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DR. RAVAL: Every person living with pulmonary arterial hypertension is different—one size does not fit all. So as a physician, or as a member of a pulmonary arterial hypertension care team, we have to understand their individual needs and where they are in their journey.

My name is Dr. Abhijit Raval and I'm a pulmonologist at a regional clinical care center that specializes in the treatment of patients with pulmonary arterial hypertension. My team is dedicated to helping patients learn how to manage their condition, whether they are newly diagnosed or have been living with pulmonary arterial hypertension for a long time.

For my patients, I emphasize the importance of continuous communication with myself and the whole care team.

My focus is making sure we are staying on top of every aspect of my patients' care together. I educate my patients to ensure they understand pulmonary arterial hypertension is high blood pressure in the arteries of their lungs and how it affects the body, I tailor our discussion to meet each patient's level of knowledge. I find visual tools help my patients understand: for my newly diagnosed patients, I have a diagram of the heart and lungs, and for those with more advanced knowledge levels, I have one showing how a right-heart catheterization works, along with echocardiogram images.

I know there is a broad spectrum of how much someone living with pulmonary arterial hypertension wants to know about the condition. Some of my patients even want to read scientific data and literature. It really comes down to: have we given them the right amount and right level of information they want about the disease, and how we are going to manage it together?

I don't want them to learn ONLY from the internet, as there can be misinformation out there. I want to provide them with credible, reliable, current resources. And we are always counseling about the importance of ongoing, timely follow-ups and monitoring their status.

Through routine tests, we keep a close eye on patients' disease status. These tests include the 6-minute walk test, blood tests, and other factors to identify signs of progression, such as a patient's WHO Functional Class, that signal a patient's pulmonary arterial hypertension may be changing.

We use routine monitoring tests at the 3-month and 6-month visits to identify trends that can help my care team know when to consider adjusting a patient's treatment plan or try something new.

So, as clinicians, we are looking at the data to get a comprehensive view of all patients is experiencing. Because the more knowledge our patient has, the more power they hold over their treatment plan.

We also discuss factors that can affect overall health, including diet, exercise, and managing stress. Continuing to provide information and resources for patients, regardless of where they are in their treatment journey, can help them to better manage the disease.

It's important that we give our patients many opportunities to ask questions. Questions during, and even in between appointments, can help patients be their own advocates. It's common for patients to realize they have questions they forgot to ask during their appointment, so we ask them to keep a diary to keep track of their questions, experiences and medications between visits. My patients who use our patient portal can send their questions there, too.

I also encourage my patients to join support groups where they can talk to support group leaders, listen to the speakers, and talk to their peers. They have the opportunity to learn a lot there and interact with people who may be going through the similar experiences. That connection can be very impactful for a person living with pulmonary arterial hypertension. Support groups can potentially help my patients think about their personal goals for treatment. It's important that we align these with our clinical goals, which include delaying disease progression and reducing the risk of pulmonary arterial hypertension-related hospitalization.

I work to be a helpful guide to my patients as they make decisions based on where they are in their personal journey. Disease progression may occur with or without symptoms, so I believe it is important to talk to my patients about how to recognize the signs or symptoms they may be experiencing that may indicate a change in the progression of their pulmonary arterial hypertension.

It's vital they notice when one-time occurrences become patterns so, together, we can know when to make changes in the way we are managing their pulmonary arterial hypertension. I try to explain how disease progression could impact them.

We will look at the data from their tests—like the 6-minute walk test, the echocardiogram, the right heart catheterization, the physical exam, and the blood tests. We interpret their clinical test results and their symptoms and talk to them about what they may still be able to do—and what they may not be able to do.

For example, a number of my patients love gardening. And we tell them they might be able to spend time in the garden, but there will be limits. It can be hard to talk about, but with better partnership, we can work together to set and evaluate individual goals for each patient and evolve them as appropriate over time.

I think ongoing counseling to help prepare my patients physically and emotionally is an incredibly important part of my role in helping someone manage pulmonary arterial hypertension successfully. I encourage patients to be open with people—their friends, their spouse, their family and loved ones, and also their doctors and nurses about how they are feeling—to share the good days and the bad. We all must play a part in supporting someone living with pulmonary arterial hypertension.

Nurses may help patients manage their side effects, assist with the procedures and help patients during their office visits. The broader care team can also help patients make sure they have access to the therapies, and getting the medication refills, and follow up to answer questions between the appointments.

Everything we do has to have a patient-centered approach. We have to work together to make decisions. As the care team, we educate them about the possibilities so they can see when change is necessary, especially when it comes to something like escalating treatment. It's important to know how well their medications are working, and I tell them we have many options for managing their pulmonary arterial hypertension. Our goal is to get them to respond well to therapies.

When I feel prescribing UPTRAVI® may be appropriate for my patients, I help them understand how UPTRAVI® can help delay, or slow down, disease progression and lower the risk of being hospitalized for pulmonary arterial hypertension, and what they might experience.

NARRATOR: UPTRAVI® 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. Tablet not shown at actual size.

DR. RAVAL: I tell them: we are going to start with lowest dose of UPTRAVI® and slowly increase based on how they adjust to the treatment, and I explain why.

NARRATOR: Do not take UPTRAVI® if you take gemfibrozil because this medicine may affect how UPTRAVI® works and cause side effects.

DR. RAVAL: I don't give them a goal dose. It should be the right dose for that specific person. We talk about finding their personal dose and managing the side effects.

It is important for me to help manage expectations. The better prepared they are, the more likely we can work together to get them to their personal dose. So I go over the side effects they may experience on UPTRAVI® such as headache, diarrhea, jaw pain, nausea, muscle pain, vomiting, pain in the arms or legs, or flushing.

If they experience side effects they are unable to tolerate, then we lower the dose. My patients sometimes feel they need to reach the highest dose, but I try to remind them that higher isn't always better and we work together to find the dose that is right for them.

When prescribing UPTRAVI® to my patients, I also ask them if they have liver problems, are pregnant or planning to become pregnant, or have any other medical conditions. And of course, I will ask about all the other medications they may be taking. Taking UPTRAVI® and other medications may affect each other and cause side effects.

I don't want people to get anxious or frustrated and discontinue therapy. Patients need a lot of help when we are finding their personal dose, so we follow up with them frequently. We encourage patients to call the office to check in and then we schedule separate follow-up

appointments, as necessary. There are various checkpoints in our treatment plan to help our patients navigate challenges. It really helps boost patients' morale to know they are not going through this journey by themselves.

Pulmonary arterial hypertension patients may each have different goals, which can include things that others take for granted. Such as, being able to walk more, and spend time with their families without it feeling like a physical struggle all the time.

I encourage all my patients not to give up—keep asking questions and keep talking to their care team about good days and bad days so they can be best cared for. In my opinion, that's how we as healthcare professionals can be the best partner with pulmonary arterial hypertension patients to try to get them on a successful pathway.

### **IMPORTANT SAFETY INFORMATION**

- **Do not take UPTRAVI® if you** take gemfibrozil because this medicine may affect how UPTRAVI® works and cause side effects
- **Do not take UPTRAVI® if you** are allergic to selexipag or any of the ingredients in UPTRAVI®
- **Before you take UPTRAVI®, tell your healthcare provider about all your medical conditions, including 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
  - 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
- Diarrhea
- Jaw pain
- Nausea

- Muscle pain
- Vomiting
- Pain, redness or swelling at the injection site with UPTRAVI® for injection
- Pain in arms or legs
- Temporary reddening of the skin (flushing)
- Joint pain
- Low red blood cell count
- Less appetite than usual
- 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®?

### UPTRAVI® Tablets

- 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

### UPTRAVI® given by intravenous (IV) injection

- Your healthcare provider will give you UPTRAVI® into your vein through an intravenous (IV) line
- Your healthcare provider will decide how much UPTRAVI® for injection you will receive each day based on your current dose of UPTRAVI® tablets

### 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](http://UPTRAVI.com).

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### References:

1. UPTRAVI® (selexipag) full Prescribing Information. Actelion Pharmaceuticals US, Inc.