

GETTING STARTED ON UPTRAVI® (selexipag)

3 IMPORTANT STEPS THAT SHOULD HAVE ALREADY OCCURRED

You may be receiving this brochure because you and your healthcare team have decided that starting treatment with UPTRAVI® was right for you and your health. Here's a step-by-step guide to help you stay on track with your pulmonary arterial hypertension (PAH) journey. You can also find Steps to Expect on **page 26** of this brochure.

- A Janssen CarePath Care Coordinator should have contacted you to discuss any insurance-related questions and work with you to get your medication from the pharmacy. If you did not receive a call, please call a Janssen CarePath Care Coordinator at 866-228-3546, Monday–Friday, 8 AM–8 PM ET.
- Your PAH medicine is only available through a Specialty Pharmacy and they will deliver your prescription to your home.

 Your Specialty Pharmacy should have contacted you to schedule your shipment. They should continue to call you every month to confirm your prescription order and schedule the following month's delivery. If you did not receive a call, please call your Janssen CarePath Care Coordinator at 866-228-3546, Monday–Friday, 8 AM–8 PM ET.

NOTE: The Specialty Pharmacy may not ship your medicine without speaking to you first. If you miss a call from the Specialty Pharmacy, please be sure to call them back to confirm your medicine shipment and save the number to your phone so you recognize future calls.

You should have received your medication. If it has not been delivered, please call your Janssen CarePath Care Coordinator at 866-228-3546, Monday–Friday, 8 AM–8 PM ET.

Important Contact Information: Make sure that you scan the QR code using the camera on your smartphone and that your phone is connected to the internet. This will ensure that the contact information is added directly to your contacts and will also help you identify when your Care Team is calling you. If you don't have a smartphone, we encourage you to manually input and save the contact information above to your phone.



*Data rates may apply

Please see Important Safety Information throughout and on pages 16–17, and <u>click here</u> for full Prescribing Information and Patient Product Information.

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YOUR PAH MAY BE PROGRESSING SILENTLY

Your pulmonary arterial hypertension may be progressing silently, even when your symptoms seem stable. This brochure will guide you in your PAH journey by providing information and resources about treatment and living with PAH. Continue reading to find out more about UPTRAVI® (selexipag) clinical trial results, get connected with financial support, and other important information.

WHAT IS UPTRAVI® (selexipag)?

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®

ABOUT PAH PATHWAYS

CONSIDERING EVERY PATHWAY IS KEY

Arteries in your lungs may be affected when there is either too much or too little of the three naturally occurring substances in your body: prostacyclin, endothelin, and/or nitric oxide. There are PAH treatments available that target each of these three key pathways.

Endothelin Pathway

Endothelin receptor antagonists (ERAs) work here.

Nitric Oxide Pathway

Phosphodiesterase type-5 inhibitors (PDE-5is) work here.

UPTRAVI® (selexipag) WORKS HERE

Prostacyclin Pathway

Prostacyclin, prostacyclin analogs, and prostacyclin receptor agonists work here.

HOW UPTRAVI® WORKS

When you have PAH, your body may produce too little of a substance called prostacyclin. Prostacyclin activates a process that helps open arteries. UPTRAVI® works in a similar way to help open the arteries. UPTRAVI® can help when used alone or in combination with other PAH medications (ERA, PDE-5i, or both). UPTRAVI® is available as oral tablets.

IMPORTANT SAFETY INFORMATION



- Before you take UPTRAVI, tell your healthcare provider about all your medical conditions, including if you:
 - Have liver problems



To learn more about the role the three key pathways may play in PAH, hold your smartphone camera over the QR code to unlock an educational video.

UPTRAVI® (selexipag) CLINICAL STUDY

UPTRAVI® WAS STUDIED IN THE LARGEST OUTCOMES TRIAL OF ANY FDA-APPROVED PAH TREATMENT

- The trial included 1156 total PAH patients. It compared 574 patients taking UPTRAVI® with 582 patients taking a placebo (a sugar pill)
- > 80% of enrolled patients were treated with an endothelin receptor antagonist (ERA), phosphodiesterase type-5 inhibitor (PDE-5i), or both for PAH
- > 20% of enrolled patients were not on any PAH treatment

- Nearly all patients were Functional Class II or Functional Class III (PAH patients range from Functional Class I to Functional Class IV)
- Patients taking UPTRAVI® were treated for an average of 1.4 years during this study
- The trial included the most scleroderma-related PAH patients of any PAH trial (170 out of 1156)

IMPORTANT SAFETY INFORMATION



- Before you take UPTRAVI,[®] tell your healthcare provider about all your medical conditions, including if you: (continued)
 - 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

UPTRAVI® (selexipag) CLINICAL STUDY RESULTS

IN A CLINICAL TRIAL, UPTRAVI® REDUCED THE RISK OF PAH DISEASE PROGRESSION* BY 40%

The trial included 1156 total patients – 27% (155/574) of patients treated with UPTRAVI® experienced a PAH disease progression event vs 41.6% (242/582) of patients taking a placebo (sugar pill). Side effects occurring more frequently (\geq 5%) for patients on UPTRAVI® compared to patients on placebo (sugar pill) are headache, diarrhea, jaw pain, nausea, muscle pain, vomiting, pain in arms or legs, and temporary reddening of the skin (flushing).

In a clinical study of UPTRAVI, PAH disease progression was defined as a serious event, such as hospitalization for PAH; the need to start injectable PAH medications or chronic oxygen therapy; atrial septostomy (a procedure where a hole is placed between the two small chambers of the heart) or lung transplantation; a decline in 6-minute walk distance (6MWD) combined with worsening of functional class (FC) or need for additional PAH therapy; or death.

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

Please see Important Safety Information throughout and on pages 16–17, and <u>click here</u> for full Prescribing Information and Patient Product Information.

26% RISK REDUCTION OF HOSPITALIZATION WITH UPTRAVI® (selexipag) VS PLACEBO (SUGAR PILL)

This was demonstrated in the same clinical trial -13.6% (78/574) of patients treated with UPTRAVI® compared to 18.7% (109/582) of patients not taking UPTRAVI® were hospitalized.

UPTRAVI® MADE A DIFFERENCE IN HOW FAR PATIENTS WERE ABLE TO WALK

In the same clinical trial, patients taking UPTRAVI® walked an average of 12 meters (about 39 feet) farther than patients taking a placebo (sugar pill) as measured by the 6-minute walk distance (6MWD).

UPTRAVI® CAN HELP WHEN USED ALONE OR IN COMBINATION WITH OTHER PAH MEDICATIONS (ERA, PDE-5i, OR BOTH).



IMPORTANT SAFETY INFORMATION

What are the possible side effects of UPTRAVI®? (continued)



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.**



SET YOUR GOALS

Now that you and your healthcare team have decided to add UPTRAVI® (selexipag), 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

STARTING AND ADJUSTING TO UPTRAVI® (selexipag)

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

- Each person has a different dose based on how their 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 an initial part of treatment until you reach the dose that is right for you

- UPTRAVI® tablets dose strengths range from 200 mcg to 1600 mcg
- Remember, each person's dose is unique and the goal is not to reach the highest dose, 1600 mcg
- In a clinical trial, people experienced similar results when they reached their personal dose of UPTRAVI®

IMPORTANT SAFETY INFORMATION

What are the possible side effects of UPTRAVI®? (continued)



◆ Keep UPTRAVI® and all other medicines away from children.

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

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.

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.

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

HOW THE DOSE ADJUSTMENT PHASE WORKS

When you start UPTRAVI® (selexipag), your PAH healthcare team will slowly adjust your dose to find your personal dose.





Slowly increase dose to reach your personal dose





Decrease dose if current dose cannot be tolerated

Starting Dose: 200 mcg (taken twice daily)

Maintenance Dose: Your personal dose (taken twice daily)

- UPTRAVI® dose strengths range from 200 mcg to 1600 mcg
- > Tablets illustrated above are not actual size

IMPORTANT SAFETY INFORMATION

How should I take UPTRAVI®? (continued)



UPTRAVI® 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



DISCOVERING YOUR PERSONAL DOSE

- Do not change your dose or stop taking UPTRAVI® (selexipag) unless your doctor tells you to
- > Taking UPTRAVI® (selexipag) with food may help you tolerate UPTRAVI® better

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

IMPORTANT SAFETY INFORMATION

How should I take UPTRAVI®? (continued)



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

IMPORTANT SAFETY INFORMATION

What is UPTRAVI® (selexipag)?

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®
- 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

IMPORTANT SAFETY INFORMATION (continued)

What are the possible side effects of UPTRAVI® (selexipag)? (continued)

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.

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

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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	 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	Talk to your doctor about screening for an iron deficiency or medication to help cope with leg pain
JAW PAIN	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
HEADACHE	 Talk to your doctor about overthe-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

NAUSEA/ Vomiting	 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	 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	 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	 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 during the dose adjustment phase to help manage flushing



FIND YOUR MOTIVATION

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.

THE GOAL IS NOT TO GET TO THE HIGHEST DOSE OF UPTRAVI® (selexipag), 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.

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.

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 on good 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.

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 care partner 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 care partner 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

CALL YOUR PAH DOCTOR FOR ADVICE ABOUT SIDE EFFECTS

YOUR CARE PARTNER OR LOVED ONES

Think of your care partner, 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 care partner support 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 Janssensponsored 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.

Information about your insurance coverage, cost support options, and treatment support is given to you by service providers for Janssen CarePath. The information you get does not require you to use any Janssen product. The information about whether your treatment is covered by your health plan comes from outside sources. It is not a promise of coverage or payment. You should contact your health plan directly for the most current information. You are responsible for meeting your health plan requirements. Janssen CarePath cost support is not for patients in the program offered by the Johnson & Johnson Patient Assistance Foundation.

FINDING SUPPORT

SUPPORT FOR PATIENTS AND THEIR CARE PARTNERS

Janssen CarePath provides the additional support you may need to help you get started with UPTRAVI® (selexipag) treatment, once you and your healthcare provider have decided it is right for you.

Your personally assigned Janssen CarePath Care Coordinator can:

- Review your health plan benefits to help answer questions you may have about insurance coverage for UPTRAVI®
- Identify options that may help make UPTRAVI® more affordable, if needed
- Connect you with a dedicated PAH Companion for one-on-one educational conversations and support
- Identify and coordinate with a specialty pharmacy to arrange shipment of your Janssen medication
- Visit JanssenCarePath.com/UPTRAVI
- Need Help? Call a Janssen CarePath Care Coordinator at 866-228-3546, Monday-Friday, 8 AM-8 PM ET. Multilingual phone support is available

PAH Companion with Ne

Personalized Support for You

Everyone faces PAH differently. Once you and your doctor have decided that a Janssen PAH medication is right for you, you can join PAH Companion with Me—a suite of patient support resources personalized just for you. You'll receive one-on-one educational support to help with your prescribed Janssen PAH medication, plus customized tools and resources to help you take a more active role in your care.

Contact a PAH Companion

If you're receiving a Janssen PAH medication, you can learn more about PAH Companion withMe by calling 866-300-1818, Monday-Friday, 8 AM-9 PM ET.

Learn more at **pahcompanion.com**



When You Participate in PAH Companion withMe, You'll Have Access To:

My PAH Companion

One-on-one conversations with a trained, dedicated PAH Companion—someone who specializes in helping people living with PAH. Your PAH Companion will communicate with you via phone, email, or text.

My Resources

Educational resources to help with your prescribed Janssen PAH medication, tailored to your specific needs, questions, and interests.

My Tips

Ongoing support and education on living with PAH to help you partner with your healthcare team and engage with your support network.

My Community

Connection to peer support groups, current PAH information and resources, and digital tools to help you track and monitor your health.





STEPS TO EXPECT

Now that you've received your medication, there are a couple of additional PAH resources Janssen offers to help you continue treatment.

01

Connect With Your PAH Companion

Your dedicated PAH Companion will call you to learn more about you and put together personalized educational resources based on your needs and daily life. Your PAH Companion can connect you to support groups, virtual educational webinars, digital tools to monitor and track your health, and the latest PAH resources.

Your PAH Companion will call you from this number: **866-300-1818**, Monday–Friday, 8 AM–9 PM ET

Learn more at pahcompanion.com

02

A Benefits Investigation Call from Your Janssen CarePath Care Coordinator

A Janssen CarePath Care Coordinator will call to go over the results of your Benefits Investigation, including estimated co-pay and financial support options.

A Janssen CarePath Care Coordinator will call you from this number: **866-228-3546**, Monday–Friday, 8 AM–8 PM ET

PAH Companion withMe is limited to education about your Janssen PAH therapy, its administration, and/or PAH. It is not intended to provide medical advice, replace a treatment plan you receive from your healthcare team, or serve as a reason for you to start or stay on treatment.



Important Contact Information

Make sure that you scan the QR code using the camera on your smartphone and that your phone is connected to the internet. This will ensure that the contact information is added directly to your contacts and will also help you identify when your Care Team is calling you. If you don't have a smartphone, we encourage you to manually input and save the contact information on the left to your phone.

*Data rates may apply

