WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR STEPHANIE?

Not an actual patient.

MEDICAL HISTORY:

- 49-year-old Caucasian woman[1]
- Diagnosed with limited cutaneous systemic sclerosis (lcSSc) 3 years ago[1]
- Anti-nuclear antibodies (ANA)[2]
- Normal PFTs and HRCT at baseline[3]
- Current medication: methotrexate (MTX) to inhibit inflammatory pathways[4,5]

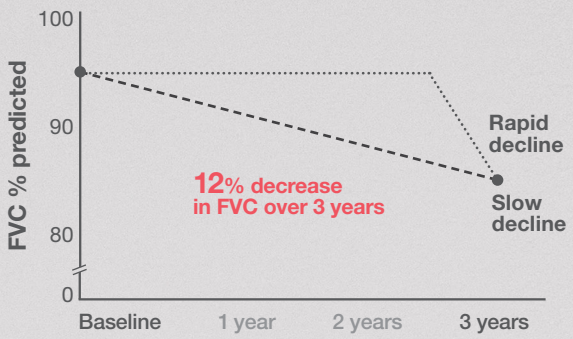
Stephanie was advised that immunosuppressive therapies, such as MTX, can increase her risk of infection[6,7]

CLINICAL EVALUATION AT 3 YEARS:

- Recent development of respiratory symptoms: dyspnea on exertion and bibasilar fine crackles on auscultation[3]
- Declined FVC, with reduced DL_{CO}[1,8,9]

Without routine monitoring of PFTs, it is unknown if Stephanie's decline was rapid or gradual[3]

Potential courses of FVC decline



REFERRAL TO PULMONOLOGIST FOR SSc-ILD EVALUATION:

- Fibrotic ILD detected on follow-up HRCT with features consistent with an NSIP pattern[10,11]
  - —Bilateral ground glass opacity with reticulation
  - —Traction bronchiectasis
  - —Subpleural sparing

Despite treatment with MTX, lung function has declined and fibrotic ILD is detected.

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

IDENTIFYING PULMONARY FIBROSIS AS EARLY AS POSSIBLE IS CRITICAL FOR TIMELY INTERVENTION

Observe for respiratory symptoms[3]:	 **Cough**	 **Dyspnea**
Listen for[3,12]:	 **Dry inspiratory crackles, typically at the lung bases**	
Order baseline PFTs and monitor regularly[3]:	 **Restrictive PFT**	 **Reduced DL_{CO}**
Order HRCT to confirm lung fibrosis[10]:	 **NSIP pattern is typical in patients with SSc-ILD, but UIP can be seen as well**	

VIGILANT AND PROACTIVE MONITORING FOR ILD IS IMPORTANT TO PROVIDE PATIENTS WITH APPROPRIATE INTERVENTION[3]

DL_{CO}, diffusing capacity for carbon monoxide; FEV_1, forced expiratory volume in 1 second; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; NSIP, nonspecific interstitial pneumonia; PFT, pulmonary function test; SSc-ILD, systemic sclerosis-associated interstitial lung disease; TLC, total lung capacity; UIP, usual interstitial pneumonia.

References: 1. Yasuoka H. *Clin Med Insights Circ Respir Pulm Med*. 2015;9(suppl 1):97-110. **2.** Jaeger VK et al. *PLoS ONE*. 2016;11(10):1-15. **3.** Silver KC, Silver RM. *Rheum Dis Clin North Am*. 2015;41(3):439-457. **4.** Kowal-Bielecka O et al. *Ann Rheum Dis*. 2017;76(8):1327-1339. **5.** Rasuvo® (methotrexate) Prescribing Information. Chicago, IL: Medac Pharma Inc; 2018. **6.** Margaritopoulos GA et al. *Eur Respir Rev*. 2017;26(143). doi:10.1183/16000617.0027-2016 **7.** Genestier L et al. *J Clin Invest*. 1998;102(2):322-328. **8.** Johnson JD et al. *Am Fam Physician*. 2014;89(5):359-366. **9.** Hu S et al. *Arthritis Res Ther*. 2018;20(1):1-9. **10.** Cappelli S et al. *Eur Respir Rev*. 2015;24(137):411-419. **11.** Kligerman SJ et al. *RadioGraphics*. 2009;29(1):73-87. **12.** Zibrak JD, Price D. *NPJ Prim Care Respir Med*. 2014;24:14054.

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