

Ofev[®] (nintedanib) – New orphan indication

- On September 6, 2019, the [FDA announced](#) the approval of Boehringer Ingelheim's [Ofev \(nintedanib\)](#), to slow the rate of decline in pulmonary function in patients with systemic sclerosis-associated interstitial lung disease (SSc-ILD)
- Ofev is also approved for the treatment of idiopathic pulmonary fibrosis (IPF).
- Scleroderma is a rare disease that causes tissue throughout the body, including the lungs and other organs, to thicken and scar. Interstitial lung disease or ILD is a condition affecting the interstitium, which is part of the lung's structure, and is one of the most common disease manifestations of scleroderma.
 - ILD is the leading cause of death among people with scleroderma, typically resulting from a loss of pulmonary function that occurs when the lungs cannot supply enough oxygen to the heart.
 - Approximately 100,000 individuals in the U.S. have scleroderma, and approximately half of scleroderma patients have SSc-ILD.
- The approval of Ofev for this new indication was based on a double-blind study in patients with SSc-ILD. A total of 580 patients were randomized to receive either Ofev or placebo for at least 52 weeks, of which 576 patients were treated. The primary endpoint was the annual rate of decline in Forced Vital Capacity (FVC) over 52 weeks. The absolute change from baseline in the modified Rodnan skin score (mRSS) at week 52 was a key secondary endpoint. Mortality over the whole trial was an additional secondary endpoint.
 - The annual rate of decline of FVC (in mL) over 52 weeks was significantly reduced by 41 mL (95% CI: 3, 79) in patients receiving Ofev vs. patients receiving placebo, corresponding to a relative treatment effect of 44%.
 - No benefit in mRSS was observed in patients receiving Ofev.
 - No difference in survival was observed in an exploratory analysis of mortality over the whole trial (Ofev: 3.5% vs. placebo: 3.1%).
- The recommended dose of Ofev for the treatment of IPF and SSc-ILD is 150 mg orally twice daily administered approximately 12 hours apart.
 - Prior to treatment initiation, liver function tests should be conducted in all patients and a pregnancy test in females of reproductive potential.