Prevalence and Characteristics of Progressive Fibrosing Intertstitial Lung Diseases

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INTRODUCTION

Interstitial lung diseases (ILDs) represent a spectrum of lung disorders involving the lung parenchyma that often result in fibrosis (scarring) of lung tissue. The prototypical fibrosingILD is idiopathic pulmonary fibrosis (IPF). A subset of non-IPF patients with fibrosing ILD develop a progressive phenotype (PF-ILD). Antifibrotic therapy may help this subset of patients; however, the local prevalence of PF-ILD unknown

OBJECTIVES

To describe the local prevalence and clinical characteristics of patients with PF-ILD, and compare them to patients with fibrosing ILD without a progressive phenotype (non-progressors)

METHODS

Participants of the Canadian Registry for Pulmonary Fibrosis (CARE-PF), McMaster University site, were studied. All participants with 1) evidence of fibrosis on CT scan, 2) diagnosis other than IPF, and had serial forced vital capacity (FVC) measurements recorded were included, from November 2015 to March 2020.

Participants were classified as PF-ILD using the INBUILD study criteria: relative forced vital capacity (FVC) decline ≥10%, or any 2 of: relative FVC decline 5-10%, worsening respiratory symptoms or worsening fibrosis on HRCT, over the preceding 24 months.

The remaining patients that did not meet criteria for PF-ILD, but had non-IPF fibrotic ILD, were classified as non-progressors.

The primary outcomes were the rate of lung function decline, as determined by the mean absolute fall in FVC over the prior year, measured in ml ± standard deviation, and the prevalence of PF-ILD.

Prevalence was reported per 100,000 persons, using Hamilton Niagara Haldimand Brant Local Health Integration Network (HNHB-LHIN) population data.

A sensitivity analysis was done stratifying prevalence estimates by the following age groups: under 40, 40-59, 60-79, and 80 and above

RESULTS

We thank Boehringer Ingelheim Canada for providing funding to support this research and the CARE-PF registry

CONCLUSIONS

• PF-ILD is prevalent in the local community, with case rates highest in those in the 60-79 age group.
• Smoking and lower baseline lung function were important risk factors for PF-ILD.
• PF-ILD were most commonly patients with CTD-ILD, specifically RA-ILD, followed by patients with chronic hypersensitivity pneumonitis.
• The rate of disease progression in patients with PF-ILD, as measured by annual FVC decline, was 173±250ml, which matches disease progression rates seen in IPF.
• The rate of lung function decline was similar to the placebo arm of the INBUILD study, a recent randomized control trial showing that nintedanib, an antifibrotic drug, slows the rate of progression of FVC decline in PF-ILD.
• This data highlights the importance of identifying patients who meet criteria for PF-ILD, so that they may benefit from evidence-based therapy with antifibrotics.

REFERENCES


ACKNOWLEDGEMENTS

• We thank Boehringer Ingelheim Canada for providing funding to support this research and the CARE-PF registry