



# Effective Health Care

## Pulmonary Fibrosis

### Nomination Summary Document

#### Results of Topic Selection Process & Next Steps

- The topic, *Pulmonary Fibrosis*, was found to be addressed two systematic reviews on idiopathic pulmonary fibrosis from the Cochrane Collaboration, both of which suggested a lack of quality evidence on treatment options for idiopathic pulmonary fibrosis. Given that the existing reviews cover this nomination and pirfenidone is unavailable in the United States, no further activity will be undertaken on this topic.
  - Richeldi L, Davies HRHR, Spagnolo P, Luppi F. Corticosteroids for idiopathic pulmonary fibrosis. Cochrane Database of Systematic Reviews 2010, Issue 3. Art. No.: CD002880. DOI: 10.1002/14651858.CD002880.
  - Spagnolo P, Del Giovane C, Luppi F, Cerri S, Balduzzi S, Walters EH, D'Amico R, Richeldi L. Non-steroid agents for idiopathic pulmonary fibrosis. Cochrane Database of Systematic Reviews 2010, Issue 9. Art. No.: CD003134. DOI: 10.1002/14651858.CD003134.pub.

#### Topic Description

**Nominator(s):** Patient with idiopathic pulmonary fibrosis

**Nomination Summary:** The nominator is interested in the comparative effectiveness of treatment options for pulmonary fibrosis including which treatments are most appropriate for specific subgroups of patients. The nomination states that multiple treatment options may be used when treating this patient population. The nominator asserts there is little understanding of these treatment options and how they should be used within the patient population.

##### **Staff-Generated PICO**

**Population(s):** Individuals with pulmonary fibrosis, including idiopathic pulmonary fibrosis (IPF)

**Intervention(s):** Treatment options including, but not limited to: corticosteroids (e.g. prednisone), immunosuppressants (e.g. azathioprine), N-acetylcysteine (NAC), supplemental oxygen therapy, pulmonary rehabilitation, and lung transplantation

**Comparator(s):** Those listed above (i.e., compared to each other)

**Outcome(s):** Quality of life, morbidity, and mortality

**Key Questions from Nominator:** What is the comparative effectiveness of combination prednisone/Imuran/NAC, pirfenidone, and no treatment?

## Considerations

- The topic meets EHC Program appropriateness and importance criteria. (For more information, see <http://effectivehealthcare.ahrq.gov/index.cfm/submit-a-suggestion-for-research/how-are-research-topics-chosen/>.)
- Pulmonary fibrosis (PF) is defined as the scarring or thickening of lung tissue, which restricts an individual's ability to move oxygen into the lung stream. It is a progressive, life-threatening illness. PF may occur due to complications from other conditions including rheumatoid arthritis (RA) and chronic obstructive pulmonary disease (COPD). PF of unknown origin is referred to as idiopathic pulmonary fibrosis (IPF).
- Treatment options for PF/IPF include, but are not limited to: corticosteroids (prednisone), immunosuppressants (azathioprine), N-acetylcysteine (NAC), supplemental oxygen therapy, pulmonary rehabilitation, and lung transplantation.
- This topic has been addressed by two systematic reviews by the Cochrane Collaboration. The reviews looked at the use of corticosteroids and non-steroidal agents in the treatment of IPF. Researchers found that there was a lack of quality studies on the use of corticosteroids in the treatment of IPF.
- Pirfenidone is not currently approved by the Food and Drug Administration (FDA) for PF/IPF and therefore does not represent a healthcare intervention that is generally available in the United States.