NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Health Technology Evaluation

Inhaled treprostinil for treating pulmonary hypertension with interstitial lung disease

Final scope

Remit/evaluation objective

To appraise the clinical and cost effectiveness of inhaled treprostinil within its marketing authorisation for treating pulmonary hypertension with interstitial lung disease.

Background

Interstitial lung disease (ILD) is a group of lung disorders that cause scarring (fibrosis) of the lung tissue, which make the lungs stiff, hampering their ability to transfer oxygen to the bloodstream and making it harder to breath. The most common form is idiopathic pulmonary fibrosis.

ILD can progress to pulmonary hypertension with ILD (PH-ILD), which is classified by the World Health Organization as group 3 pulmonary hypertension. Group 3 PH has the lowest survival of the 5 groups. Median overall survival with PH-ILD has been estimated at 15.1 months.[1] Symptoms include shortness of breath, cough, and fatigue, which limit daily activities and physical functioning, and reduce health-related quality of life. The prevalence of PH-ILD in 2019 was estimated at 0.36 per 10,000 people and incidence 0.19 per 10,000.[1]

There are no treatments that specifically target PH-ILD. Phosphodiesterase 5 inhibitors (PDE-5i; sildenafil and tadalafil) are sometimes offered on a case-by-case basis to people with severe PH-ILD.

Conventional treatment of pulmonary hypertension which may be used as supportive treatment can include diuretics, home oxygen therapy, and digoxin.

The technology

Inhaled treprostinil (brand name unknown, Ferrer International) does not currently have a marketing authorisation in the UK for treating pulmonary hypertension with interstitial lung disease. It has been studied in a clinical trial compared with placebo in people with pulmonary hypertension associated with interstitial lung disease.

Intervention(s)	Inhaled treprostinil
Population(s)	Adults with a confirmed diagnosis of pulmonary hypertension with interstitial lung disease

Subgroups	If the evidence allows the following subgroups will be considered: • different types of interstitial lung disease, for example idiopathic pulmonary fibrosis, combined pulmonary fibrosis and emphysema, idiopathic interstitial pneumonia, sarcoidosis, hypersensitivity pneumonitis
Comparators	• Established clinical management without inhaled treprostinil • Phosphodiesterase 5 inhibitors: sildenafil and tadalafil
Outcomes	The outcome measures to be considered include: • exercise capacity (for example 6-minute walking distance) and other measures of physical function • time to clinical worsening • hospitalisations • overall survival • lung function • breathlessness • haemodynamic outcomes (for example cardiac index, cardiac output, right atrial pressure, pulmonary arterial pressure and pulmonary vascular resistance) • fatigue • transplant-free survival • 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 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. 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 NICE guidelines:** Idiopathic pulmonary fibrosis in adults: diagnosis and management (2013; updated 2017) NICE guideline CG163. **Related interventional procedures:** Balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension (2016) NICE interventional procedures guidance 554 **Related quality standards:** Idiopathic pulmonary fibrosis in adults (2015) NICE quality standard 79.
Related National Policy	The NHS Long Term Plan (2019) NHS Long Term Plan NHS England (2023) Prescribed specialised services manual (version 6) Chapter 4 Adult specialist respiratory Services Chapter 14 Adult Specialist pulmonary hypertension services NHS England (2018) Interstitial Lung Disease Service Adult Service Specification 17009/S

References

1. Kiely DG, Wort SJ, Funes DF, et al. (2024) Epidemiology, survival, and healthcare resource use of patients with pulmonary hypertension associated with lung disease and/or hypoxia in the UK. Poster presented at ISPOR Europe; 17 to 20 November 2024; Barcelona, Spain