

Treprostinil (Tyvaso[®], Yutrepia[®]) Inhalation Solution and Treprostinil Dry Powder Inhaler (DPI) (U.S. Only)



pulmonaryfibrosis.org

What is treprostinil?

Treprostinil is an inhaled medication approved for the treatment of pulmonary hypertension (PH) related to interstitial lung disease (ILD) by the U.S. FDA in 2022. Pulmonary hypertension (PH) is abnormal high blood pressure in the pulmonary arteries, which connect the heart to the lungs. Interstitial lung diseases (also known as ILD), include all of the diseases that have inflammation and/or scarring in the air sacs and the tissue surrounding the air sacs in the lungs. Treprostinil has been shown to improve exercise capacity in PH related to ILD in a clinical trial. It is also used in oral and intravenous forms in PH that is not related to other lung diseases.

How does treprostinil work?

Treprostinil is a pulmonary vasodilator, meaning that it works by relaxing the blood vessels that carry blood cells through the lungs. This lowers the high pressure, or hypertension, in the pulmonary blood vessels that occurs with PH.

Who should take treprostinil?

Treprostinil is approved for people with ILD who also have PH. In most cases, a right heart catheterization is needed to diagnose PH and measure its severity.

Only you and your physician can determine the best approach to treating your condition, and if you have PH related to ILD, we encourage you to consult with your physician to determine whether this drug is appropriate for you.

How should treprostinil be taken?

Inhaled treprostinil can be administered by nebulized solution or by dry powder inhaler (DPI). Treprostinil inhalation solution is taken by inhaling the medication using a special nebulizer device that is supplied by the manufacturer. It cannot be used with any other type of nebulizers such as mesh, jet or ultrasonic. Treprostinil DPI is taken by inserting a cartridge into a special inhaler device and inhaling the medication through the mouth into the lungs. The medication is inhaled four times per day with doses at least four hours apart for both devices.

When starting either form of inhaled treprostinil, you will start with a low dosage that is increased every 1-2 weeks until a target maintenance dose (which is determined by your doctor) is reached. For nebulized treprostinil, the dose is increased by adding to the number of breaths taken via nebulizer with each dose. For treprostinil DPI, the dose is increased by changing to a higher dose cartridge strength in the inhaler.

How can I obtain treprostinil?

Treprostinil is only available by prescription through a specialty pharmacy. A prior authorization is required. Once approved, the specialty pharmacy will arrange for delivery of the medication and device, and a nurse from the specialty pharmacy will come to your home or doctor's office to show you how to take the medication.

Will my insurance pay for treprostinil?

Insurance coverage will vary by insurer and your individual policy. We recommend that you contact your insurance provider directly to determine your specific coverage. The manufacturer of treprostinil

has set up programs to help patients afford their medications. You can contact **ASSIST Access Solutions** at 1-877-864-8437 or enroll in the **Tyvaso Co-Pay Assistance Program**. Contact the PFF Help Center at **844.TalkPFF** (844.825.5733) for information about other organizations that may be able to assist with the cost of your medication

What side effects can occur from treprostinil?

Common side effects of inhaled treprostinil include cough, headache, throat irritation and pain, nausea, flushing of the face and neck, fainting and loss of consciousness, dizziness, diarrhea, and shortness of breath. Eating or drinking something to coat the throat or using a numbing throat spray prior to taking your medicine can help with throat irritation. An over the counter pain medication, such as acetaminophen, can help with headache.

Serious side effects can include low blood pressure, particularly if you have low blood pressure prior to taking the medication; bleeding problems, especially in people taking blood thinners; and wheezing or bronchospasm.

Talk to your doctor if any of these side effects occur.

How will my doctor monitor me while I am taking treprostinil?

Your doctor will monitor you for side effects, especially while you are first taking treprostinil at increasing doses. If you have side effects, your doctor might add additional medications, lower your dose, or temporarily or permanently stop treprostinil.

What drug interactions should I know about?

Diuretic medications and those used to treat high blood pressure may increase the risk of low blood pressure if taken with treprostinil. Anticoagulants, or medications that decrease blood clotting, may increase risk for bleeding. Gemfibrozil, a medication used to treat high cholesterol, and rifampin, an antibiotic, can also interact with treprostinil.

It is important to update your physician and pharmacy about any medication changes or additions including all prescriptions, over the counter medications, vitamins, and herbal supplements.

Are there some people who should not take treprostinil?

The safety of treprostinil in pregnancy and breastfeeding has not been established.

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