

Clinical Trial Results Signal an Advance for Idiopathic Pulmonary Fibrosis

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Pulmonary Fibrosis Medications Approved

The U.S. Food and Drug Administration has approved two medications for treatment of idiopathic pulmonary fibrosis. Pirfenidone (Esbriet) and nintedanib (Ofev) are the first medications approved in the United States for treatment of the disease.

Denver, CO — After a string of failed clinical trials in [idiopathic pulmonary fibrosis](#), the results from studies of two experimental medications signal progress in the treatment of this deadly disease, according to researchers at National Jewish Health and around the nation. The trial results were presented May 18 at the 2014 American Thoracic Society annual conference and simultaneously published online in the *New England Journal of Medicine*.

“We are taking measurable steps forward,” said [Kevin K. Brown, MD](#), vice chair of Medicine at National Jewish Health and co-author of one of the trials. National Jewish Health has one of the largest programs for the treatment and study of idiopathic pulmonary fibrosis. Dr. Brown, his colleagues and their patients have participated in dozens of trials over the course of almost two decades.

Idiopathic pulmonary fibrosis (IPF) is a progressive, fatal scarring of the lungs. Researchers do not understand what causes IPF. There is no approved medication available in the United States, and mean survival of patients with IPF is three to five years after diagnosis, worse than for many cancers. Approximately 50,000 Americans die of IPF every year.

National Jewish Health researchers and their patients began participating in large, multi-center trials of experimental medications to treat IPF in the mid-1990s. After a long string of failures, this recent success demonstrates how persistence and flexibility in thinking about the disease has helped guide researchers to more effective therapies.

Early trials were aimed at reducing chronic inflammation, which was thought to cause scarring. Then in the mid-2000s, thinking about IPF shifted following a series of papers that carefully described its clinical features, its natural history and the characteristics associated with early death. Instead of chronic inflammation, physicians and scientists began to see the progressive fibrosis as the primary therapeutic target.

In an adult, normal wound healing requires the knitting together of damaged tissue, which involves some fibrosis or scarring. Once the wound heals, a small, fixed area of scar tissue is left. In IPF, this process seems to go awry. Scar tissue proliferates and expands, replacing normal lung tissue and reducing patients’ capacity to absorb oxygen.

In recent years, researchers developed medications aimed directly at slowing the abnormal proliferation of scar tissue in IPF patients. The recent trials demonstrate the results of that strategy.

Separate trials of two medications, nintedanib and pirfenidone each slowed the rapid loss of lung function that is seen in patients with IPF. Nintedanib slowed the loss of lung capacity by about 50% when compared with placebo during a 52-week trial, with similar findings in the pirfenidone study. There were fewer deaths during the studies in those taking either nintedanib or pirfenidone, although the differences were not considered statistically significant.

A third trial, of N-acetylcysteine, did not appear to have a major benefit, though additional study was recommended.

“With the steadfast commitment of the entire pulmonary fibrosis community — the patients, their families, the physicians, researchers and funding organizations — we are seeing measurable progress, progress that keeps the search for a cure alive,” said Dr. Brown.

National Jewish Health is the leading respiratory hospital in the nation. Founded 125 years ago as a nonprofit hospital, National Jewish Health today is the only facility in the world dedicated exclusively to groundbreaking medical research and treatment of children and adults with respiratory, cardiac, immune and related disorders. Patients and families come to National Jewish Health from around the world to receive cutting-edge, comprehensive, coordinated care. To learn more, visit the [media resources](#) page.

Media Resources

We have many faculty members, from bench scientists to clinicians, who can speak on almost any aspect of respiratory, immune, cardiac and gastrointestinal disease as well as lung cancer and basic immunology.

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Media Contacts

Our team is available to arrange interviews, discuss events and story ideas.

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