Selixipag (Uptravi)
Updated June 2023

Selixipag is an oral medication to treat pulmonary arterial hypertension (PAH), also known as Group 1 pulmonary hypertension. The goal of this therapy is to delay disease progression and reduce the risk of hospitalization.

The Food and Drug Administration approved Uptravi to treat PAH in 2015.

**How selixipag works**
Selixipag is an oral prostacyclin IP receptor agonist that is structurally unique from prostacyclin. It is converted to the active form of the drug in the liver, which is 37 times more potent than the parent molecule (compound). Selixipag and its metabolite selectively bind to the IP receptor which works to widen the blood vessels in the lungs which helps reduce the blood pressure in the lungs.

**Dosing and administration**
Dosing is adjusted incrementally to determine the most effective dosing for you without adverse effects. Selixipag is available in dosages ranging from 200 microgram to 1600 mcg.

- The tablet is taken twice a day, once in the morning and once in the evening.
- Take the tablet with food for better toleration.
- Swallow tablets whole. Do not split, crush or chew.
- Your doctor will start you at the lowest dose — one 200 mcg tablet — and slowly increase the dosage to find what works best for you. Dosage is generally increased 200 mcg weekly.
- Long-term (maintenance) dosage is determined by how well you tolerate side-effects. Your doctor will increase the dose until side effects are intolerable.

Selixipag comes in 60-tablet bottles in the following doses: 200 mcg, 400 mcg, 600 mcg, 800 mcg, 1,000 mcg, 1,200 mcg, 1,400 mcg or 1,600 mcg.

Selixipag also comes in a dose titration pack that includes a 140-count bottle of 200 mcg tablets and a 60-count bottle of 800 mcg tablets.

**Note:** If you can't eat or drink for any reason, your doctor should have an alternative treatment plan ready for periods when you can't ingest anything by mouth.

**How selixipag is supplied**
You’ll need a prescription from your pulmonary hypertension specialist. The prescribing doctor must complete a patient enrollment form to obtain prior authorization through insurance.

**Insurance coverage**

https://phassociation.org/patients/treatments/selexipag/
Most health insurance plans will pay part of the cost of this medication. However, some plans leave patients with high out-of-pocket costs. Ask your insurance company for more accurate information about copays or out-of-pocket costs. Depending on your insurance plan, you may be eligible for assistance from the company that manufactures the drug or copay assistance from a non-profit charitable organization. See the list [www.PHAssociation.org/Help](https://www.PHAssociation.org/Help) or call 301-565-3004.

**Common side effects**

- Headache
- Diarrhea.
- Jaw pain.
- Nausea.
- Muscle pain.
- Vomiting.
- Pain in arms and legs.
- Flushing.
- Joint pain.
- Anemia.
- Decreased appetite.
- Rash.

**Special considerations**

**Pregnancy and breastfeeding:** There are no adequate and well-controlled studies in pregnant women. However, in most circumstances, women shouldn’t become pregnant while being treated for PAH. In animal studies, maternal and fetal weights fell slightly when pregnant rats received 47 times more than the maximum human dose during the stage of pregnancy when tissues develop into organs. However, there was no evidence of other abnormalities. No developmental abnormalities were seen when pregnant rabbits were given selexipag at 50 times higher than the maximum human dose.

Selexipag has been detected in milk from lactating rats. Although it's unknown whether it shows up in human milk, many drugs pass to milk. Nursing mothers are advised to discontinue either breastfeeding or selexipag because of possible adverse effects.

**Children:** Safety and effectiveness for pediatric patients is unknown.

**Older adults:** No significant differences have been seen or reported in older patients compared to younger patients, but a greater sensitivity to the medication can’t be ruled out.

**Liver disease:** No dose adjustment is required for people with mild liver impairment (Child-Pugh Class A). Selexipag should be taken once daily for people with moderate liver impairment (Child-Pugh Class B). People with
severe liver impairment (Child-Pugh Class C) shouldn't take selexipag.

**Kidney disease:** No dose adjustment is required in patients with a glomerular filtration rate (GFR) > 15 ml/min/1.73 m². There is no clinical experience in patients with a GFR < 15 ml/min/1.73 m² or who are undergoing dialysis.3

**Drug Interactions**

Avoid concurrent use of strong CYP2C8 inhibitors, such as gemfibrozil, which can increase exposure to selexipag and its active metabolite.

No dose adjustments are recommended for other PAH medications, warfarin or lopinavir/ritonavir.

**Allergies:** Possible but unlikely.