

DR. OUDIZ: It surprises a lot of people—but, if I were to put a couple of people living with pulmonary arterial hypertension, or P-A-H for short, at a table with other people, and start chatting with them all, you might 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 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, and I 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 overwhelm 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 throughout their journeys.

Patients consume a lot of information from the internet. So, one of the first things I do, especially early on in their diagnosis, is refer them to reliable resources. I aim to give new patients hope and understanding. I strive to be sure the information they obtain after leaving my office is accurate.

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 don't 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 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- to 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 at home.

Patient symptoms are also highly variable. Most of the time, the manifestation is shortness of breath, and/or getting really dizzy. But it could also be retaining water, or even passing out.

If a patient comes in with that binder of his or her records, it's very helpful.

But it's also valuable to get the perspective of someone who lives with or cares for 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're 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 the 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 hospitalization.

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, or a 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. This means that 27% of patients treated with UPTRAVI experienced a PAH disease progression event versus 41.6% of patients not taking the UPTRAVI. In this clinical trial, disease progression was defined as a serious event, such as hospitalization for PAH; the need to start injectable PAH medications or chronic oxygen therapy; lung transplantation; a decline in 6-minute walk distance often referred to as 6MWD, combined with the worsening of functional class, often referred to as FC, or need for additional PAH therapy; or death.

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 the arms or legs, and flushing and I encourage them to tell me about all the medicines that 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 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.

## **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](#) on [\*\*UPTRAVI.com\*\*](#).

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Dr. Oudiz is a paid speaker for Janssen. The information does not replace or substitute medical advice from your healthcare providers. Please consult with your healthcare team for treatment and medical advice.