NATIONAL INSTITUTE FOR HEALTH AND CARE EXCELLENCE

Single Technology Appraisal

Nintedanib for treating idiopathic pulmonary fibrosis in people with a forced vital capacity above 80% predicted (part-review of technology appraisal guidance 379) [ID4062]

Final scope

Draft remit/evaluation objective

To appraise the clinical and cost effectiveness of nintedanib within its marketing authorisation for treating idiopathic pulmonary fibrosis in people with a forced vital capacity above 80% predicted.

Background

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive lung disease in which scarring (fibrosis) occurs. The cause of IPF is unknown although it is thought to be related to an abnormal immune response to an unknown cause. It is a difficult disease to diagnose and requires a multidisciplinary team. Most people with IPF experience symptoms of breathlessness, which may initially be only on exertion. Cough, with or without sputum, is a common symptom. Over time, these symptoms are associated with a decline in lung function, reduced quality of life, disability and death.

The median survival for people with IPF in the UK is approximately 3-4 years from the time of diagnosis.[1,2] However, about 20-30% of people with IPF survive for more than 5 years.[1,2] The rate of disease progression can vary greatly. Prognosis is difficult to estimate at the time of diagnosis and may only become apparent after a period of careful follow-up.

The prevalence rate of IPF is about 50 per 100,000 [3] with an incidence of approximately 9 per 100,000 per year [4], which equates to around 6000 new diagnoses each year in the UK.[3,4] The incidence is higher in men than women, and increases with age (85% of diagnoses are made in people aged over 70 years).[3] IPF co-exists with chronic obstructive pulmonary disease in around 8-15% of people.

The aim of treatment is to manage the symptoms and slow progression. NICE clinical guideline 163 on the diagnosis and management of suspected idiopathic pulmonary fibrosis recommends that best supportive care (including symptom relief, management of co-morbidities, withdrawal of therapies suspected to be ineffective or causing harm and end of life care) should be offered to people from diagnosis and be tailored according to disease severity, rate of progression and the person's preference. If pharmacological treatment is considered appropriate, the guideline recommends use of pirfenidone and nintedanib if a person's forced vital capacity (FVC) is between 50% and 80% of their expected value in line with recommendations in NICE technology appraisal guidance 504 and 379. Treatment with pirfenidone and nintedanib should be discontinued if there is evidence of disease progression (a decline in per cent predicted FVC of 10% or more within any 12 month period). Lung transplantation is an option if there are no contraindications.

The existing recommendations in NICE technology appraisal guidance 379 are optimised for a narrower population than covered by the marketing authorisation. Stakeholders have indicated that there may be new information to warrant an update of the current guidance which only recommends nintedanib when a person has a forced vital capacity (FVC) between 50% and 80% predicted.

The technology

Nintedanib (Ofev, Boehringer Ingelheim) has a UK marketing authorisation for treating idiopathic pulmonary fibrosis in adults. It is administered orally.

Intervention(s)	Nintedanib
Population(s)	Adults with idiopathic pulmonary fibrosis with a forced vital capacity above 80% predicted
Comparators	Established clinical management without nintedanib
Outcomes	The outcome measures to be considered include: • pulmonary function parameters • physical function • exacerbation rate • 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.
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' (2016) NICE Technology appraisal guidance 379. Under review, anticipated date of publication TBC. 'Pirfenidone for treating idiopathic pulmonary fibrosis' (2018). NICE Technology appraisal guidance 504. Under review, anticipated date of publication TBC. **Related Guidelines:** 'COVID-19 rapid guideline: interstitial lung disease' (2020). NICE guideline 177. 'Idiopathic pulmonary fibrosis in adults: diagnosis and management (2013). NICE guideline 163. **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 Adult highly specialist respiratory services, chapter 4. NHS England (2018/2019) NHS manual for prescribed specialist services (2018/2019)

References

1. NHS. Idiopathic pulmonary fibrosis. [online accessed March 2022]

2. Khor, Y et.al. (2020) Prognosis of idiopathic pulmonary fibrosis without anti-fibrotic therapy: a systematic review. *European Respiratory Review*. 29: 190158

3. British Lung Foundation (2016) The battle for breath – the impact of lung disease in the UK. [online accessed March 2022]

4. Hutchinson, J (2015) Global incidence and mortality of idiopathic pulmonary fibrosis: a systematic review. *European Respiratory Society*. 46: 795-806