NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health Technology Evaluation

Nerandomilast for treating idiopathic pulmonary fibrosis or progressive pulmonary fibrosis

Final scope

Remit/evaluation objective

To appraise the clinical and cost effectiveness of nerandomilast within its marketing authorisation for treating idiopathic pulmonary fibrosis or progressive pulmonary fibrosis.

Background

Interstitial lung disease (ILD) is an overarching term used to describe a large group of many different and often rare disorders that cause inflammation or scarring (fibrosis) of the functional lung tissue. Some ILDs progress and become fibrotic and are referred to as progressive fibrosing ILD (PF-ILD).

Idiopathic pulmonary fibrosis (IPF) and progressive pulmonary fibrosis (PPF) are both forms of PF-ILD and occur primarily in older adults.[1] IPF is a specific form of chronic PF-ILD of unknown cause and associated with a pattern of usual interstitial pneumonia. PPF occurs in cases of PF-ILD other than IPF and is defined by worsening respiratory symptoms and evidence of disease progression.[2] It is defined based on clinical symptoms, lung function and chest imaging, regardless of the underlying condition.[3] IPF and PPF are difficult diseases to diagnose and require a multidisciplinary team.

IPF and PPF have similar pathogenetic mechanisms and disease behaviours.[3] The most common symptoms are breathlessness (which may initially be only on exertion), cough, and fatigue. Over time, these symptoms are associated with a decline in lung function, reduced quality of life, disability and shortened life expectancy. It is estimated that around 3,500 people die of IPF every year and the survival of people with PPF is similar to that of IPF.[4,5]

In the UK around 32,500 people are living with IPF, a prevalence of around 50 per 100,000.[6] There are around 6,000 new cases of IPF diagnosed every year.[6] The prevalence of PPF is difficult to measure as data are usually for specific conditions. PPF can be caused by a variety of conditions, including sarcoidosis, systemic autoimmune rheumatic disease (SARD-ILD), hypersensitivity pneumonitis and asbestosis.

Established clinical management for IPF includes treatment with antifibrotic therapy alongside pulmonary rehabilitation and cessation of smoking and drugs associated with pulmonary toxicity.[7] For PPF, established clinical management may include antifibrotic therapy, immunosuppresants, corticosteroids, infliximab or rituximab.[8] NICE technology appraisals 379 and 864 recommend nintedanib for treating idiopathic pulmonary fibrosis. NICE technology appraisal 747 recommends nintedanib for treating progressive fibrosing interstitial lung diseases . NICE technology appraisal 504 recommends pirfenidone for treating idiopathic pulmonary

fibrosis. People may also receive non-pharmacological care from a multidisciplinary team.

The technology

Nerandomilast (brand name unknown, Boehringer Ingelheim) does not currently have a marketing authorisation in the UK for treating idiopathic pulmonary fibrosis or progressive pulmonary fibrosis. It has been studied in double-blind, randomised clinical trials for treating idiopathic pulmonary fibrosis and other types of progressive fibrosing interstitial lung diseases.

Intervention(s)	Nerandomilast
Population(s)	People with idiopathic pulmonary fibrosis or progressive pulmonary fibrosis
Subgroups	If the evidence allows the following subgroups will be considered: • Idiopathic pulmonary fibrosis • Progressive pulmonary fibrosis • Disease severity defined by Forced Vital Capacity (FVC)
Comparators	Established clinical management without nerandomilast, including but not limited to: For idiopathic pulmonary fibrosis: • nintedanib • pirfenidone (in people who have a FVC of 50% to 80% predicted) • best supportive care. For progressive pulmonary fibrosis: • nintedanib • immunosuppressants, such as methotrexate, azathioprine, cyclophosphamide or mycophenolate • corticosteroids • infliximab • rituximab • best supportive care.

Outcomes	The outcome measures to be considered include: • pulmonary function • physical function • exacerbation rate • hospitalisations • lung transplantation • mortality • adverse effects of treatment • health-related quality of life.
Economic analysis	The reference case stipulates that the cost effectiveness of treatments should be expressed in terms of incremental cost per quality-adjusted life year. The reference case stipulates that the time horizon for estimating clinical and cost effectiveness should be sufficiently long to reflect any differences in costs or outcomes between the technologies being compared. Costs will be considered from an NHS and Personal Social Services perspective. The availability of any commercial arrangements for the intervention, comparator and subsequent treatment technologies will be taken into account. The availability and cost of biosimilar and generic products should be taken into account.
Other considerations	Guidance will only be issued in accordance with the marketing authorisation. Where the wording of the therapeutic indication does not include specific treatment combinations, guidance will be issued only in the context of the evidence that has underpinned the marketing authorisation granted by the regulator.
Related NICE recommendations	**Related technology appraisals:** Nintedanib for treating idiopathic pulmonary fibrosis when forced vital capacity is above 80% predicted (2023) NICE technology appraisal guidance 864 Nintedanib for treating progressive fibrosing interstitial lung diseases. (2021) NICE Technology appraisal guidance 747. Review date to be confirmed. Pirfenidone for treating idiopathic pulmonary fibrosis (2018) NICE technology appraisal guidance 504 Nintedanib for treating idiopathic pulmonary fibrosis (2016) NICE technology appraisal guidance 379

	Related technology appraisals in development: Nintedanib for treating fibrosing interstitial lung disease in people aged 6 to 17. NICE technology appraisal guidance [ID6194] Publication expected: to be confirmed **Related NICE guidelines:** Idiopathic pulmonary fibrosis in adults: diagnosis and management (2013) NICE Clinical Guideline 163. Last updated: 23 May 2017

References

1. Olson AL, Gifford AH, Inase N, Fernández Pérez ER, Suda T. The epidemiology of idiopathic pulmonary fibrosis and interstitial lung diseases at risk of a progressive-fibrosing phenotype. Eur Respir Rev. 2018 Dec 21;27(150):180077. doi: 10.1183/16000617.0077-2018. PMID: 30578336; PMCID: PMC9489016.

2. Raghu G, Remy-Jardin M, Richeldi L, et al. Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults: An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. Am J Respir Crit Care Med. 2022 May 1;205(9):e18-e47. doi: 10.1164/rccm.202202-0399ST. PMID: 35486072; PMCID: PMC9851481.

3. Kang HK, Song JW. Progressive Pulmonary Fibrosis: Where Are We Now? Tuberc Respir Dis (Seoul). 2024 Apr;87(2):123-133. doi: 10.4046/trd.2023.0119. Epub 2023 Dec 18.

4. Gupta R, Morgan AD, George PM, et al. Incidence, prevalence and mortality of idiopathic pulmonary fibrosis in England from 2008 to 2018: a cohort study. Thorax 2024;79:624–631

5. Simpson T, Barratt SL, Beirne P, et al. The burden of progressive fibrotic interstitial lung disease across the UK. European Respiratory Journal. 2021. 58;1.

6. Snell N, Strachan D, Hubbard R, et al P272 Epidemiology of idiopathic pulmonary fibrosis in the uk: findings from the british lung foundation's 'respiratory health of the nation' project Thorax 2016;71:A236.

7. BMJ Best Practice. Idiopathic pulmonary fibrosis. Available at: https://bestpractice.bmj.com/topics/en-gb/446/treatment-algorithm. Accessed August 2025

8. BMJ Best Practice. Sarcoidosis. Available at: Sarcoidosis - Treatment algorithm | BMJ Best Practice. Accessed August 2025