patients with PAH, arteries in the lungs may be affected when there is too much or too little of three substances, nitric oxide, endothelin, and/or prostacyclin, that occur naturally in your body. There are PAH treatments available that target each of these three key pathways.

JENNIFER: After I was diagnosed, we started to treat two of the three key pathways for PAH using an endothelin receptor antagonist, also known as an ERA, and a phosphodiesterase type 5 inhibitor, commonly called a PDE-5 inhibitor. I was on this combination for some time, but then my doctors said they wanted to treat the third key pathway using an oral prostacyclin pathway agent, called UPTRAVI.

NARRATOR: Arteries in your lungs may narrow when there is too little prostacyclin in your body. UPTRAVI (selexipag) targets the prostacyclin pathway.

JENNIFER: My doctor told me about UPTRAVI and how it can help delay disease progression and lower the risk of being hospitalized for PAH. He reviewed the important safety information with me. We discussed what I should tell my doctor at each visit, if I'm experiencing side effects such as reddening of the skin, headache, nausea, or vomiting. Also if I am pregnant or planning on getting pregnant.

You need to look at your life and say this is who I was before this diagnosis, and this is who I want to be after. And then, make small goals.

Little benchmarks that lead to the next goal I plan for myself. I think that keeps you positive. When you reach that goal, you can say look, I did this.

I don't have to run a marathon to live a normal life. My priorities are being able to take care of my girls and to work. And my main goal is to live the life I want to live.

## **IMPORTANT SAFETY INFORMATION**

### **What is the most important information about UPTRAVI?**

#### **Who should not take UPTRAVI?**

- Do not take UPTRAVI if you take gemfibrozil because this medicine may affect how UPTRAVI works and cause side effects

### **What should I tell my doctor before taking UPTRAVI?**

Tell your doctor if you:

- Have liver problems
- Have narrowing of the pulmonary veins (veins in your lungs). This is called pulmonary veno-occlusive disease (PVOD)
- Are pregnant or plan to become pregnant. It is not known if UPTRAVI will harm your unborn baby
- Are breastfeeding or plan to breastfeed. It is not known if UPTRAVI passes into your breast milk. You and your doctor should decide if you will take UPTRAVI or breastfeed. You should not do both
- Have any other medical conditions

- Are taking any other prescription or over-the-counter medicines, vitamins, or herbal supplements

### **What are the possible side effects of UPTRAVI?**

The most common side effects are:

- Headache
- Nausea
- Pain in arms or legs
- Low red blood cell count
- Diarrhea
- Muscle pain
- Temporary reddening of the skin (flushing)
- Less appetite than usual
- Jaw pain
- Vomiting
- Joint pain
- Rash

Talk to your doctor if you have a side effect that bothers you or does not go away. These are not all the possible side effects of UPTRAVI. For more information, ask your doctor or pharmacist.

You may report side effects to **FDA at 1-800-FDA-1088** or [www.fda.gov/medwatch](http://www.fda.gov/medwatch).

Keep UPTRAVI and all other medicines away from children.

### **What other medicines might interact with UPTRAVI?**

- UPTRAVI and other medicines may affect each other, causing side effects. Tell your doctor about all the medicines you are taking. Do not start any new medicine until you check with your doctor

### **How should I take UPTRAVI?**

- Take UPTRAVI exactly as your doctor tells you to take it. Usually, your doctor will have you take UPTRAVI twice a day. Taking UPTRAVI with food may help you tolerate UPTRAVI better
- Swallow UPTRAVI tablets whole. Do not split, crush, or chew tablets
- Tell your doctor if you have any form of liver disease. Your doctor may need to change your dose of UPTRAVI
- UPTRAVI is measured in micrograms (mcg). Tablets come in the following strengths: 200, 400, 600, 800, 1000, 1200, 1400, and 1600 mcg

## What is UPTRAVI?

- UPTRAVI® (selexipag) is a prescription medicine used to treat pulmonary arterial hypertension (PAH, WHO Group 1), which is high blood pressure in the arteries of your lungs.
- UPTRAVI can help delay (slow down) the progression of your disease and lower your risk of being hospitalized for PAH.
- It is not known if UPTRAVI is safe and effective in children.

Please see full [Prescribing Information](#) and [Patient Product Information](#).

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