WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR DAVID?

MEDICAL HISTORY:

- 52-year-old Caucasian male[1-3]
- Former smoker, 30 pack-years[1,3]
- Presented with dyspnea and fatigue in hospital; consolidation found on chest X-ray[3]
- Treated for pneumonia with broad spectrum antibiotics[3]
  - —Cultures consistently negative

INITIAL CLINICAL EVALUATION:

- Exertional dyspnea, mild fatigue[3]
- Velcro®-like crackles on auscultation[3]
- Reduced PFTs with restriction[1]

69% FVC % predicted

48% DL_{CO} % predicted

- Rheumatologic exam was negative[3]
- Rheumatoid factor and anticitrullinated C-peptide positive, slightly increased creatinine kinase and CRP levels[3]

Treated with steroids and alkylating agent and showed some symptomatic improvement[3]

1 YEAR FOLLOW-UP:

- Respiratory symptoms have worsened[3]
- Underlying etiology unclear[3]
- Stable PFTs[4]
- Extensive fibrosis on HRCT[3]
  - —Ground glass opacities, traction bronchiectasis, mosaic attenuation, and reticulation

After review of clinical, radiographic, and histologic findings by an MDT did not produce a conclusive diagnosis, ILD was considered unclassifiable[3]

CRP, C-reactive protein; DL_{CO}, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; MDT, multidisciplinary team; PFT, pulmonary function test; UIIP, unclassifiable idiopathic interstitial pneumonia.

FVC is not declining, but symptoms and fibrosis are worsening, which are indicators of progressive disease.[4]

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

Not an actual patient.

HRCT scan courtesy of Leung SC et al. *Respirol Case Rep*. 2015;3(3):85-88. Published by John Wiley & Sons Ltd.

~1 IN 4 PATIENTS WITH ILD MAY DEVELOP A PROGRESSIVE PHENOTYPE[5]*

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs[6-8]

- 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[7,8]:


Worsening respiratory symptoms


Accelerated decline in lung function


Worsening quality of life


Early mortality

EARLY IDENTIFICATION OF ILD IS CRITICAL

Observe for respiratory symptoms[9]:


Cough


Dyspnea

Listen for[9]:


Velcro®-like crackles on lung auscultation

Order baseline PFTs and monitor regularly[9]:


Restrictive PFT


Reduced DL_{CO}

Order HRCT if ILD is suspected[9,10]:


Presence of fibrotic ILD

EARLY IDENTIFICATION OF PROGRESSIVE PULMONARY FIBROSIS CAN HELP ENSURE PATIENTS RECEIVE APPROPRIATE INTERVENTION TO SLOW PROGRESSION[7]

*Data from a global, online survey of physicians (n=486).[5]

IPF, idiopathic pulmonary fibrosis.

References: 1. Ryerson CJ et al. *Eur Respir J.* 2013;42(3):750-757. **2.** Hyldgaard C et al. *Respirology.* 2017;22(3):494-500. **3.** Leung SC et al. *Respirol Case Rep.* 2015;3(3):85-88. **4.** Skolnik K, Ryerson CJ. *Respirology.* 2016;21(1):51-56. **5.** Wijsenbeek M et al. *Curr Med Res Opin.* 2019;35(11):2015-2024. **6.** Demedts M et al. *Eur Respir J.* 2001;18(suppl 32):2s-16s. **7.** Cottin V et al. *Eur Respir Rev.* 2018;27(150). doi:10.1183/16000617.0076-2018. **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.** Walsh SLF et al. *Eur Respir Rev.* 2018;27(150):976-986.

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