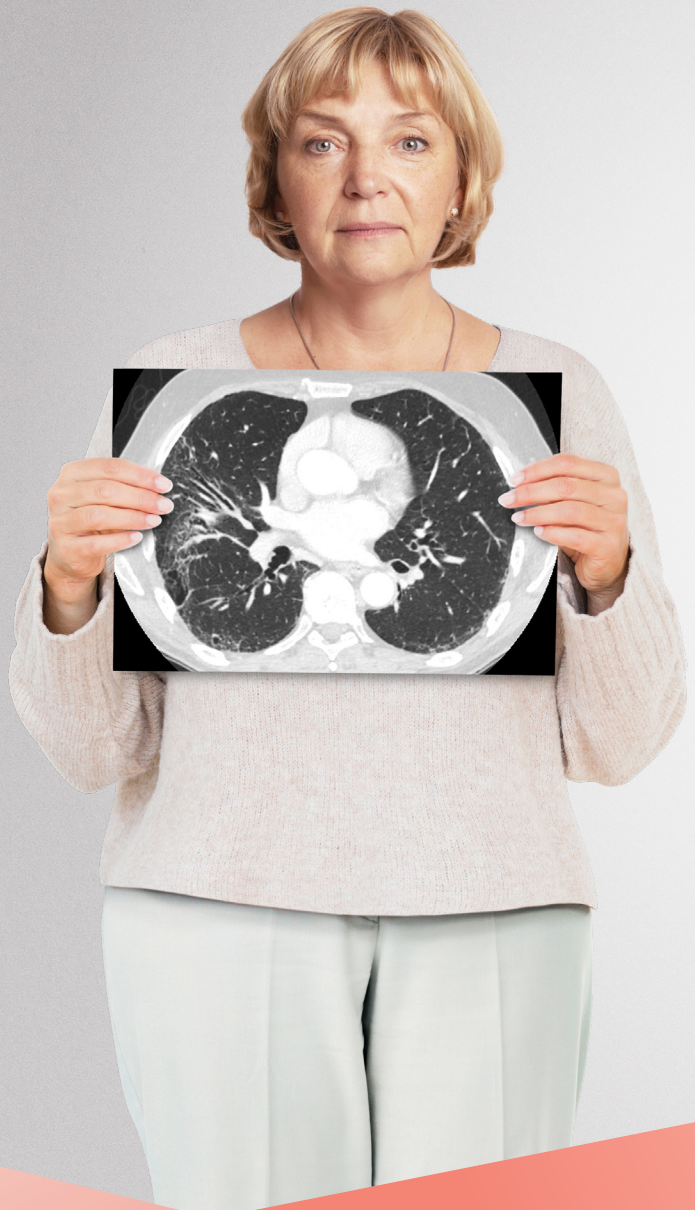
WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR LISA?



MEDICAL HISTORY:

- 50-year-old Caucasian woman[1,2]
- Diagnosed with polymyositis 3 years ago[1,2]
- High levels of anti-Jo-1 antibodies[3]
- Current treatment: immunosuppressant and corticosteroid[2,4,5]

Presence of anti-Jo-1 indicates an increased risk of ILD in patients with myositis[3]

CLINICAL EVALUATION:

- Development of pulmonary symptoms: dyspnea, cough[4-6]
- Dry inspiratory crackles at the lung bases, pneumonia-like symptoms[6]
- Reduced PFTs with restriction[4,6]

RADIOLOGIC FINDINGS:

- Fibrotic ILD confirmed by features consistent with a UIP-like pattern[4,6,7]
  - —Reticulation
  - —Bilateral traction bronchiectasis

Diagnosis: myositis-associated ILD[3,6]

MONITORING:

- Lung function declined over 6 months[4]

10% decrease in FVC % predicted[4]
5% decrease in DL_{CO} % predicted[4]

Declining lung function is a sign of progressive disease[5]

DL_{CO}, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; ILD, interstitial lung disease; PFTs, pulmonary function tests; UIP, usual interstitial pneumonia.

Fibrotic ILD is detected and lung function is worsening despite immunomodulatory therapy.[5]

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

Not an actual patient.

HRCT scan courtesy of Dr. Hani Salam, Radiopaedia.org, rID: 45584.

~1 IN 4 PATIENTS WITH ILD MAY DEVELOP A PROGRESSIVE PHENOTYPE[8]*

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs[7,9,10]

- 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 BY[9,10]:


Worsening respiratory symptoms


Accelerated decline in lung function


Worsening quality of life


Early mortality

EARLY IDENTIFICATION OF ILD IS CRITICAL

Observe for respiratory symptoms[11]:


Cough


Dyspnea

Listen for[12,13]:


Dry inspiratory crackles, typically at the lung bases

Order baseline PFTs and monitor regularly[11]:


Restrictive PFT


Reduced DL_{co}

Order HRCT if ILD is suspected[11,14]:


Presence of fibrotic ILD

EARLY IDENTIFICATION OF PROGRESSIVE PULMONARY FIBROSIS CAN HELP ENSURE PATIENTS RECEIVE APPROPRIATE INTERVENTION[9]

*Data from a global, online survey of physicians (N=486).[8]

HRCT, high-resolution computed tomography.

References: 1. Furst DE et al. *Muscle Nerve.* 2012;45(5):676-683. **2.** Smoyer-Tomic KE et al. *BMC Musculoskelet Disord.* 2012;13:103. doi:10.1186/1471-2474-13-103 **3.** Bernstein RM et al. *Br Med J (Clin Res Ed).* 1984;289(6438):151-152. **4.** Fathi M et al. *Arthritis Rheum.* 2008;59(5):677-685. **5.** Morisset J et al. *Chest.* 2016;150(5):1118-1128. **6.** Fujisawa T et al. *PLoS ONE.* 2014;9(6):e98824. doi:10.1371/journal.pone.0098824 **7.** Demedts M et al. *Eur Respir J.* 2001;18(suppl 32):2s-16s. **8.** Wijsenbeek M et al. *Curr Med Res Opin.* 2019;35(11):2015-2024. **9.** Cottin V et al. *Eur Respir Rev.* 2018;27(150). doi:10.1183/16000617.0076-2018 **10.** Wells AU et al. *Eur Respir J.* 2018;51(5). doi:10.1183/13993003.00692-2018 **11.** Ryu JH et al. *Mayo Clin Proc.* 2007;82(8):976-986. **12.** Silver KC, Silver RM. *Rheum Dis Clin North Am.* 2015;41(3):439-457. **13.** Zibrak JD, Price D. *NPJ Prim Care Respir Med.* 2014;24:14054. **14.** Walsh SLF et al. *Eur Respir Rev.* 2018;27(150):180073. doi:10.1183/16000617.0073-2018

Boehringer Ingelheim