MEDIA FACT SHEET

Progressive Fibrosing Interstitial Lung Disease

A proportion of patients with a wide variety of interstitial lung diseases (ILDs) develop a 'progressive fibrosing phenotype'[1,2]

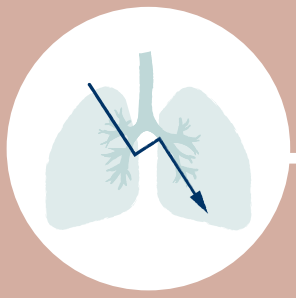
Patients with this phenotype have a disease course similar to the patients with idiopathic pulmonary fibrosis (IPF)[1,2]

In these patients, lung fibrosis is believed to become self sustaining,[1,3–7] and leads to irreversible[7–9] and progressive damage[8–10] to both lung structure and function[6–10]

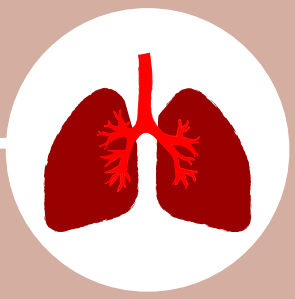
PROGRESSIVE FIBROSING PHENOTYPE

PROGRESSIVE

Progressive disease with declining lung function,[1,11–14] worsening of symptoms[1,13,15,16] and quality of life[15–17]



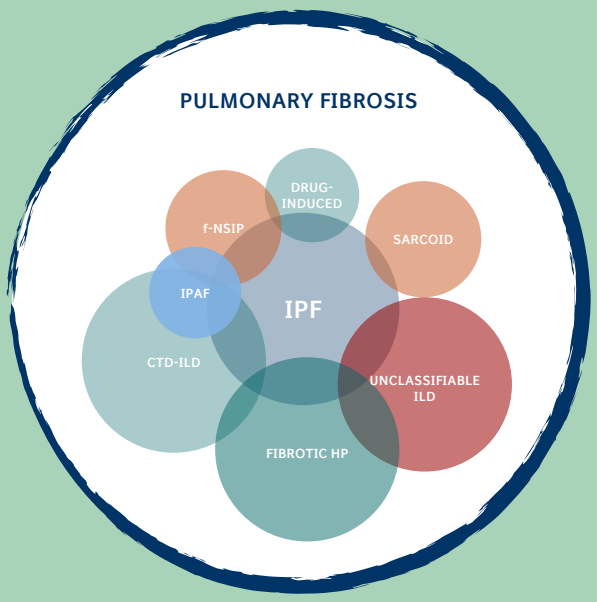
FIBROSING



Self-sustaining fibrosis causing irreversible,[7–9] diffuse[1] fibrotic destruction of lung architecture[6–10]

OVER 200 TYPES OF ILDs IDENTIFIED[1,2,18,19]

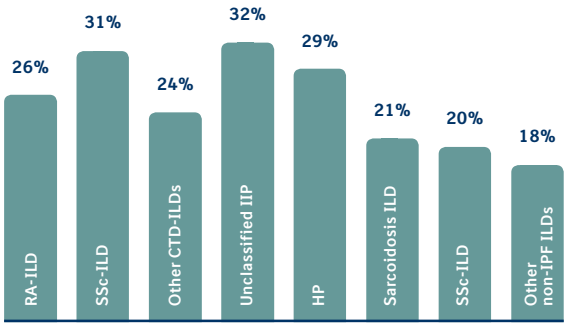
ILDs THAT MAY BE ASSOCIATED WITH A PROGRESSIVE FIBROSING PHENOTYPE[1,20]



There is an overlap in longitudinal disease behaviour between idiopathic pulmonary fibrosis and other disorders. The size of the circles reflects the approximate prevalence of individual diseases and counted at ILD centres.[20]

Adapted from Flaherty KR, et al. BMJ Open Resp Res 2017;4:e000212; Wells AU, et al. Eur Respir J 2018; 51(5):pii:1800692.

ESTIMATED PERCENTAGE OF PATIENTS WITH NON-IPF ILD THAT DEVELOP PROGRESSIVE FIBROSING PHENOTYPE*†‡[21]



Based on online physician survey*†‡[21]

18–32%

of patients diagnosed with ILD develop a progressive fibrosing phenotype

Delayed referral to a specialist is likely to delay diagnosis and management of progressive fibrosing ILD

Survival time in patients with non-IPF progressive fibrosing ILD is believed to be similar to that of patients with IPF

*Progressive fibrosing was defined as fibrosis detected by HRCT (i.e. reticular abnormality with traction bronchiectasis with or without honeycombing) that was progressive in terms of worsening of lung function (FVC and/or D_{LCO}) and/or respiratory symptoms and/or chest images.
†Research funded by Boehringer Ingelheim.
‡Estimates of percentage of patients with non-IPF ILD that develop progressive fibrosis based on online survey of total 486 physicians (243 pulmonologists, 203 rheumatologists and 40 internist) from the United States, Japan, France, Germany, Italy, Spain and the United Kingdom.

PROGRESSIVE FIBROSING ILDs HAVE A SIGNIFICANT IMPACT ON PATIENTS[11,15]

Deteriorating lung function[1,11–14]
Worsening respiratory symptoms[1,1,3,15,16]
Reduced quality of life[15–17]
Early mortality[1,2,13,14,22]

THERE ARE SIGNIFICANT UNMET NEEDS FOR PATIENTS AND THEIR PHYSICIANS

NO DRUGS

are currently approved for treatment of patients with progressive ILDs, other than IPF[1]

CLOSE COLLABORATION

between pulmonologists, rheumatologists, and radiologists is important in the diagnosis and management of these complex patients[23]

ABBREVIATIONS

References

CTD, connective tissue disease; D_{LCO}, diffusing capacity of the lungs for carbon monoxide; f-NSIP, fibrotic non-specific interstitial pneumonia; FVC, forced vital capacity; HP, hypersensivity pneumonitis; HRCT, high-resolution computed tomography; IIP, Idiopathic interstitial pneumonia; ILD, interstitial lung disease; IPAF, interstitial pneumonia with autoimmune features; IPF, idiopathic pulmonary fibrosis; non-IPF ILD, non-idiopathic pulmonary fibrosis interstitial lung disease; RA-ILD, rheumatoid arthritis-associated interstitial lung disease; SSc-ILD, systemic sclerosis-associated interstitial lung disease.

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Nintedanib is approved in Europe for the treatment of adult patients with idiopathic pulmonary fibrosis (IPF).
Nintedanib is an investigational compound in progressive fibrosing interstitial lung diseases other than IPF. Its safety and efficacy in the clinical trials mentioned here have not yet been established. This information is presented solely to provide an overview of these clinical trials and should not be interpreted as a recommendation for use in these indications.

Boehringer Ingelheim