

LEARN ABOUT

GETTING TO YOUR
PERSONAL DOSE


SETTING GOALS
AND EXPECTATIONS

FINDING YOUR PAH
SUPPORT TEAM

STAYING MOTIVATED
THROUGH TREATMENT

DOSE ADJUSTMENT & GOAL GUIDE

Please see Important Safety Information throughout and on page 23, and [click here](#) for full Prescribing Information and Patient Product Information.

A woman with dark hair is standing in a shower. She is wearing a green towel wrapped around her waist. She is looking directly at the camera with a neutral expression. The background consists of dark red or maroon square tiles. To her right, there is a metal shower rack with several bottles on it. To her left, there is a vertical metal bar, likely part of the shower door or wall.

This brochure contains QR codes that you can activate using your smart phone with a QR code scanner. To activate a QR code:



- Step 1. Open the camera on your phone
- Step 2. Point your camera at the QR code
- Step 3. Follow the prompt that appears on-screen to visit UPTRAVI.com

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TAKE ON TREATMENT

PAH can feel overwhelming. That's why we're here to help guide you through starting and staying on prescribed treatment with UPTRAVI® (selexipag). With support at each step, you can face PAH with all you've got.

USE THIS GUIDE TO:

UNDERSTAND

The Dose Adjustment Phase with UPTRAVI®

IDENTIFY

What you want to get out of treatment

LEARN

How to work with your caregiver and healthcare team to get there

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.

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®

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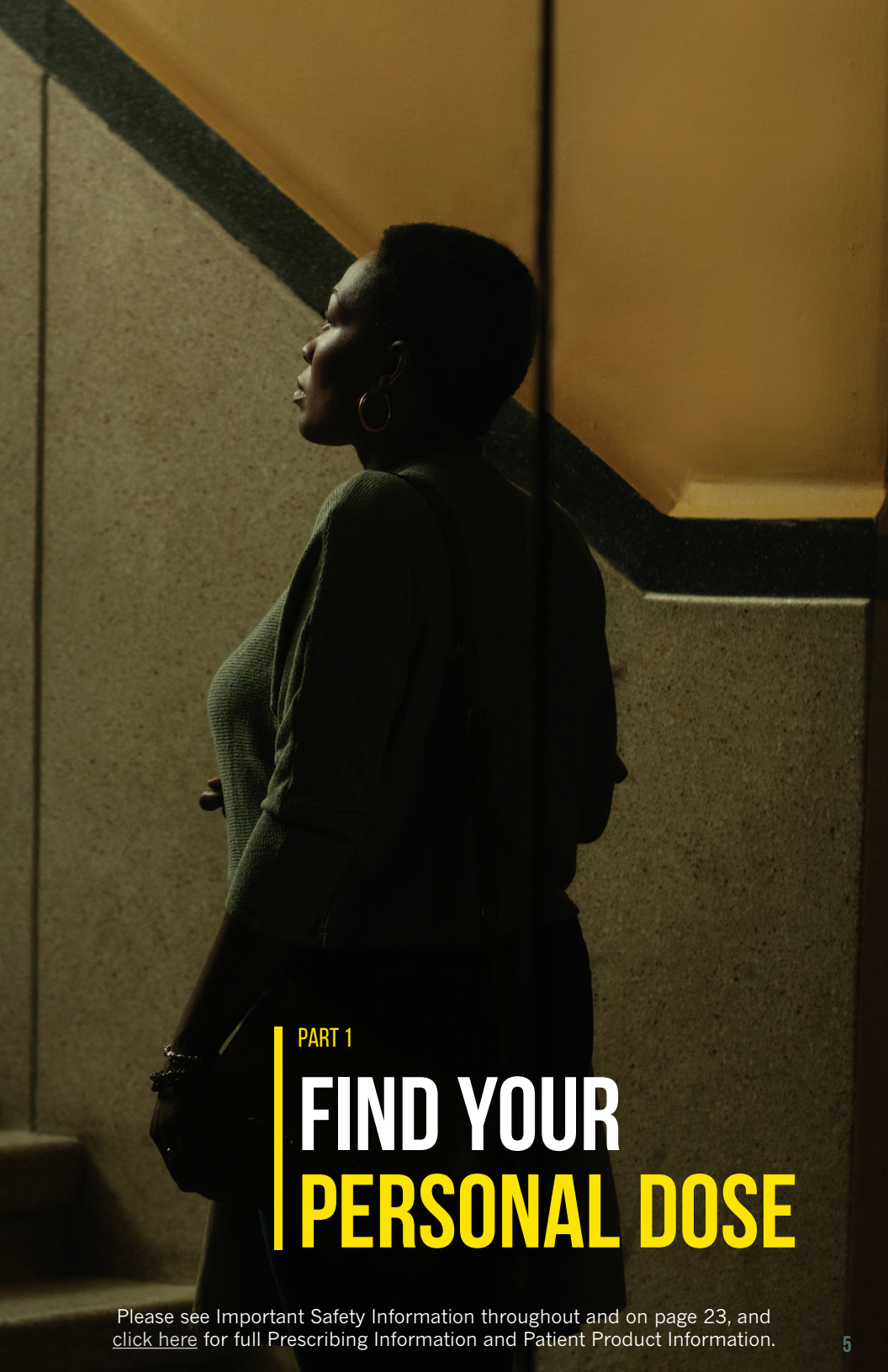
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PART 1

FIND YOUR PERSONAL DOSE

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SET YOUR GOALS

Now that you and your healthcare team have decided to add UPTRAVI®, think about why you made this choice. Setting goals is an important part of your prescribed treatment plan. You can discuss these topics with your doctor:

01 What you're looking for from your treatment

02 Goals you are working towards

03 What's most important to you

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STARTING AND ADJUSTING TO TREATMENT

There isn't one specific dose of UPTRAVI® that all people take

- ▶ Each person has a different dose based on how his or her body responds and adjusts to the medicine
- ▶ The Dose Adjustment Phase (also called titration) is how you and your healthcare team find your personal dose
- ▶ This is just a temporary part of treatment until you reach the dose that is right for you
- ▶ Remember, each person's dose is unique and the goal is *not* to reach the highest dose, 1600 mcg
- ▶ In a clinical trial, there were similar results when people reached the right UPTRAVI® dose for them

EVERYONE'S BODY IS UNIQUE. THAT MEANS PEOPLE WILL RESPOND DIFFERENTLY TO TREATMENT WITH UPTRAVI®.

THINK OF ROOM TEMPERATURE AS AN EXAMPLE



Some people feel more comfortable at **higher temperatures**



Others feel better at **lower temperatures**

You have to adjust the thermostat to determine which temperature is right for you. The same goes for UPTRAVI®: **Each person's dose is unique.**

IMPORTANT SAFETY INFORMATION



- **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

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HOW THE DOSE ADJUSTMENT PHASE WORKS

WHEN YOU START UPTRAVI[®], YOUR PAH HEALTHCARE TEAM WILL SLOWLY ADJUST YOUR DOSE TO FIND YOUR PERSONAL DOSE.

SLOWLY INCREASE DOSE TO REACH YOUR PERSONAL DOSE



Starting dose
200 mcg
(taken twice daily)

DECREASE DOSE IF CURRENT DOSE CANNOT BE TOLERATED



Maintenance Dose
Your personal dose
(taken twice daily)

- ▶ UPTRAVI[®] dose strengths range from 200 mcg to 1600 mcg
- ▶ Tablets are not actual size

During the Dose Adjustment Phase, your doctor will change your dose, usually once a week. Be sure to take UPTRAVI[®] exactly as directed by your doctor. Remember, in a clinical trial, there were similar results when people reached the right UPTRAVI[®] dose for them. It's okay to speak with your doctor about lowering your dose. The goal is not to reach the highest dose, 1600 mcg, so don't hesitate to be honest with your doctor about any side effects you cannot tolerate.

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WORKING WITH A SPECIALTY PHARMACY

Anticipate a call from your specialty pharmacy to alert you of your next shipment.

- ▶ **DO NOT CHANGE YOUR DOSE OR STOP** taking UPTRAVI® unless your doctor tells you to
- ▶ **TRY TO TAKE UPTRAVI® WITH FOOD** This may help you tolerate the dose better if you are having side effects with UPTRAVI®

REACHING YOUR PERSONAL DOSE

After the Dose Adjustment Phase, you will have found your personal dose of UPTRAVI®.

Your healthcare team may refer to this as your maintenance dose. This will be the dose you continue to take on a regular basis, unless your doctor tells you otherwise.

WHAT SHOULD I DO IF I MISS A DOSE?

- ▶ If you miss a dose of UPTRAVI®, take it as soon as you remember. If your next scheduled dose is due within 6 hours, skip the missed dose. Take the next dose at your regular time
- ▶ If you miss 3 or more days of UPTRAVI®, call your doctor to see if your dose needs to be changed
- ▶ If you take too much UPTRAVI®, call your healthcare team or go to the nearest hospital emergency room right away



SUGGESTIONS FOR DISCUSSING SIDE EFFECTS WITH YOUR DOCTOR

DOSE ADJUSTMENT MAY COME WITH SIDE EFFECTS. THESE WILL VARY FROM PATIENT TO PATIENT. IF YOU'RE HAVING TROUBLE WITH SIDE EFFECTS, CALL YOUR DOCTOR'S OFFICE RIGHT AWAY TO LET THEM KNOW.

The following table has been adapted and modified from Kingman M, Archer-Chicko C, Bartlett M, et al. Management of prostacyclin side effects in adult patients with pulmonary arterial hypertension. *Pulm Circ.* 2017; 7(3):598-608. This table reflects the authors' experiences with PAH and other disease states and contains common approaches to managing potential side effects. **Always consult with your doctor before attempting any of the following suggestions to ensure that your individual needs and considerations are taken into account.**

PAIN MANAGEMENT	<ul style="list-style-type: none">• Talk to your doctor about pain management strategies, referrals to specialists, and medication to help manage your pain• To help manage pain, it's okay to speak with your doctor about lowering your dose• Ask your doctor about methods (other than medication) for promoting relaxation and pain relief
LEG PAIN	<ul style="list-style-type: none">• Talk to your doctor about screening for an iron deficiency or medication to help cope with leg pain
JAW PAIN	<ul style="list-style-type: none">• Talk to your doctor about strategies to help ease jaw pain, such as taking slow bites or sips of water, sucking on saltine crackers or hard candy, and chewing gum before eating

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HEADACHE	<ul style="list-style-type: none"> • Talk to your doctor about over-the-counter pain medication to help cope with mild to moderate headaches • If your headaches become severe, talk to your doctor about appropriate next steps and treatment options
DIZZINESS	<ul style="list-style-type: none"> • If you are prescribed blood pressure medication, talk to your doctor about decreasing the dose as you adjust to UPTRAVI®. Monitor your blood pressure as directed by your doctor • Talk to your doctor about strategies such as drinking more water and being careful about changing positions too quickly • To help manage dizziness, talk to your doctor about decreasing your dose
NAUSEA/ VOMITING	<ul style="list-style-type: none"> • Talk to your doctor about medication to help cope with nausea and vomiting • Talk to your doctor if you think you may be pregnant • Ask your doctor about a referral to a gastrointestinal specialist • To help manage nausea and vomiting, talk to your doctor about decreasing your dose
LOSS OF APPETITE/ WEIGHT LOSS	<ul style="list-style-type: none"> • Consult with your doctor about dietary changes to promote weight gain, such as higher-calorie meals, or smaller and more frequent meals • Talk to your doctor about other causes of weight loss
DIARRHEA	<ul style="list-style-type: none"> • To help manage diarrhea, talk to your doctor about decreasing your dose • Ask your doctor about dietary modifications or medication to help cope with diarrhea • Ask your doctor about a referral to a gastrointestinal specialist
FLUSHING/ RASH	<ul style="list-style-type: none"> • Talk to your doctor about anxiety-reducing techniques • Talk to your doctor about applying a cold compress or ice pack to the back of your neck • Talk to your doctor about slowing down your increase in dosages of UPTRAVI® to help manage flushing

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PART 2

FIND YOUR MOTIVATION

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YOU ARE STRONGER THAN YOUR BAD DAYS

WE KNOW TREATMENT IS NOT EASY. THAT'S WHY IT'S IMPORTANT TO REMEMBER THAT EVEN WHEN YOU'RE NOT FEELING YOUR BEST, YOU'RE STILL TRYING YOUR BEST.

- ▶ Stick with the plan that you and your healthcare team decide is best for you
- ▶ Remember to stay hopeful and focused on the big picture of how UPTRAVI® can help you
- ▶ Be honest and tell your doctor about any side effects you have while taking UPTRAVI®
- ▶ Advocate for yourself and your health by speaking up to your doctor

**THE GOAL IS NOT TO GET TO THE HIGHEST DOSE OF UPTRAVI®,
IT'S TO GET TO THE DOSE THAT'S RIGHT FOR YOU.**

Don't be discouraged if you have to reduce your dose or stay on a lower one. In a clinical trial, there were similar results when people reached the right UPTRAVI® dose for them.

IMPORTANT SAFETY INFORMATION

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**.

Keep UPTRAVI® and all other medicines away from children.

WHOM CAN I REACH OUT TO WITH QUESTIONS?

- **PAH healthcare team**
Share any side effects and symptoms with your doctor and nurses. This is your source for medical advice
- **Specialty pharmacy nurse**
When you were prescribed UPTRAVI®, your doctor may have requested Janssen-sponsored one-on-one educational support and visits from a specialty pharmacy nurse to help you through the Dose Adjustment Phase. Always consult your doctor for medical advice
- **PAH Companion**
If you have enrolled in the PAH Companion* program, your PAH Companion is here for you during treatment with one-on-one educational conversations and support. Always consult your doctor for medical advice

*PAH Companion is limited to education about your Janssen therapy, its administration, and/or PAH. It is intended to supplement your understanding of your therapy, and does not provide medical advice or replace a treatment plan from your doctor, nurse, or healthcare team.

FOR IMMEDIATE HELP DURING TREATMENT, I SHOULD TALK TO:

My PAH doctor or healthcare team member

IMPORTANT SAFETY INFORMATION

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.

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PERSIST WITH SOME HELP ALONG THE WAY

PAH progression doesn't stop. That's why you shouldn't either. Here are some tips and questions to discuss with your care team.

- ▶ Pay close attention to your good days, your not-so-good days, and every day in between
- ▶ Are there specific things you notice you're doing those days in terms of diet, activities, etc.?
- ▶ Is there anything that triggers side effects or helps relieve them?
- ▶ On days you're not feeling well, don't do anything too strenuous
- ▶ Take care of your body and focus on creating the most comfortable conditions you can

YOUR PAH TEAM

Remember, you're not alone in your treatment. Managing PAH is about teamwork. Each person plays a key role. Knowing each person's role can help you during the Dose Adjustment Phase and throughout treatment with UPTRAVI®.

IT ALL STARTS WITH THE MOST IMPORTANT PERSON ON THE TEAM—YOU

Each team member is essential, but think of yourself as the leader.

You're in charge of talking with the rest of the team members about how you feel. Being transparent with your caregiver and other team members will help them support you when you need it.

YOUR PAH DOCTOR & HEALTHCARE TEAM

Your PAH doctor and staff rely on you to speak up about how you're feeling.

- ▶ In order to make things as smooth as possible, you and your caregiver should keep track of how you're feeling. Let them know if you are having any problems during the Dose Adjustment Phase or anytime during treatment
- ▶ Don't be afraid to speak up if you think the dose you're on is not right for you. Your care team will help you work on feeling better or reduce your dose if needed

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YOUR CAREGIVER OR LOVED ONES

Think of your caregiver, loved ones, and/or family as the members of your team.

- ▶ They can help support you physically, mentally, and emotionally on a regular basis
- ▶ They can also give you and your PAH doctor their insight on how you're doing, especially during the Dose Adjustment Phase



Get resources to help your caregiver partner with you by holding your smartphone camera over this QR code

SPECIALTY PHARMACY/SPECIALTY PHARMACY NURSE

Discuss how you're feeling with the specialty pharmacist during your regular dose adjustment calls; they're here to help you reach your goals.

If your PAH doctor requests support services from a specialty pharmacy nurse, they can provide Janssen-sponsored one-on-one educational support and answer any questions you may have during the Dose Adjustment Phase.

If you're unsure whether you have access to specialty pharmacy nurse services, you can reach out to your healthcare provider to find out.

The specialty pharmacy nurse/pharmacist is not a substitute for the care you receive from your PAH doctor and healthcare team.

CALL YOUR PAH DOCTOR FOR ADVICE ABOUT SIDE EFFECTS

IMPORTANT SAFETY INFORMATION

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

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PART 3

FIND YOUR STRENGTH

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“STAY ON TRACK” TRACKER

Part of staying on treatment is staying motivated. This is a space to record how you're feeling and what's inspiring you to stop at nothing. Fill this out before a scheduled dose change of UPTRAVI® and use it to be prepared for doctor visits. It's important to be honest about how you're feeling on each dose of UPTRAVI® so you can be prepared for honest discussions with your doctor.

Week of:

My current dose of UPTRAVI® is ___ mcg twice

daily **How am I feeling this week?**

GOOD

JUST OK

NOT GOOD

Explain:

What inspires me this week?

What motivates me to stay on treatment?

At or before my scheduled dose change, I talked about how I was feeling with:

My PAH doctor or healthcare team member

My caregiver

My specialty pharmacy nurse/pharmacist*

Their recommendations were:

Some questions I may have for my PAH doctor are:

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**HERE IS A SPACE FOR YOUR LOVED ONES
TO ADD MESSAGES OF SUPPORT.**



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- Joint pain
- Nausea
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FIND YOUR STRENGTH

FACING PAH ISN'T
EASY. THAT'S WHY
IT'S IMPORTANT TO
REMEMBER WHY
YOU'RE ON TREATMENT
IN THE FIRST PLACE.

- ▶ Stay focused on your treatment goals
- ▶ Be honest about how you're feeling
- ▶ Remember, each person's dose is unique and the goal is not to reach the maximum 1600 mcg dose
- ▶ In a clinical trial, there were similar results when people reached the right UPTRAVI® dose for them
- ▶ You have a whole team of people behind you
- ▶ Celebrate your victories
- ▶ Do not take UPTRAVI® if you take gemfibrozil because this medicine may affect how UPTRAVI® works and cause side effects

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

