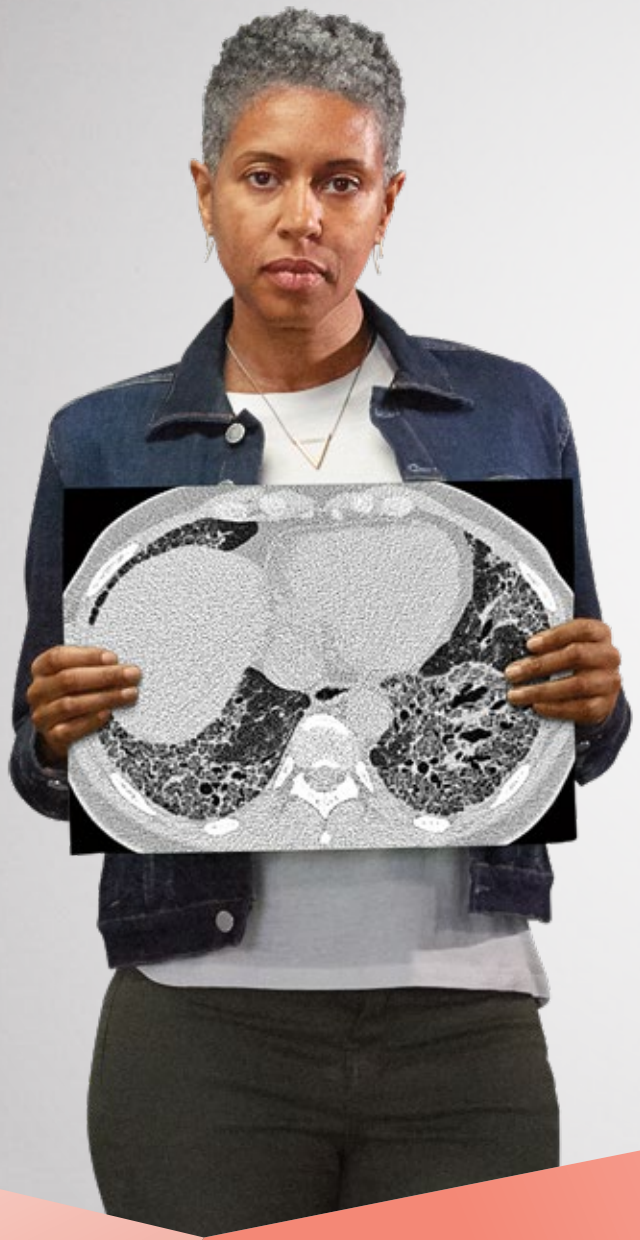
WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR ZOE?



Not an actual patient.

MEDICAL HISTORY:

- 44-year-old African-American woman[1]
- Diagnosed with diffuse cutaneous systemic sclerosis (dcSSc) 1 year ago[1]
- Anti-topoisomerase antibodies (ATA) and anti-nuclear antibodies (ANA)[2,3]
- Normal PFTs at baseline, but HRCT results reveal minimal parenchymal abnormalities[4]
- Current medication: mycophenolate (MMF) to inhibit inflammatory pathways[5,6]

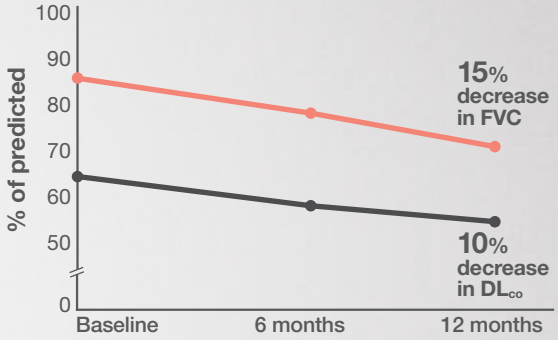
CLINICAL EVALUATION AT 1 YEAR:

INFECTIONS

- Recurrent esophageal candidiasis, resolved with treatment[6,7]

PULMONARY DECLINE

- Development of respiratory symptoms: dyspnea and cough[1]
- Normal breath sounds on lung auscultation[4]
- Declined FVC, with reduced DL_{CO}[1,3,5,8,9]



Immunosuppressive therapies, such as MMF, can increase the risk of infection[6,10]

REFERRAL TO PULMONOLOGIST FOR SSc-ILD EVALUATION:

- Worsening fibrotic ILD detected on follow-up HRCT with features consistent with a probable UIP pattern[11]
  - —Traction bronchiectasis
  - —Ground glass opacity
  - —Reticulation

SSc-ILD is more likely to be severe in African American patients[12*]

*Severe ILD is defined as pulmonary fibrosis with at least one of the following: FVC <50% of predicted, requirement for oxygen, lung transplantation, or death due to SSc-ILD.[12]

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

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

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

Observe for respiratory symptoms[4]:	 **Cough**  **Dyspnea**
Listen for[4,13]:	 **Dry inspiratory crackles, typically at the lung bases**
Order baseline PFTs and monitor regularly[4]:	 **Restrictive PFT**  **Reduced DL_{CO}**
Order HRCT to confirm lung fibrosis[11]:	 **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[4]

DL_{CO}, diffusing capacity for carbon monoxide; 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; UIP, usual interstitial pneumonia.

References: 1. Yasuoka H. *Clin Med Insights Circ Respir Pulm Med.* 2015;9(suppl 1):97-110. **2.** van den Hoogen F et al. *Arthritis Rheum.* 2013;65(11):2737-2747. **3.** Jaeger VK et al. *PLoS ONE.* 2016;11(10):1-15. **4.** Silver KC, Silver RM. *Rheum Dis Clin North Am.* 2015;41(3):439-457. **5.** Tashkin DP et al. *Lancet Respir Med.* 2016;4(9):708-719. **6.** Cellcept® (mycophenolate mofetil) Prescribing Information. South San Francisco, CA: Genentech USA, Inc; 2019. **7.** Juárez M et al. *Rheum Dis Clin North Am*. 2003;29(1):163-184. **8.** Johnson JD et al. *Am Fam Physician.* 2014;89(5):359-366. **9.** Volkmann ER et al. *Ann Am Thorac Soc.* 2016;13(11):2045-2056. **10.** Margaritopoulos GA et al. *Eur Respir Rev.* 2017;26(143). doi:10.1183/16000617.0027-2016 **11.** Cappelli S et al. *Eur Respir Rev.* 2015;24(137):411-419. **12.** Steen V et al. *Arthritis Rheum.* 2012;64(9):2986-2994. **13.** Zibrak JD, Price D. *NPJ Prim Care Respir Med*. 2014;24:14054.

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