What is pirfenidone?
Pirfenidone is an oral medication approved for the treatment of idiopathic pulmonary fibrosis (IPF) by the U.S. FDA in 2014. Pirfenidone has been shown to slow the progression of IPF in clinical trials.

How does pirfenidone work?
Pirfenidone slows down the buildup of scar tissue (fibrosis) in the lung by interfering with the ability of cells to make scar tissue. Pirfenidone does not improve the function of the lungs. Instead, the disease progresses more slowly over time.

Who should take pirfenidone?
Pirfenidone has been recommended by professional medical societies for use in adults with IPF. These recommendations state that a patient’s preferences should be incorporated into the decision to start taking pirfenidone and that some patients may decide not to take medication after consideration of the potential benefits and adverse reactions.

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

How should pirfenidone be taken?
Pirfenidone comes in 267mg capsules, 267mg tablets, and 801mg tablets. The FDA approved dose is 801mg by mouth three times a day.

When starting pirfenidone, the initial dose is 267mg (one capsule or tablet) three times a day. If tolerated, the dose is increased to 534mg (two capsules or tablets) three times a day after one week. If that dose is tolerated, the dose is increased to 801mg (three capsules or tablets) three times a day. After a while, the 801mg tablet can be used instead of using three 267mg capsules or tablets.

It is important to take pirfenidone with full meals.

How can I obtain pirfenidone?
Pirfenidone is only available by prescription through a Specialty Pharmacy. A Prior Authorization is required. Once approved, the Specialty Pharmacy will schedule mail delivery of pirfenidone to your home.

Will my insurance pay for pirfenidone?
Insurance coverage will vary by insurer and your individual policy. We recommend that you directly contact your insurance provider to determine your specific coverage. The supplier of pirfenidone (Esbriet®) has set up programs to help patients afford their medications. You can contact Esbriet Access Solutions at (866) 422-2377 and the Esbriet Co-pay Program at 844-693-7274. We recommend that you contact these programs directly as they will assist you in working with your insurance company as well.

There are also several organizations that can help those living with pulmonary fibrosis with financial and insurance issues. You can contact Patient Services Inc. at 1-800-366-7741, the HealthWell Foundation at 1-800-675-8416, or the Patient Advocate Foundation at 1-800-532-5274 to find out more about how they can provide assistance.
**What side effects can occur from pirfenidone?**
Common side effects include nausea, rash, abdominal pain, diarrhea, fatigue, headache, upset stomach, dizziness, vomiting, loss of appetite, heartburn (acid reflux), weight loss and joint pain. The gastrointestinal symptoms can often be reduced by taking pirfenidone with food, following the dosing schedule carefully, and taking additional medications recommended by your doctor (such as antacids and anti-nausea medication).

A skin rash in reaction to the sun (“photosensitivity”) can also occur. This reaction can be minimized or avoided by using sunblock every day (SPF 50 or greater), wearing a hat with a brim, wearing pants and shirts with long sleeves, and avoiding direct and indirect sunlight and sunlamps.

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

Elevations in blood tests for liver function can occur. In the post-marketing period, non-serious and serious cases of drug-induced liver injury, including severe liver injury with fatal outcome, have been reported. Talk to your doctor if you experience symptoms like fatigue, anorexia, right upper abdominal discomfort, dark urine, or jaundice. Low white blood cell counts and an allergic reaction called “angioedema” have been reported to the FDA following approval of the medication.

**How will my doctor monitor me while I am taking pirfenidone?**
You doctor will monitor you for side effects and will perform blood tests to check your liver function monthly for the first 6 months and every 3 months after that. If you have side effects, your doctor might add additional medications, lower your dose, or temporarily or permanently stop pirfenidone.

**What drug interactions should I know about?**
Fluvoxamine (an anti-depressant) and high doses of ciprofloxacin (an antibiotic) can increase levels of pirfenidone in your body.

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

**Are there some people who should not take pirfenidone?**
In general, women who are pregnant or breastfeeding should not take pirfenidone. Pirfenidone may not be appropriate for some people with kidney or liver problems. Smoking can lower blood levels of pirfenidone. If you smoke, you should talk to your doctor about how they can help you quit.

---

**Acknowledgements**
The Pulmonary Fibrosis Foundation is thankful to the following for their assistance in writing and reviewing this fact sheet:

David J. Lederer, MD, MS
Former PFF Senior Medical Advisor, Education and Awareness

Kevin R. Flaherty, MD, MS
University of Michigan Health System
Ann Arbor, MI
Steering Committee Chair, PFF Care Center Network and PFF Patient Registry