

WHEN PULMONARY FIBROSIS IS SUSPECTED, WHAT'S NEXT FOR ROBERT?

CLINICAL EVALUATION:

- 68-year-old male presenting with worsening dyspnea and unexplained weight loss¹
- Inspiratory crackles on lung auscultation¹
- Restrictive pattern on PFTs^{2,3}
- Positive HP panels with precipitating antibodies against mold⁴
- Broncho-alveolar lavage lymphocytes >50%⁴

RADIOLOGIC FINDINGS:

- Fibrotic ILD confirmed by HRCT¹:
 - Areas of mosaic lung attenuation on inspiratory image are confirmed to be air trapping on expiratory images
 - Peripheral reticulation; no honeycombing
 - Ground glass opacity present
 - Upper and lower lobe involvement

Treated for chronic hypersensitivity pneumonitis (cHP) with removal of exposure (mold) and corticosteroid⁴

3-MONTH FOLLOW-UP:

- Dyspnea continues to worsen despite removal of inciting exposure and treatment with corticosteroid for 3 months²
- PFTs have declined²:
 - 5% decrease in FVC % predicted**
 - 3% decrease in DL_{co} % predicted**

Lung function decline and worsening respiratory symptoms are indicators of progressive disease⁵

cHP, chronic hypersensitivity pneumonitis; DL_{co}, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; HP, hypersensitivity pneumonitis; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; PFTs, pulmonary function tests.

Despite removal of the inciting exposure and immunosuppressive therapy, lung function has declined and fibrotic ILD is detected.²

WHAT IS THE NEXT STEP IN ROBERT'S DISEASE MANAGEMENT PLAN?

~1 IN 4 PATIENTS WITH ILD MAY DEVELOP A PROGRESSIVE PHENOTYPE^{6*}

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs^{5,7,8}

- Idiopathic pulmonary fibrosis
- Systemic sclerosis-associated ILD
- Rheumatoid arthritis-associated ILD
- Other connective tissue disease-associated ILDs
- Hypersensitivity pneumonitis
- Occupational exposure-related ILDs
- Idiopathic nonspecific interstitial pneumonia
- Unclassifiable interstitial pneumonia
- Sarcoidosis

SIMILAR TO IPF, SOME ILDs CAN DEVELOP A PROGRESSIVE FIBROSING PHENOTYPE CHARACTERIZED BY^{5,8}:



Worsening respiratory symptoms



Accelerated decline in lung function



Worsening quality of life



Early mortality

EARLY IDENTIFICATION OF ILD IS CRITICAL

Observe for respiratory symptoms⁹:

Listen for⁹:

Order baseline PFTs and monitor regularly⁹:

Order HRCT if ILD is suspected^{9,10}:



Cough



Velcro[®]-like crackles on lung auscultation



Restrictive PFT



Presence of fibrotic ILD



Dyspnea



Reduced DL_{co}

EARLY IDENTIFICATION OF PROGRESSIVE PULMONARY FIBROSIS CAN HELP ENSURE PATIENTS RECEIVE APPROPRIATE INTERVENTION TO SLOW PROGRESSION⁵

*Data from a global, online survey of physicians (n=486).⁶

IPF, idiopathic pulmonary fibrosis.

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