

GIVE IT ALL YOU'VE GOT

PATIENT GUIDE

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 28** of this brochure.

01 **A J&J withMe Care Coordinator should have contacted you** to discuss any insurance-related questions and work with you to get your medicine from the pharmacy. If you did not receive a call, please call a J&J withMe Care Coordinator at **866-228-3546**, Monday–Friday, 8 AM–8 PM ET.

02 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 J&J withMe 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.

03 **You should have received your medicine.** If it has not been delivered, please call your J&J withMe 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.



Scan QR code or [click here](#).

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

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PART 1: PAH AND UPTRAVI (selexipag)

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 adults and children 2 years of age and older with 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 younger than 2 years of age.

IMPORTANT SAFETY INFORMATION



- **Do not take UPTRAVI if you or your child** take gemfibrozil because this medicine may affect how UPTRAVI works and cause side effects
- **Do not take UPTRAVI if you or your child** are allergic to selexipag or any of the ingredients in UPTRAVI

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ABOUT PAH FOUNDATIONAL PATHWAYS

CONSIDERING EVERY FOUNDATIONAL PATHWAY IS KEY

Arteries in your lungs may be affected when there are imbalances of one or more naturally occurring substances in your body. These may include prostacyclin, endothelin, and/or nitric oxide. There are PAH treatments available that target each of these three foundational pathways.

Endothelin Pathway

Endothelin receptor antagonists (ERAs) work here.

Nitric Oxide Pathway

Phosphodiesterase type 5 inhibitors (PDE5is) 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 medicines (ERA, PDE5i, or both). UPTRAVI is available as oral tablets.

IMPORTANT SAFETY INFORMATION



- **Before you or your child 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 foundational pathways may play in PAH, hold your smartphone camera over the QR code.

Please see Important Safety Information throughout and on [pages 14–15](#).
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UPTRAVI (selexipag) CLINICAL STUDY

UPTRAVI WAS STUDIED IN THE LARGEST OUTCOMES TRIAL IN ADULTS 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 (PDE5i), 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 or your child 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

Please see Important Safety Information throughout and on [pages 14–15](#).
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ABOUT UPTRAVI (selexipag)

UPTRAVI is the only oral prostacyclin pathway therapy approved to: delay disease progression* and reduce the risk of PAH-related hospitalization. UPTRAVI can also improve the ability to exercise (as measured by the 6-minute walk distance).

Side effects occurring more frequently (by $\geq 3\%$) for adult patients on UPTRAVI compared to patients on placebo (sugar pill) are headache, diarrhea, jaw pain, nausea, muscle pain, vomiting, pain in arms or legs, temporary reddening of the skin (flushing), joint pain, low red blood cell count, less appetite than usual, and rash.

*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.

UPTRAVI IS THE ONLY ORAL PROSTACYCLIN PATHWAY THERAPY APPROVED TO:

 Delay disease progression*

&

 Reduce the risk of
PAH-related hospitalization

UPTRAVI can also improve the ability to exercise
(as measured by the 6-minute walk test).

IMPORTANT SAFETY INFORMATION

What are the possible side effects of UPTRAVI?



The most common side effects in adults and children are:

- Headache
- Nausea
- Diarrhea
- Muscle pain
- Jaw pain
- Vomiting
- Pain in arms or legs
- Flushing

Additional common side effects of UPTRAVI in adults include:

- Pain in joints
- Low red blood cell count
- Redness, pain or swelling at the injection site for UPTRAVI for injection
- Rash
- Decreased appetite

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Please [click here](#) for full Prescribing Information and Patient Product Information.

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

➤ 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).

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, PDE5i, OR BOTH).

IMPORTANT SAFETY INFORMATION

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



- Additional common side effects in children include stomach (abdominal) pain. Talk to your doctor if you or your child 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**.

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Please [click here](#) for full Prescribing Information and Patient Product Information.





PART 2: FINDING YOUR PERSONAL DOSE

SET YOUR GOALS

Now that you and your healthcare team have decided to add UPTRA VI (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 UPTRA VI (selexipag)

There isn't one specific dose of UPTRA VI 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
- UPTRA VI tablets typical dose strengths for adults range from 200 mcg to 1600 mcg
- Remember, each adult'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 UPTRA VI

IMPORTANT SAFETY INFORMATION

What are the possible side effects of UPTRA VI? (continued)



- ◀ Keep UPTRA VI and all other medicines away from children.

Please see Important Safety Information throughout and on [pages 14–15](#).
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PEOPLE MAY 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 adult 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 THE ADULT 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

Starting Dose: **200 mcg (taken twice daily)**



Decrease dose if current dose cannot be tolerated

Maintenance Dose: **Your personal dose (taken twice daily)**

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

IMPORTANT SAFETY INFORMATION

How should you or your child 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 or crush tablets. If you or your child cannot swallow tablets whole, see instructions located in the Patient Product Information for alternate options of taking UPTRAVI

Please see Important Safety Information throughout and on [pages 14–15](#).
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THE ADULT DOSE ADJUSTMENT PHASE

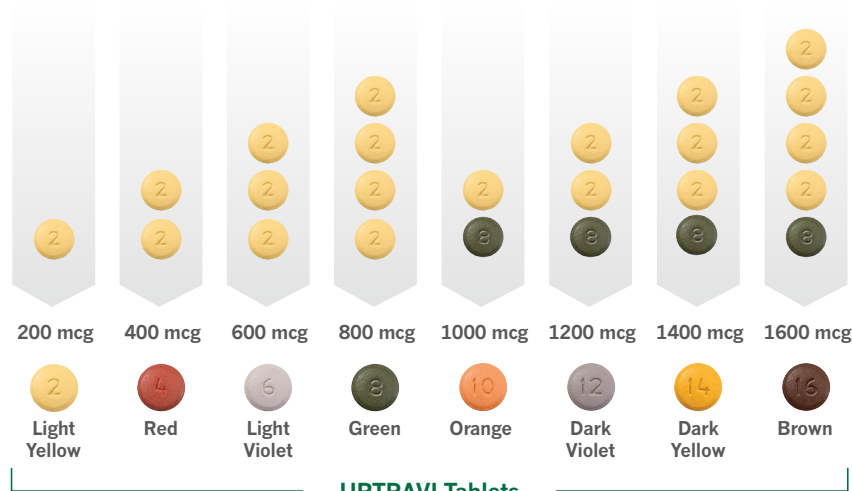
1 The Dose Adjustment Phase

You will receive a starting supply of **200-mcg and 800-mcg tablets**. During your dose-adjustment phase, your PAH healthcare team will instruct you to simply **add or remove a single 200-mcg tablet** to increase or decrease your dose as needed. **See the table on the right for the different tablet combinations at each dose.**

2 The Maintenance Phase

Once you have achieved your personal dose, your doctor will replace the 200-mcg and 800-mcg tablets you were taking during the dose adjustment phase with a single tablet.

There are 10 different tablet strengths of UPRAVI.*



UPRAVI Tablets

(Tablets are not actual size.)

*The 100 mcg and 150 mcg tablets are not shown.

IMPORTANT SAFETY INFORMATION

How should you or your child take UPRAVI?



UPRAVI Tablets

- Tell your doctor if you or your child have any form of liver disease. Your doctor may need to change your dose of UPRAVI
- UPRAVI is measured in micrograms (mcg). Tablets come in the following strengths: 100, 150, 200, 400, 600, 800, 1000, 1200, 1400, and 1600 mcg

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.



- ▶ **Do not change your dose or stop** taking UPTRAVI unless your doctor tells you to

- ▶ **Taking UPTRAVI 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

**Please see Important Safety Information throughout and on pages 14–15.
Please [click here](#) for full Prescribing Information and Patient Product Information.**

INDICATION AND IMPORTANT SAFETY INFORMATION

What is UPTRAVI (selexipag)?

UPTRAVI (selexipag) is a prescription medicine used to treat adults and children 2 years of age and older with 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 younger than 2 years of age.

Important Safety Information



- **Do not take UPTRAVI if you or your child** take gemfibrozil because this medicine may affect how UPTRAVI works and cause side effects
- **Do not take UPTRAVI if you or your child** are allergic to selexipag or any of the ingredients in UPTRAVI
- **Before you or your child 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 in adults and children are:

- | | | |
|------------|---------------|------------------------|
| • Headache | • Nausea | • Pain in arms or legs |
| • Diarrhea | • Muscle pain | • Flushing |
| • Jaw pain | • Vomiting | |

Additional common side effects of UPTRAVI in adults include:

- | | | |
|------------------|----------------------------|---|
| • Pain in joints | • Low red blood cell count | • Redness, pain or swelling at the injection site for UPTRAVI for injection |
| • Rash | • Decreased appetite | |

IMPORTANT SAFETY INFORMATION (continued)

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

Additional common side effects in children include stomach (abdominal) pain.

Talk to your doctor if you or your child 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 you or your child 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 or crush tablets. If you or your child cannot swallow tablets whole, see instructions located in the Patient Product Information for alternate options of taking UPTRAVI
- Tell your doctor if you or your child 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: 100, 150, 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

Please [click here](#) for full Prescribing Information and Patient Product Information.

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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 medicine 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 medicine) for promoting relaxation and pain relief
LEG PAIN	• Talk to your doctor about screening for an iron deficiency or medicine 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 over-the-counter pain medicine 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 medicine 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 medicine 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



PART 3: STOP AT NOTHING

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.

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

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

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

Scan QR code or
[click here.](#)



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 Johnson & Johnson-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.

Information about your insurance coverage, cost support options, and treatment support is given to you by service providers for J&J withMe.

The information you get does not require you to use any medicine by Johnson & Johnson. 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.

**Please see Important Safety Information throughout and on [pages 14–15](#).
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FINDING SUPPORT

SUPPORT FOR PATIENTS AND THEIR CARE PARTNERS

Once you and your doctor decide that UPTRAVI is right for you, J&J withMe provides the additional support you may need to help you get started with UPTRAVI (selexipag) treatment.

Your personally assigned J&J withMe 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 medicine by Johnson & Johnson
- **Visit JNJwithMe.com**
- **Need Help?** Call a J&J withMe Care Coordinator at **866-228-3546**, Monday–Friday, 8 AM–8 PM ET. Multilingual phone support is available

PAH Companion withMe

Personalized Support for You

Everyone faces PAH differently. Once you and your doctor have decided that a PAH medicine by Johnson & Johnson is right for you, you can join PAH Companion withMe — a part of the J&J withMe suite of patient support resources personalized just for you.

You'll receive one-on-one educational support to help with your prescribed PAH medicine by Johnson & Johnson, 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 PAH medicine by Johnson & Johnson, 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



PAH Companion is available for adult patients ≥ 18 years of age. The support and resources provided by PAH Companion withMe and J&J withMe are not intended to provide medical advice, replace a treatment plan you receive from your doctor or nurse, or serve as a reason for you to start or stay on treatment.

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 PAH medicine by Johnson & Johnson, 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.

PAH Companion is available for adult patients ≥ 18 years of age.

STEPS TO EXPECT

Now that you've received your medicine, there are a couple of additional PAH resources Johnson & Johnson 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 J&J withMe Care Coordinator

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

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

The support and resources provided by PAH Companion withMe and J&J withMe are not intended to provide medical advice, replace a treatment plan you receive from your doctor or nurse, or serve as a reason for you to start or stay on treatment.

Scan QR code
or [click here](#).



**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.



PEDIATRIC DOSING

ABOUT UPTRAVI (selexipag)

UPTRAVI is indicated for the treatment of PAH (WHO Group I) in pediatric patients aged two years and older. UPTRAVI reduces NT-proBNP and is expected to slow down disease progression and lower your risk of being hospitalized for PAH.

WHAT IS A BNP TEST?

A B-type Natriuretic Peptide (BNP) test measures the amount of BNP in your body. NT-proBNP develops into BNP inside of your body. BNP is a hormone that increases when blood pressure or fluid volume in the lung's blood vessels is high. High BNP can mean that the heart is working harder to manage increased fluid volume and blood pressure.

IMPORTANT SAFETY INFORMATION

What is UPTRAVI (selexipag)?

UPTRAVI (selexipag) is a prescription medicine used to treat adults and children 2 years of age and older with 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 younger than 2 years of age.

**Please see Important Safety Information throughout and on pages 14–15.
Please [click here](#) for full Prescribing Information and Patient Product Information.**

THE PEDIATRIC DOSE ADJUSTMENT PHASE

When starting UPTRA^{VI} (selexipag), your child’s PAH healthcare team will slowly adjust your child’s dose until they reach their personal maintenance dose.

Starting Dose: **Weight dependent**



Slowly increase dose to reach your child’s personal dose, taken twice daily

Maintenance Dose: **Your child’s personal dose**



Decrease dose if current dose cannot be tolerated, taken twice daily

Body weight (lb)*	Recommended starting dose, twice daily	Recommended weekly titration increments, twice daily	Maximum dose allowed, twice daily
19 lb to less than 55 lb	100 mcg	100 mcg	800 mcg
55 lb to less than 88 lb	150 mcg	150 mcg	1200 mcg
88 lb to less than 110 lb	150 mcg	150 mcg	1600 mcg
110 lb and up	200 mcg	200 mcg	1600 mcg

➤ UPTRA^{VI} dose strengths range from 100 mcg to 1600 mcg

Before you or your child take UPTRA^{VI}, tell your healthcare provider if the patient has any liver problems. Once-daily dosing is recommended in patients with moderate liver problems. UPTRA^{VI} should be avoided in patients with severe liver problems.

**Patient bodyweight is an approximate conversion of kilograms to pounds*

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

DISCOVERING YOUR CHILD'S PERSONAL DOSE

1 The Dose Adjustment Phase

Depending on your child's body weight, you will receive a starting supply of **100 mcg, 150 mcg or 200 mcg tablets**. During the dose-adjustment phase, your child's PAH healthcare team will instruct you on how to increase or decrease the child's dose as needed.

UPTRAVI comes in different strengths. Be sure to follow your doctor's instructions closely so you take the right dose as it is adjusted.

2 The Maintenance Phase

During the dose adjustment phase, the doctor may prescribe a combination of multiple tablet strengths to achieve your child's personal dose. Depending on the personal dose achieved, some patients may be switched to a single tablet that matches their personal dose, while other patients may need to continue taking multiple tablets or a combination of different tablet strengths to maintain their personal dose.

WHAT TO DO IF A DOSE IS MISSED?

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

IMPORTANT SAFETY INFORMATION



- **Do not take UPTRAVI if you or your child** take gemfibrozil because this medicine may affect how UPTRAVI works and cause side effects
- **Do not take UPTRAVI if you or your child** are allergic to selexipag or any of the ingredients in UPTRAVI

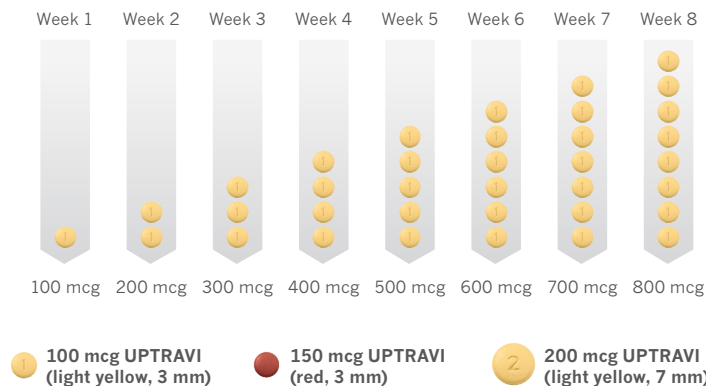
Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

DISCOVERING YOUR CHILD'S PERSONAL DOSE (APPROXIMATELY 19 LB TO LESS THAN 55 LB*)

CHILDREN THAT CANNOT SWALLOW TABLETS WHOLE

For children who cannot swallow the tablets whole, 100 mcg UPTRAVI tablets can be dissolved and administered in apple or orange juice. (**Do not** use water or milk to dissolve the tablets, see full instructions for dissolving tablets on page 35.)

Alternatively, 100-mcg UPTRAVI tablets can also be administered with soft foods such as yogurt, applesauce or mashed banana.

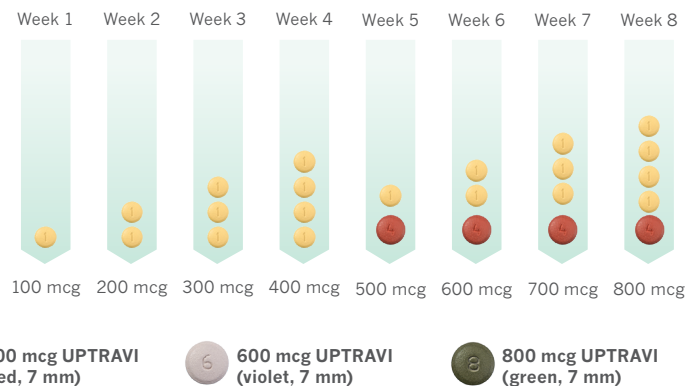


Example of dose adjustment. Actual schedule may vary.

*Approximate from conversion of kilograms to pounds.

CHILDREN CAPABLE OF SWALLOWING TABLETS

For children capable of swallowing tablets ≥ 200 mcg (7 mm), they should swallow the UPTRAVI tablets whole. Do not split or crush the tablets.



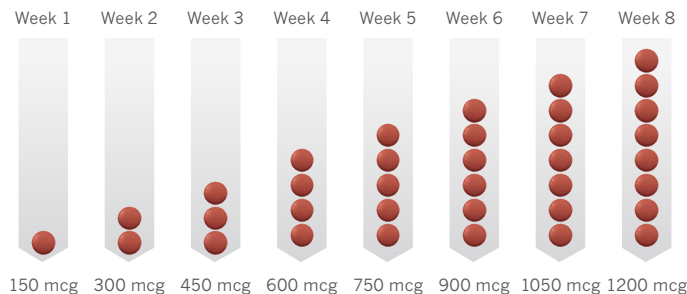
Please see Important Safety Information throughout and on pages 14–15.
Please [click here](#) for full Prescribing Information and Patient Product Information.

DISCOVERING YOUR CHILD'S PERSONAL DOSE (APPROXIMATELY 55 LB TO LESS THAN 110 LB*)

CHILDREN THAT CANNOT SWALLOW TABLETS WHOLE

For children who cannot swallow the tablets whole, 100 mcg UPTRAVI tablets can be dissolved and administered in apple or orange juice. (**Do not** use water or milk to dissolve the tablets, see full instructions for dissolving tablets on page 35.)

Alternatively, 150-mcg UPTRAVI tablets can also be administered with soft foods such as yogurt, applesauce or mashed banana.



1 100 mcg UPTRAVI
(light yellow, 3 mm)

150 mcg UPTRAVI
(red, 3 mm)

2 200 mcg UPTRAVI
(light yellow, 7 mm)

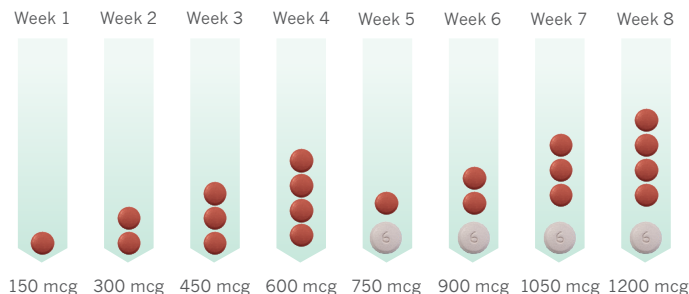
4 400 mcg UPTRAVI
(red, 7 mm)

6 600 mcg UPTRAVI
(violet, 7 mm)

8 800 mcg UPTRAVI
(green, 7 mm)

CHILDREN CAPABLE OF SWALLOWING TABLETS

For children capable of swallowing tablets ≥ 200 mcg (7 mm), they should swallow the UPTRAVI tablets whole. Do not split or crush the tablets.



Patients weighing 88 lbs to less than 110 lbs can continue titrating to a maximum dose of 1600 mcg. Example of dose adjustment. Actual schedule may vary.

*Approximate from conversion of kilograms to pounds.

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

DISCOVERING YOUR CHILD'S PERSONAL DOSE (APPROXIMATELY 110 LB AND UP*)

1 The Dose Adjustment Phase

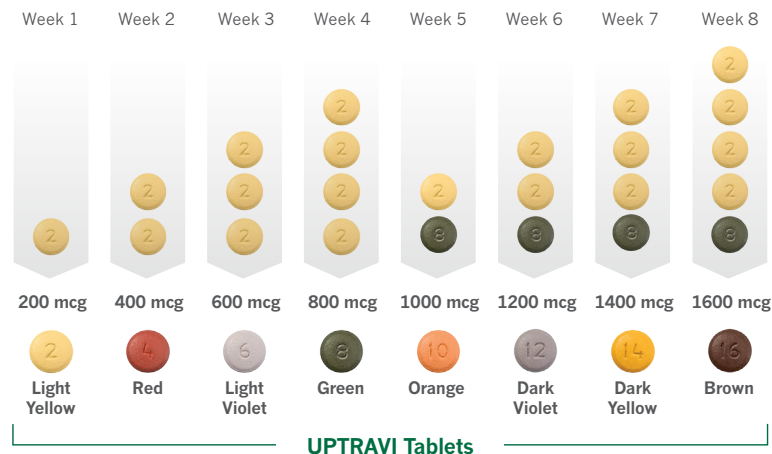
Your child will receive a starting supply of **200-mcg and 800-mcg tablets**. During your child's dose adjustment phase, your child's PAH healthcare team will instruct you and your child to simply **add or remove a single 200-mcg tablet** to increase or decrease your dose as needed. **See the table on the right for the different tablet combinations at each dose.**

2 The Maintenance Phase

Once your child has achieved their personal dose, your child's doctor will replace the 200-mcg and 800-mcg tablets your child was taking during the dose adjustment phase with a single tablet. **There are 10 different tablet strengths of UPTRAVI.**

Example of dose adjustment. Actual schedule may vary.

**Approximate from conversion of kilograms to pounds.*



(Tablets are not actual size.) The 100-mcg and 150-mcg tablets are not shown.

IMPORTANT SAFETY INFORMATION



- **Before you or your child 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

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.



Please see Important Safety Information throughout and on [pages 14–15](#).
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SUGGESTIONS FOR DISCUSSING SIDE EFFECTS WITH YOUR DOCTOR

Stomach-Related Side Effects

Compared to adults, a higher frequency of vomiting and abdominal pain was observed in pediatric patients on UPTRAVI. UPTRAVI can be taken with or without food. Taking UPTRAVI with food may help your child tolerate UPTRAVI better. If your child experiences side effects, speak with their healthcare provider about options for side effect management.

Some patients taking UPTRAVI may experience decreased appetite.

Weight and height gain that may be lower than expected:

Your healthcare provider will prescribe your child's UPTRAVI starting dose based on their body weight.

Monitor your child's growth, if you notice that they are not gaining weight as expected, talk with your child's healthcare provider.

These are not all the possible side effects of UPTRAVI. Talk to your doctor for medical advice about side effects, even if they do not seem serious.

IMPORTANT SAFETY INFORMATION



- **Before you or your child take UPTRAVI, tell your healthcare provider about all your medical conditions, including if you: (continued)**

- 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

**Please see Important Safety Information throughout and on pages 14–15.
Please [click here](#) for full Prescribing Information and Patient Product Information.**



Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

INSTRUCTIONS FOR DISSOLVING

▶ If your child cannot swallow the tablets:

- Dissolve the 100-mcg or 150-mcg tablet(s) in apple or orange juice. **Do not** dissolve the tablet(s) in water or milk.

▶ Place the required number of tablet(s) per prescribed dose in a cup. **Do not crush or split the tablet(s).**

▶ Add 1 or 2 teaspoons (at least 1 mL per tablet) of apple or orange juice to the prescribed number of tablets.

▶ Wait for 5 minutes, then stir the juice and the tablet(s) until dissolved.

▶ Administer the mixture immediately.

- Alternative administration with soft food: Cover the required number of 100-mcg or 150-mcg tablets per prescribed dose with a small amount of soft food, such as yogurt, applesauce or mashed banana. Administer the mixture immediately. Make sure no remaining medicine is left in the container by adding more soft food and swallowing mixture immediately.

▶ **Do not** store UPTRAVI tablets that are already mixed with juice or soft food for later use.

▶ If you are using a combination of different tablet strengths, only the 100-mcg and 150-mcg tablets can be dissolved. The 200-mcg to 1600-mcg tablets are not designed to be dissolved.

IMPORTANT SAFETY INFORMATION

What are the possible side effects of UPTRAVI?



The most common side effects in adults and children are:

- | | | |
|------------|---------------|------------------------|
| • Headache | • Nausea | • Pain in arms or legs |
| • Diarrhea | • Muscle pain | • Flushing |
| • Jaw pain | • Vomiting | |

Additional common side effects of UPTRAVI in adults include:

- | | | |
|------------------|----------------------------|---|
| • Pain in joints | • Low red blood cell count | • Redness, pain or swelling at the injection site for UPTRAVI for injection |
| • Rash | • Decreased appetite | |

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.

WHAT TO EXPECT

There is additional support from Johnson & Johnson to help you continue treatment.

A Benefits Investigation Call from Your J&J withMe Care Coordinator

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

A J&J withMe Care Coordinator will call you from this number:

866-228-3546, Monday–Friday, 8 AM–8 PM ET

The support and resources provided by PAH Companion withMe and J&J withMe are not intended to provide medical advice, replace a treatment plan you receive from your doctor or nurse, or serve as a reason for you to start or stay on treatment.

Please see Important Safety Information throughout and on [pages 14–15](#).
Please [click here](#) for full Prescribing Information and Patient Product Information.



