

## Raising awareness of covert killer: pulmonary fibrosis

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Despite claiming an estimated 40,000 lives annually - roughly the same number as breast cancer or prostate cancer - pulmonary fibrosis remains one of the least known covert killers in the country.

"Survivors of [breast cancer](#) come together in droves to raise awareness," said Teresa Barnes, vice president of patient outreach and program support for the Coalition for Pulmonary Fibrosis, based in Culver City, Calif. "Pulmonary fibrosis leaves no survivors," except a relative handful who receive [lung transplants](#).

,Pulmonary fibrosis causes progressive lung scarring and eventually suffocation.

Hope is on the horizon. The first treatment options available, InterMune's pirfenidone and Boehringer Ingelheim's nintedanib, are being evaluated by the Food and Drug Administration and may become available as early as this fall.

The disease has an annual fundraising walk/run in the summer in Pittsburgh organized by Tami Rippy, whose mother died of the disease in March 2009. "Knowing she was suffering, and literally suffocating, that there was no treatment, no idea where it came from or why it happened makes it devastating," said Rippy. "People assume my mom got it because she did this or that or smoked, but she was healthy all these years."

The terminal diagnosis often disguises itself in generic symptoms, such as dry cough and fatigue. It typically affects people over 50 but does not discriminate, debilitating runners and smokers alike. The overwhelming majority of cases are idiopathic, meaning no known cause is ever identified and the condition is known as idiopathic [pulmonary fibrosis](#) or IPF.

According to the coalition, pulmonary fibrosis patients may have an exaggerated or uncontrolled healing response that, over time, produces excessive fibrous scar tissue - or fibrosis - in the lungs. It's not known what sets this abnormal tissue-repair process in motion.

"This is worse than a cancer diagnosis," said Kevin Gibson, clinical director of a center at the University of Pittsburgh Medical center that specializes in lung disease. "With cancer you're offered treatment, but with IPF there is no treatment apart from transplant." Fewer than 1 percent of these patients receive them.

The two drugs are considered breakthrough therapies, meaning the FDA will expedite the process of their development and review. The drugs work by trying to reduce the onset and progression of the disorder, which also destroys blood vessels, diminishing the ability to circulate oxygen in the body.

"If they're approved it's great, but the treatment's effects are modest at best," said Dr. Gibson. "It is still a long way until a cure."

Although the condition has been recognized for the past century, relatively little is known about pulmonary fibrosis, the "disease course being highly variable and unpredictable," he said.

Despite being five times as prevalent as [cystic fibrosis](#), which receives about \$85 million in federal funding annually, the condition receives

only \$18 million a year, according to the Coalition for Pulmonary Fibrosis.

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