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**Project ECHO for Interstitial Lung Disease  
Case Form**

ECHO ID: ILDECHO-056

 New Case Follow-up Case

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Current or suspected ILD diagnosis (if known): MCTD-ILD

How was this diagnosis made (select all that apply)?

- Radiology  
 Pathology  
 Multidisciplinary discussion

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**Main Question:**

By presenting this case, I hope to obtain:

- Help making a diagnosis                       Help with lab/test interpretation  
 Help with clinical management  
 Other (please specify):

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**Patient Demographic Information:**

Male    Female    Transgender                      Age (years): 33

State of primary residence: VA

Ethnicity:  Hispanic or Latino    Not Hispanic or Latino

Race (check all that apply):  American Indian or Alaskan Native    Asian

Black or African American    Native Hawaiian/Pacific Islander    White

Other    Unknown

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**Case Summary:**

32F with pmh of remote Crohn's not on therapy, MCTD with ILD who is now pregnant. Initially presented with asymptomatic LFT elevation in spring 2024 (AST>ALT) and found to have elevated CPK. She presented to rheumatology 5/2024 with Raynaud's, arthralgias, fatigue, GERD, nonexertional dyspnea, dry

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eyes, weakness in her hands. She was found to have on exam hand discoloration, abnormal nailfold capillaroscopy with sclerodactyly, slight weakness, arthralgias.

She had progressive weakness, dyspnea and fatigue in her legs and arms as well as myalgias when she was referred to pulmonary for concern of ILD. Started on nifedipine for Raynaud's in the fall 2024. Became pregnant at end of 2024.

She works as a clinic manager. Has a hypoallergenic dog. Only hobby is baking.

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**Exposure History:**

- Previous or current bird ownership    Down products in the home
- Woodworking    Well water    Hay exposure    Hot tub/jacuzzi/sauna
- Humidifiers    Dusty environments    Water damage or mold at work or home
- Farmland/barns    Amiodarone    Chemotherapy past/present
- Chest radiation past/present    Asbestos

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**Past Medical History:**

Crohn's disease on anti-TNF therapy 2000-2012

GERD

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**Medications:**

Nifedipine

Omeprazole

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**Focused ROS:**

- Rashes    Skin thickening    Arthralgias    Myalgias    Muscle weakness
- Dry mouth    Dry eyes    Red or painful eyes    Raynaud's    Oral ulcers
- Alopecia    Dysphagia    Heartburn/reflux    Fevers    Night sweats
- Palpitations    Weight loss

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**Smoking History:**

- Never smoked
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- Current smoker (          packs per day)
- Pack years:                  Quit date (if applicable):
- Cocaine use (route)
- Vaping/e-cigarettes (frequency)
- Inhaled marijuana (quantity)

**Occupations, current and previous (if industrial or factory work, please provide specific details):**

Clinic office manager

**Travel and Residential History:**

From midwest, then North Carolina before moving to DMV. No remarkable travel.

**Family History:**

- Pulmonary fibrosis or interstitial lung disease
- RA, Lupus, or other autoimmune diseases"
- Premature gray hair     Cirrhosis of the liver     Bone marrow disorders
- Leukemias

*Comments:* Crohn's mother

**Physical Exam:**

*Vital signs:*

BP: 98/65    Height: 5' 4"    Wt: 53.5kg    BMI: 20

Oxygen saturation: 98%    Ambulatory saturation: 96%

*HEENT:*

- Scleral injection     Dry mucous membranes     Poor dentition     Ptosis

*Pulmonary:*

- Wheezes     Rhonchi     Crackles
- Squeaks     Stridor     Dullness to percussion     Pleural Rub
- Bronchial breath sounds

Other:

*Cardiac:*

- Murmur                       Gallop  
 RV heave    Pulmonary tap    JVD    Irregular  
 Edema       Other:

*Abdomen:*

- Distended    Tender    Tympanitic    Pulsatile liver    Fluid wave  
 Other:

*Skin/Nails:*

- Rash (location, description) Telangiectasias on chest  
 Clubbing    Ragged cuticles    Telangiectasias  
 Abnormal nailfold capillaroscopy  
 Digital swelling / Sclerodactyly    Ulcerations  
 Mechanics hands

*Joints:*

- Synovitis                       Deformity    Tenderness    Erythema

*Neuro:*

- Proximal muscle weakness UE deltoid bilaterally  
 Other:

*Other pertinent findings:*

**Relevant Studies (please list key findings):**

PFTs:

<b>MM/YYYY:</b>	<b>06/2024</b>	<b>08/2024</b>
FVC	2.78 (79%)	2.58 (73%)
FEV1	2.29 (77%)	2.28 (77%)
FEV1/FVC	82%	88%

DsbHb		14.4 (68%)
TLC	3.75 (73%)	3.66 (72%)
VA		3.60

- CXR                                      Uploaded to Ambra:  Yes  No  
 CT chest                                      Uploaded to Ambra:  Yes  No  
 Echocardiogram                              Uploaded to Ambra:  Yes  No  
 Right heart catheterization:  
 Other relevant testing:

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### Relevant Labs:

Has the patient been evaluated by a Rheumatologist?  Yes  No

Date of last labs (month/year): 08/2024

CBC/differential: WBC 5.4 Hg 10.4 PLT 250

ANA:  Neg  Titer 1:1280 Pattern speckled

ANA #2:  Titer                      Pattern

ANA #3:  Titer                      Pattern

Smith:  Neg  Titer >8.0

SSA/Ro60:  Neg  Titer 1.2

SSB/La:  Neg  Titer

dsDNA:  Neg  Titer

Scl-70:  Neg  Titer

Centromere:  Neg  Titer

RNA polymerase III:  Neg  Titer

RNP:  Neg  Titer >8

RF:  Neg  Titer 169.2

CCP:  Neg  Titer

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ANCA:  Neg  Titer      Pattern

MPO:  Neg  Titer

PR3:  Neg  Titer

PM1-Scl:  Neg  Titer

Ro52:  Neg  Titer 70

Jo-1:  Neg  Titer

EJ:  Neg  Titer

OJ:  Neg  Titer

PL-7:  Neg  Titer

PL-12:  Neg  Titer

MDA5:  Neg  Titer

KU:  Neg  Titer

MI-2:  Neg  Titer

P155/140 (TIF1y):  Neg  Titer

NXP-2:  Neg  Titer

SRP:  Neg  Titer weak positive

SAE-1:  Neg  Titer

HP panel:  Neg  Pos

C3/C4:  WNL  Low

ESR:

CRP: low

CK: 1355

Aldolase:

ACE:

Urinalysis: normal Ur prot/cr

*Additional Comments:*

U3RNP neg; PM-SCL100 neg, PMSCL 75 neg, Anti-Th/To neg, anticardiolipin IgA IgG, IgM neg; U1-RNP 200, U2-RNP weak positive

Beta-2 glycoprotein IAb IgM H(131), IgG H (21), IgA neg; dRVVT and PTT-LA neg

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**Questions:**

- 1) What is the likely rheumatological diagnosis?
  - 2) How would you manage the lung disease at this stage in light of her pregnancy?
  - 3) How would you interpret the +SSA in terms of pregnancy risk?
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