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 fibrotic hypersensitivity pneumonitis (HP) 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_∞ % predicted

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

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 PHENOTYPE6*

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs5,7,8

- Idiopathic pulmonary fibrosis (IPF)
- 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 ILD
- Sarcoidosis

SIMILAR TO IPF, SOME ILDs CAN DEVELOP A PROGRESSIVE FIBROSING PHENOTYPE CHARACTERIZED BY5,8:



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 for10,11:

Order baseline PFTs and monitor regularly⁹:

Order HRCT if ILD is suspected^{9,12}:



Cough



Dry inspiratory crackles, typically at the lung bases



Restrictive PFT



Presence of fibrotic ILD





Reduced DL_{co}

EARLY IDENTIFICATION OF PROGRESSIVE PULMONARY FIBROSIS CAN HELP ENSURE PATIENTS RECEIVE APPROPRIATE INTERVENTION⁵

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

IPF, idiopathic pulmonary fibrosis.

References: 1. Gimenez A et al. *Thorax*. 2018;73(4):391-392. 2. Jacob J et al. *BMC Pulm Med*. 2017;17(1):81. 3. Johnson JD, Theurer WM. *Am Fam Physician*. 2014;89(5):359-366. 4. Vourlekis JS et al. *Am J Med*. 2004;116(10):662-668. 5. Cottin V et al. *Eur Respir Rev*. 2018;27(150). doi:10.1183/16000617.0076-2018 6. Wijsenbeek M et al. *Curr Med Res Opin*. 2019;35(11):2015-2024. 7. Demedts M et al. *Eur Respir J*. 2001;18(suppl 32):2s-16s. 8. Wells AU et al. *Eur Respir J*. 2018;51(5). doi:10.1183/13993003.00692-2018 9. Ryu JH et al. *Mayo Clin Proc*. 2007;82(8):976-986. 10. Silver KC, Silver RM. *Rheum Dis Clin North Am*. 2015;41(3):439-457. 11. Zibrak JD, Price D. *NPJ Prim Care Respir Med*. 2014;24:14054. 12. Walsh SLF et al. *Eur Respir Rev*. 2018;27(150):976-986.

