PILOT™ Education Transforming PULMONARY CARE

A Closer Look at ILD, Sarcoidosis and CTEPH

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Case 1: Eugene

Eugene: Presentation

- Eugene is a 56-year-old male
- He presents with progressive dyspnea for 18 months
  - First noted symptoms when traveling to higher altitudes
  - Now notes symptoms climbing a flight of stairs
- Over the last six months, he has had a non-productive cough
- He saw his PCP, who heard “crackles” and is referred to you for additional evaluation
- He has no other symptoms

History

- **PMHx**
  - Obstructive sleep apnea, on BIPAP
  - Depression
- **Medications**
  - Ibuprofen prn
  - Sertraline 50 mg/day
- **SHx**
  - Current smoker, one pack-per-day for 40 years
- **FHx**
  - Father died of “lung disease”
- **Environmental/occupational Hx**
  - Two years ago had a flood in basement, this was remediated

History

- Signs or symptoms of a systemic autoimmune disorder?
- Clinically relevant exposures (occupational and environmental)?
- Drugs that may account for the presence of lung disease?
- Relevant family history?

Physical Exam

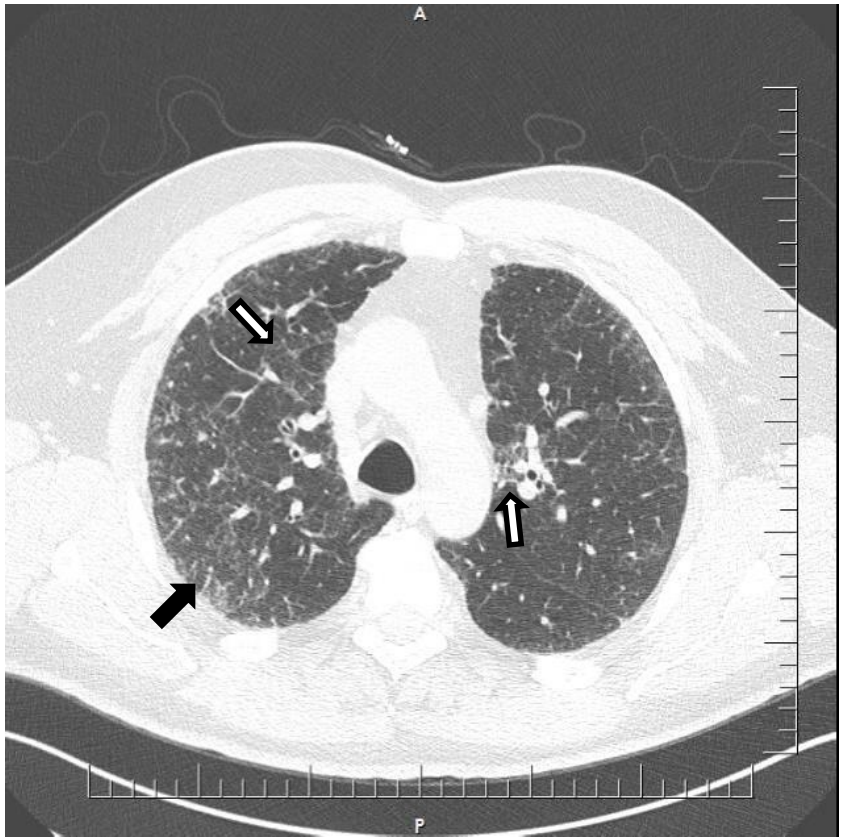
- BP = 132/68, HR = 63 , RR = 16, SpO2 = 90%
- on 2L of oxygen

- Pertinent findings
  - Inspiratory crackles at bases bilaterally
  - No edema, clubbing, skin thickening or rash
  - No joint deformities or evidence for synovitis

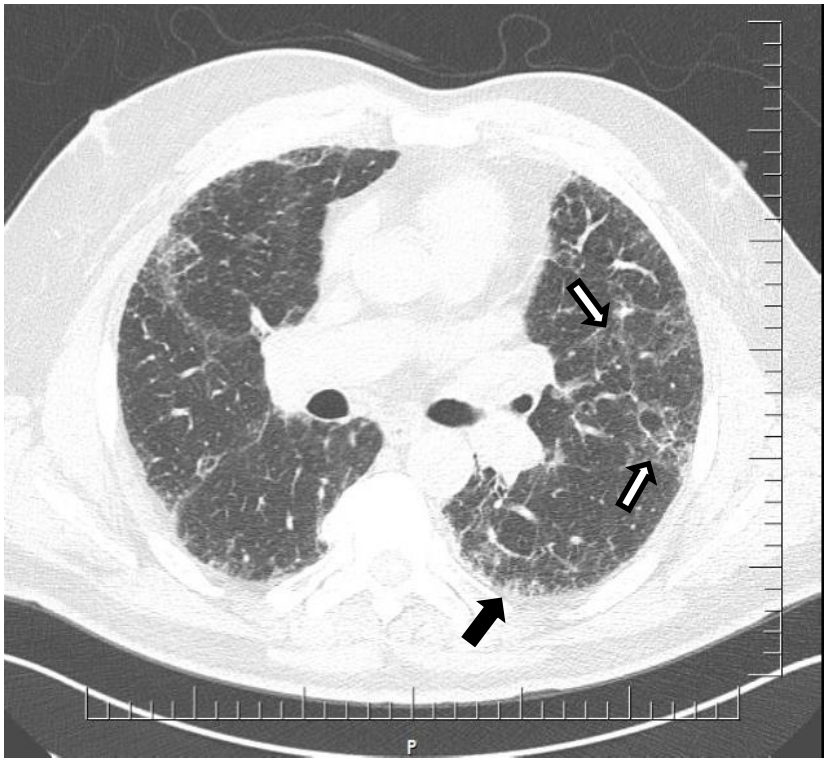
Data

- PFTs
  - TLC = 5.65 (76% of predicted)
  - FVC = 3.33 (62% of predicted)
  - FEV1 = 3.03 (74% of predicted)
  - FEV1/FVC = 91%
  - DLCO = 24.39 (53% of predicted)
  - DL/VA = 4.42 (85% of predicted)

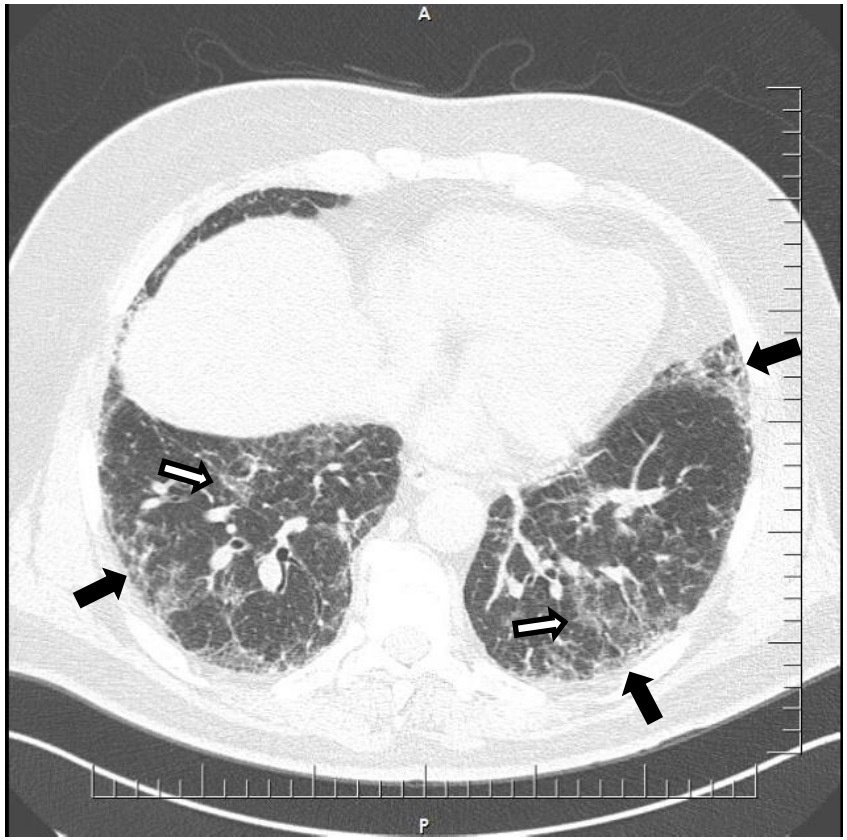
Eugene HRCTs



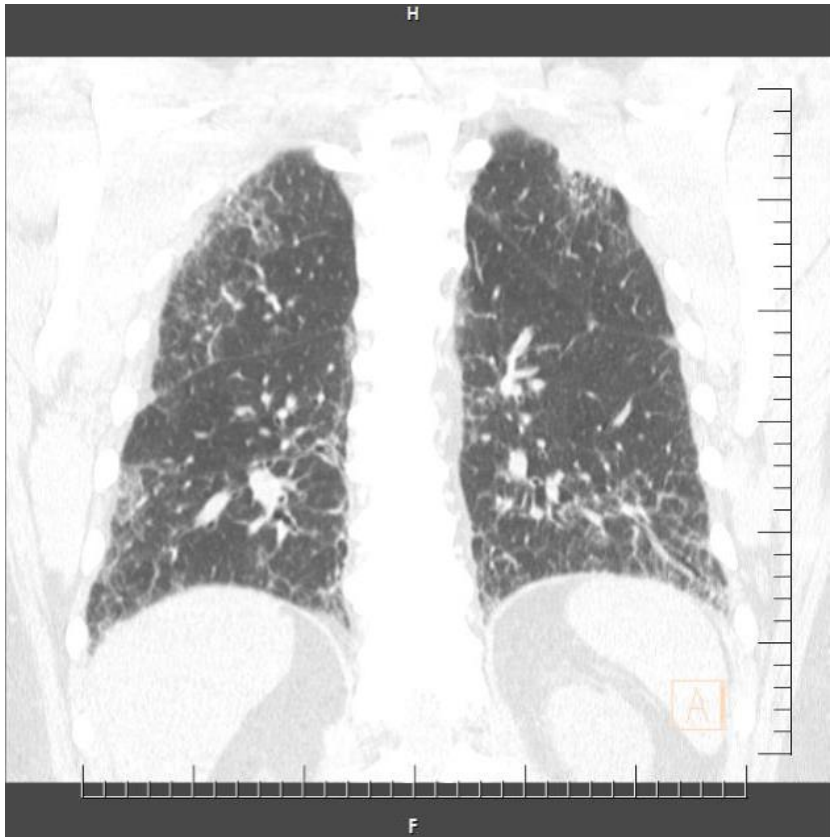
Eugene HRCTs



Eugene HRCTs

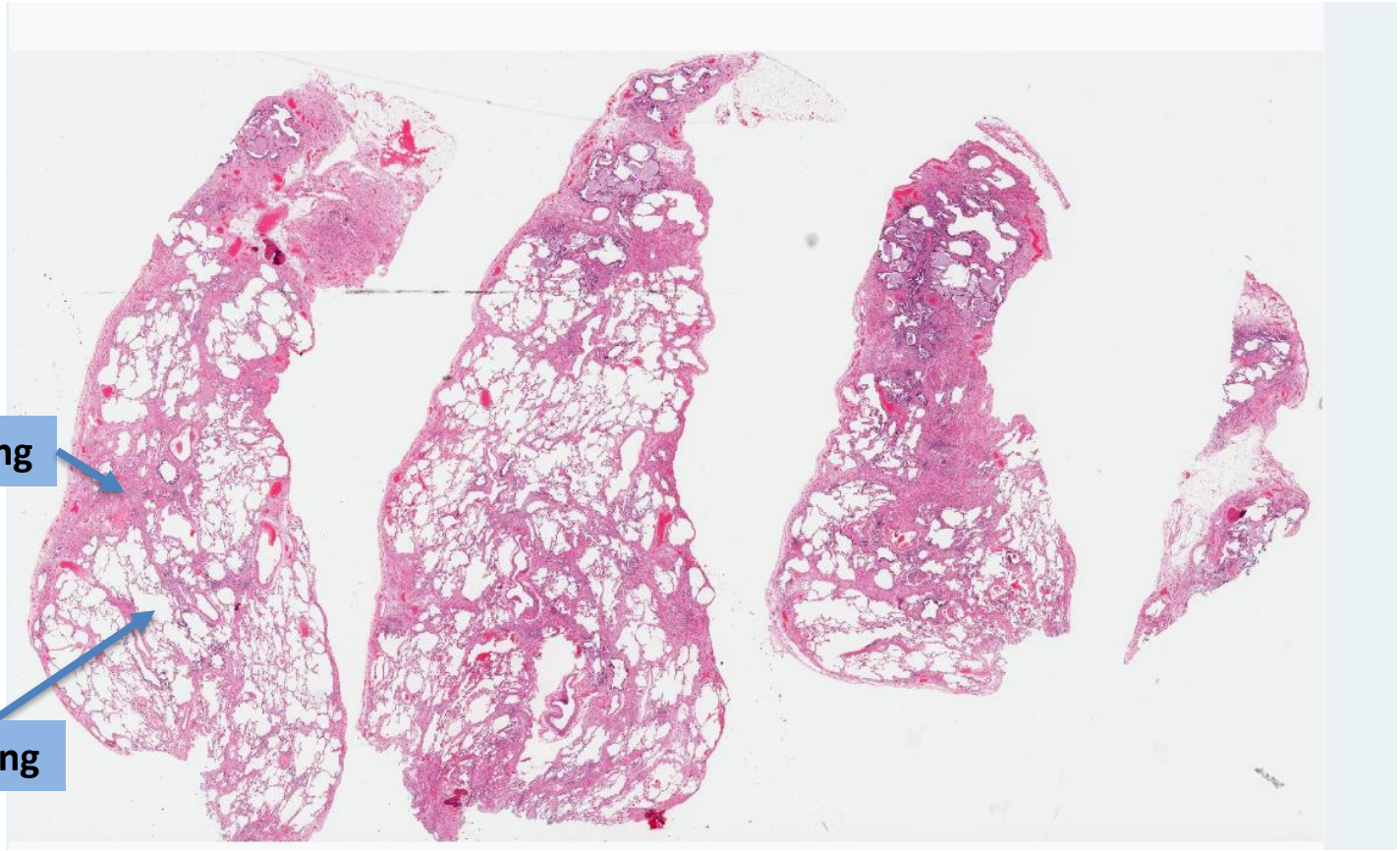


Eugene HRCTs

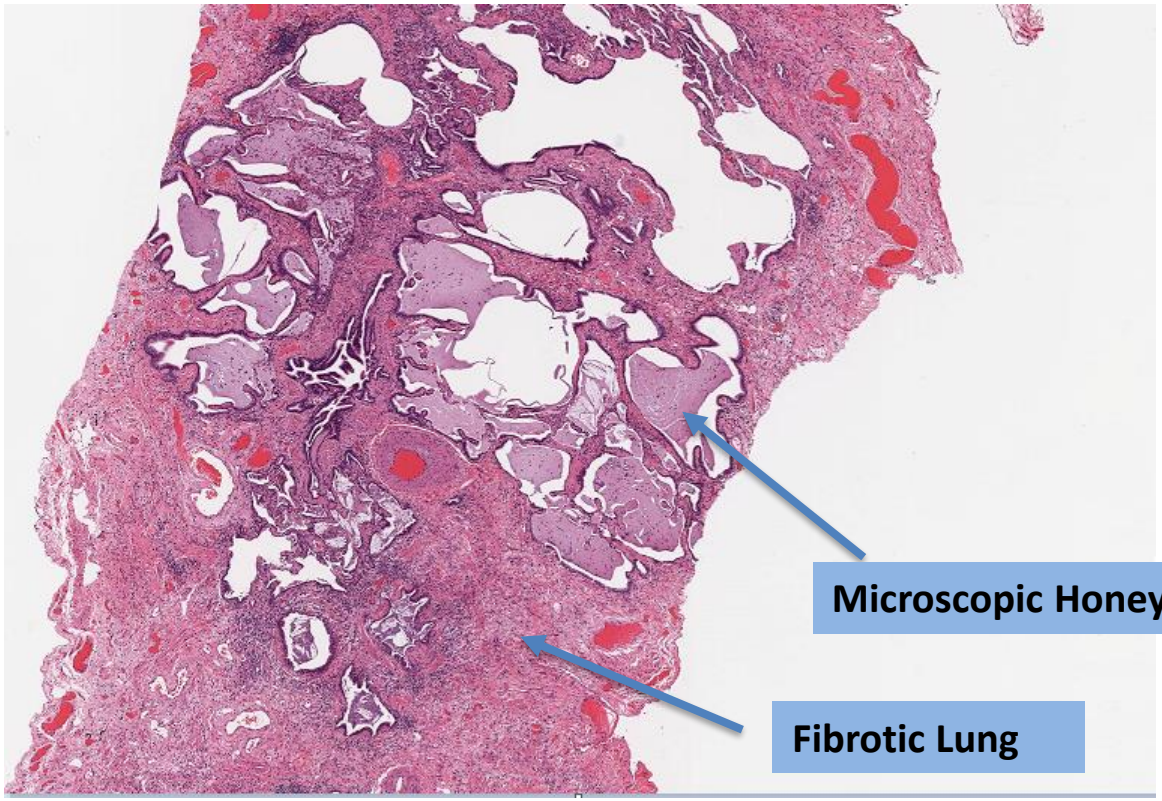


What can be concluded from Eugene's imaging?

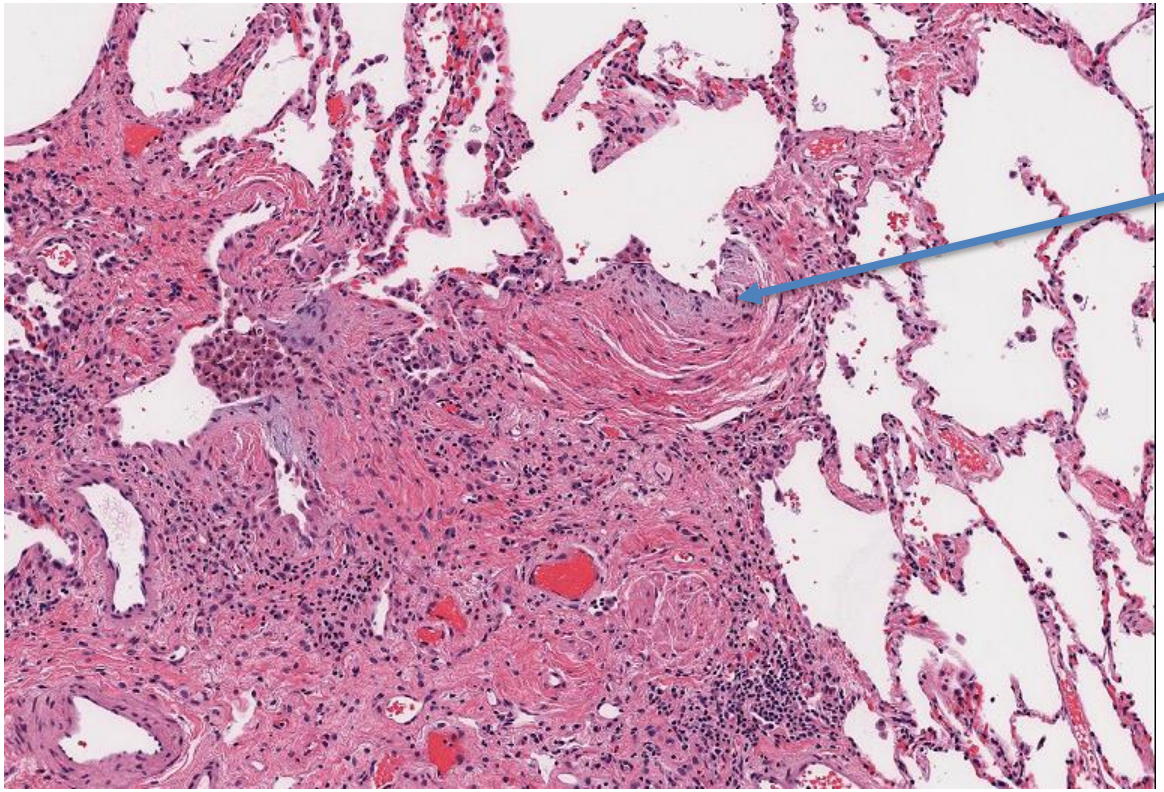
Eugene Pathology
Temporal Heterogeneity



Eugene Pathology



Eugene Pathology



Case 2: Ina

Ina: Presentation

- Ina is a 56-year-old female
- She presents with progressive dyspnea and cough for two years
  - She can do her ADLs without breathlessness, but any other activities cause dyspnea
  - The cough is worse when she is at home
- She has some joint pain in the distal finger joints bilaterally

History

- **PMHx**
  - Breast cancer diagnosed in 2012 treated with Cytoxan and radiation to the right breast
  - Hypothyroidism
  - GERD
- **Medications**
  - Tamoxifen
  - Synthroid (levothyroxine)
  - Omeprazole 40 mg orally per day
- **SHx**
  - Non-smoker
- **FHx**
  - Mother with osteoarthritis and h/o breast cancer
- **Environmental/occupational Hx**
  - She became a veterinary technician (a life-long dream) after her diagnosis of breast cancer

History

- Signs or symptoms of a systemic autoimmune disorder?
- Clinically relevant exposures (occupational and environmental)?
- Drugs that may account for the presence of lung disease?
- Relevant family history?

Physical Exam

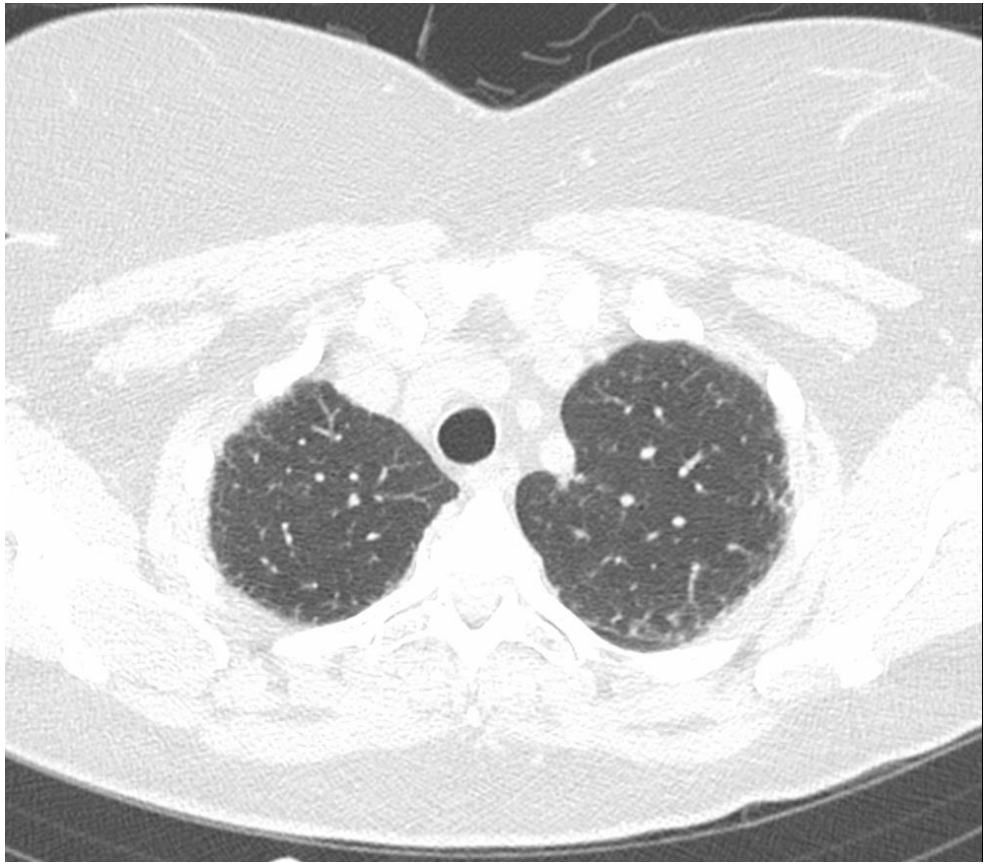
- BP = 122/73, HR = 82 , RR = 16, SpO2 = 92% on RA
- Pertinent findings
  - Inspiratory crackles at bases bilaterally and occasional inspiratory squeaks
  - No edema, clubbing, skin thickening or rash
  - Hands with Heberden's nodes in her second and third distal interphalangeal joints

Data

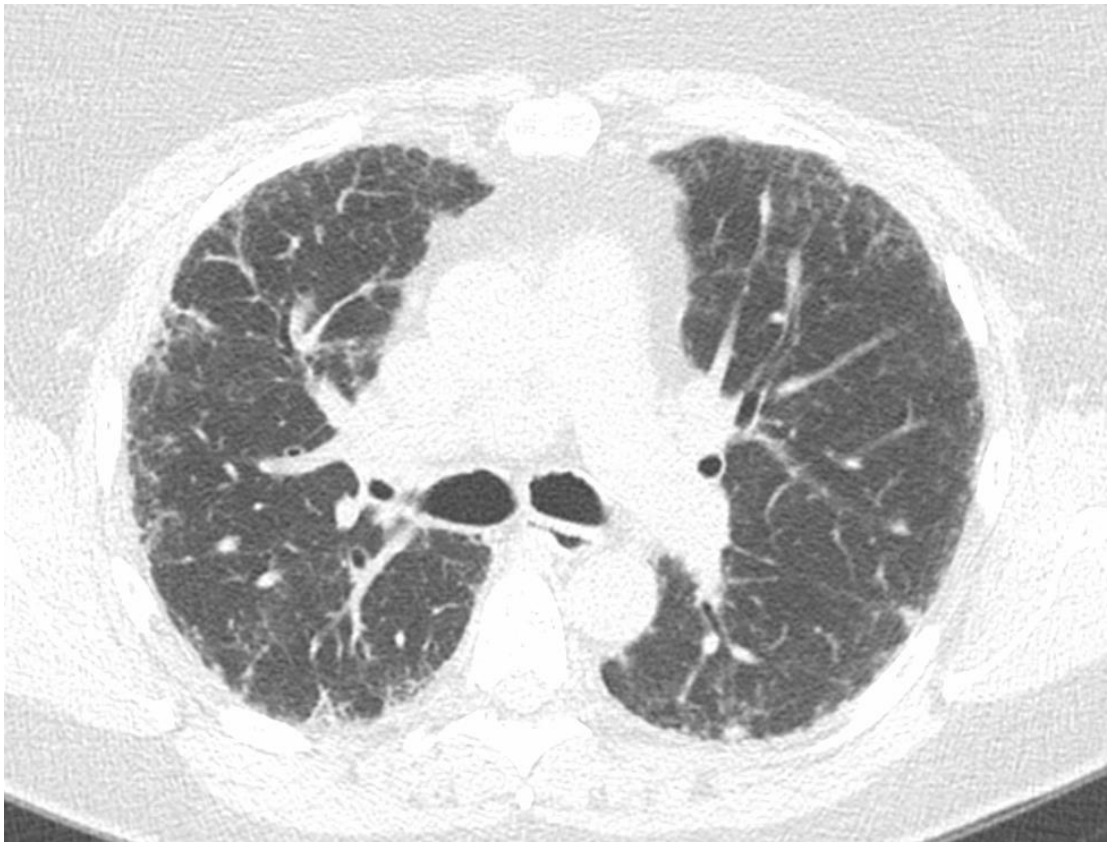
- PFTs
  - TLC = 3.19 (50% of predicted)
  - RV = 2.01 (82% of predicted)
  - FVC = 1.74 (49% of predicted)
  - FEV1 = 1.44 (50% of predicted)
  - FEV1/FVC = 89%
  - DLCO = 15.18 (58% of predicted)
  - DL/VA = 5.51 (106% of predicted)

- Oxygen titration study reveals she needs 2L of oxygen to maintain saturations > 90%

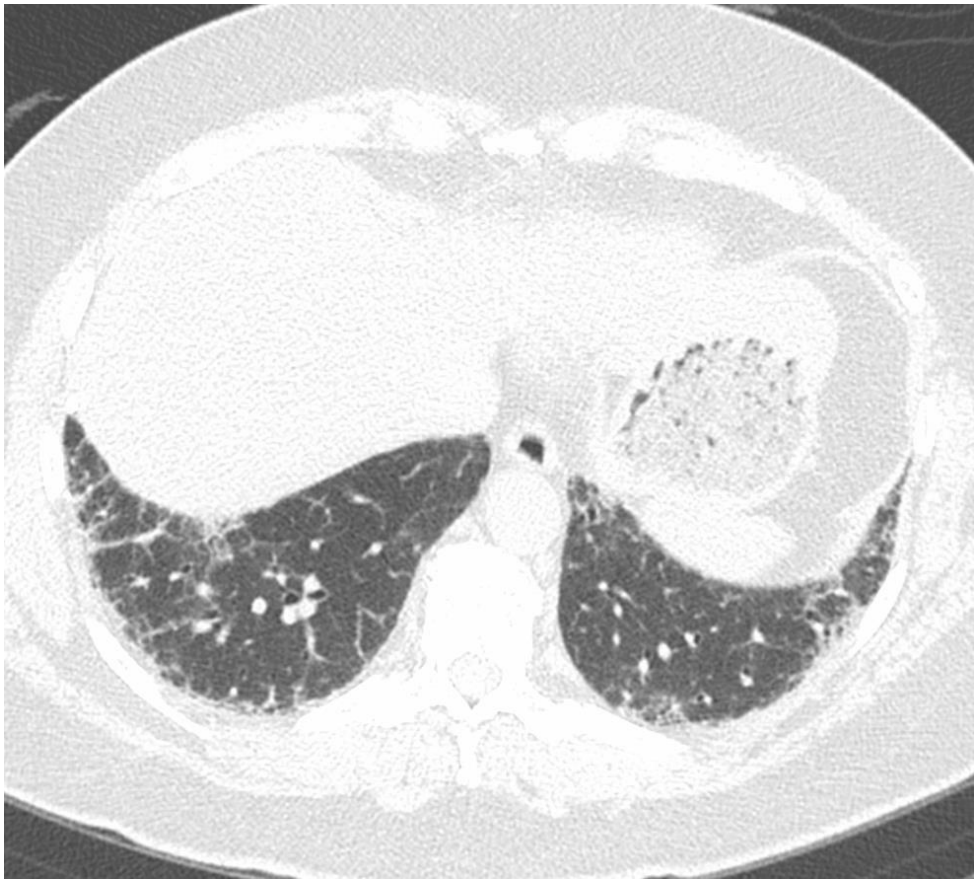
Ina HRCTs



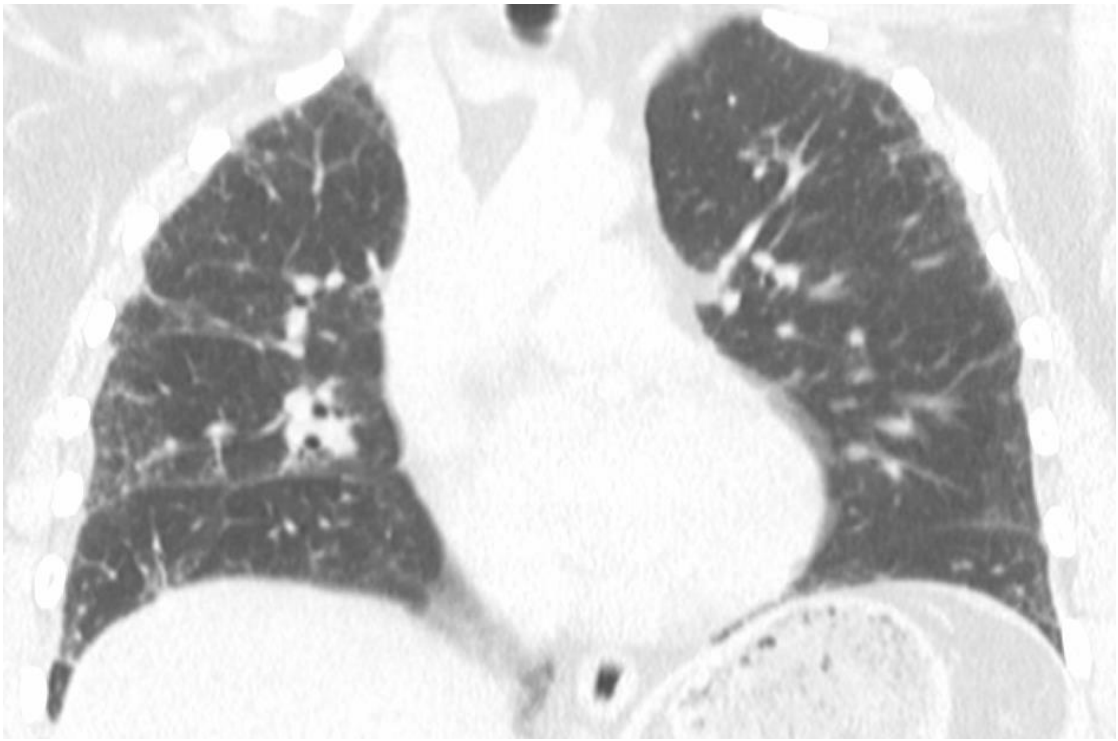
Ina HRCTs



Ina HRCTs

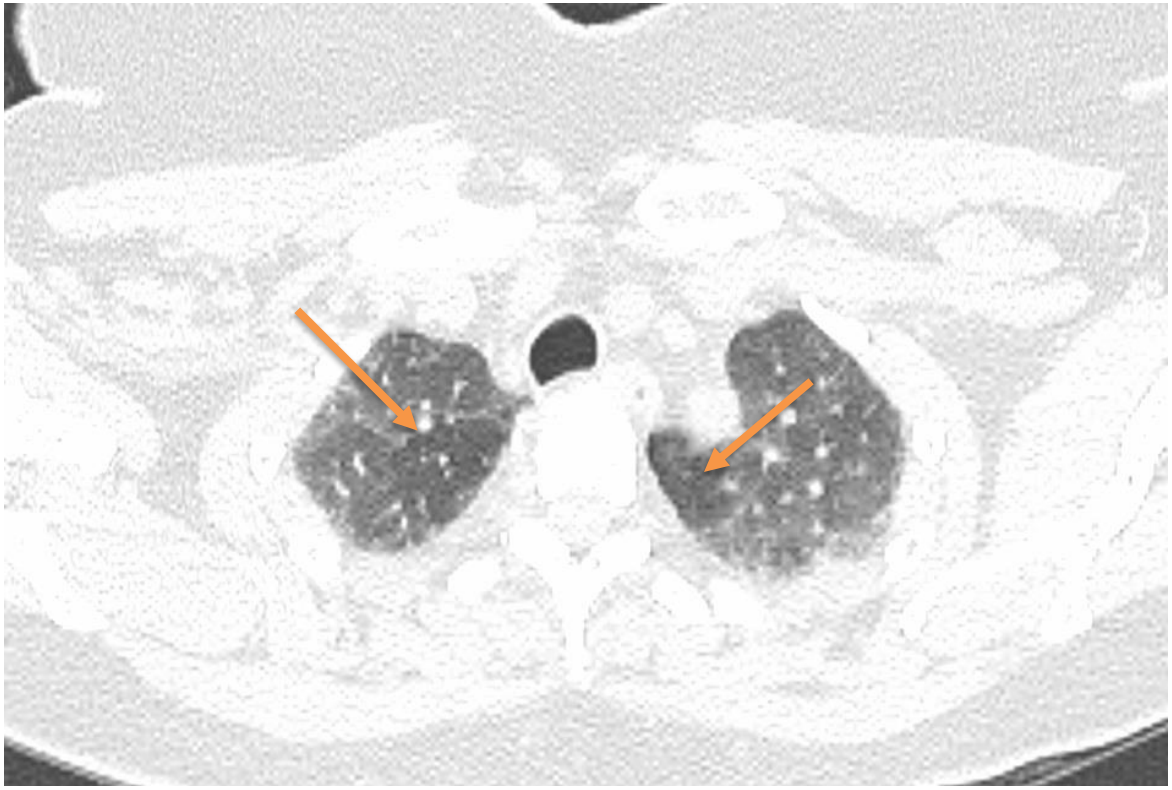


Ina HRCTs



What can be concluded from Ina's imaging?

Ina HRCTs



What can be concluded from Ina's expiratory imaging?

Additional Information

- **SEROLOGIES**
  - ANA (Antinuclear antibodies) = **negative**
  - SCL-70 antibody = **negative**
  - SSA antibody = **negative**
  - SSB antibody = **negative**
  - Rheumatoid factor = **negative**
  - CCP antibody **= negative**
  - CK and aldolase = **normal**
  - Myositis panel (includes Mi-2, Ku, PM-Scl100, PM-Scl175, Jo-1, SRP, PL-7, PL-12, EJ, OJ, Ro52) = **negative**
- **PRECIPITINS TO MOLDS**
  - **Negative**
- **PRECIPITINS TO BIRDS**
  - ☒ **Cockatiel droppings**
  - ☒ **Cockatiel serum**
  - ☒ **Macaw droppings**
  - ☒ **Macaw serum**
- **BRONCHOSCOPY (BAL)**
  - Macrophages: **45%**
  - Lymphocytes: **52%**
  - Neutrophils: **2%**
  - Eosinophils: **1%**

IPF Diagnosis: BAL Cellular Analysis

	2018 Guideline	
	HRCT Pattern of Probable UIP*, Indeterminate for UIP, and Alternative Diagnosis	HRCT Pattern of UIP*
BAL cellular analysis	We suggest performing BAL cellular analysis (conditional)	We suggest *NOT* performing BAL cellular analysis (conditional)

	BAL Cellular Analysis		
	Cell Population	**Healthy Individuals**	**IPF Relative to Other ILDs**
Ina: 2%	Neutrophils	≤ 3%	**IPF: 5.9% to 22.08%** Higher than HP, cellular NSIP, eosinophilic pneumonia
Ina: 45%	Macrophages	> 85%	**IPF: 49.18% to 83%** Higher than NSIP, eosinophilic pneumonia
Ina: 1%	Eosinophils	≤ 1%	**IPF: 2.39% to 7.5%** Lower than patients with eosinophilic pneumonia
Ina: 52%	Lymphocytes	10% to 15%	**IPF: 7.2% to 26.7%** Lower than patients with NSIP, sarcoidosis or COP

Raghu G et al. *Am J Respir Crit Care Med*. 2018 Sep 1;198(5):e44-e68.

Case 3: Margaret

Margaret: Presentation

- Margaret is a 58-year-old female
- She has had mild breathlessness and cough for the past six months
  - She runs 5Ks and has noticed her times are declining
- She has no other systemic complaints

History

- **PMHx**
  - Allergies
  - Chronic sinusitis
- **Medications**
  - Zyrtec (cetirizine)
  - Nasal washes
  - Flonase (fluticasone propionate nasal spray)
- **SHx**
  - Non-smoker
- **FHx**
  - Mother with rheumatoid arthritis
- **Environmental/occupational Hx**
  - No exposures

History

- Signs or symptoms of a systemic autoimmune disorder?
- Clinically relevant exposures (occupational and environmental)?
- Drugs that may account for the presence of lung disease?
- Relevant family history?

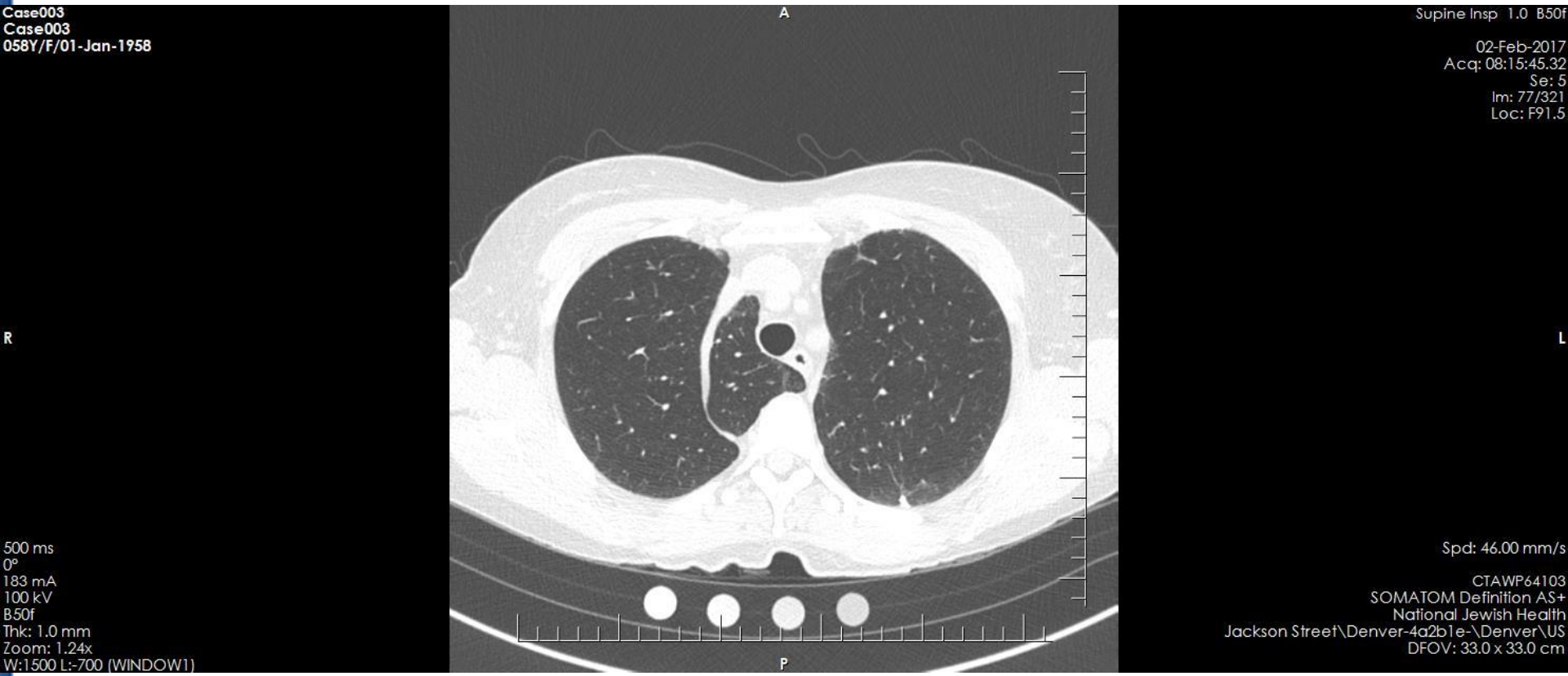
Physical Exam

- BP = 117/77, HR = 75, RR = 18, SpO2 = 97% on RA
- Pertinent findings
  - Occasional faint late inspiratory crackles at the bases
  - No edema, clubbing, skin thickening or rash

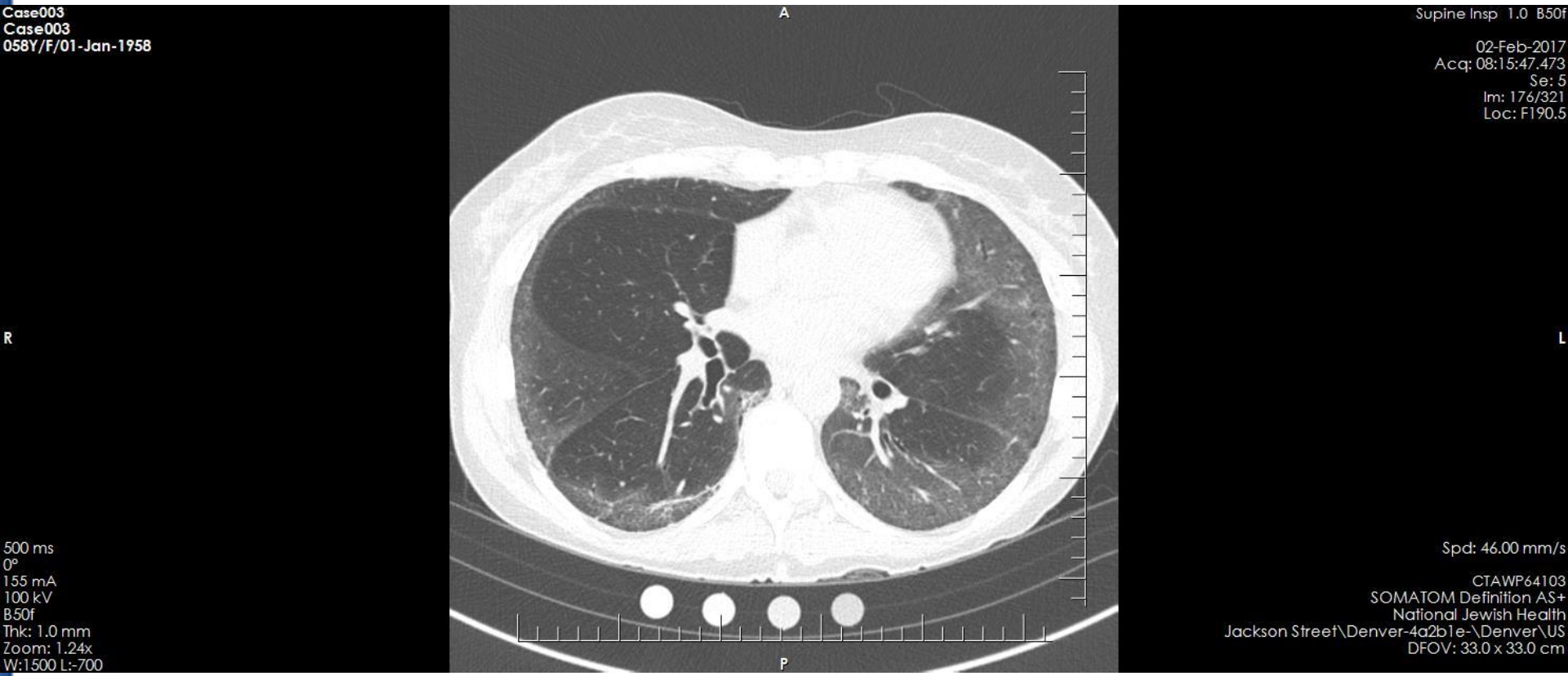
Data

- PFTs
  - TLC = 5.50 (105% of predicted)
  - FVC = 3.76 (105% of predicted)
  - FEV1 = 2.88 (104% of predicted)
  - FEV1/FVC = 77%
  - DLCO = 19.71 (76% of predicted)
  - DL/VA = 4.19 (81% of predicted)

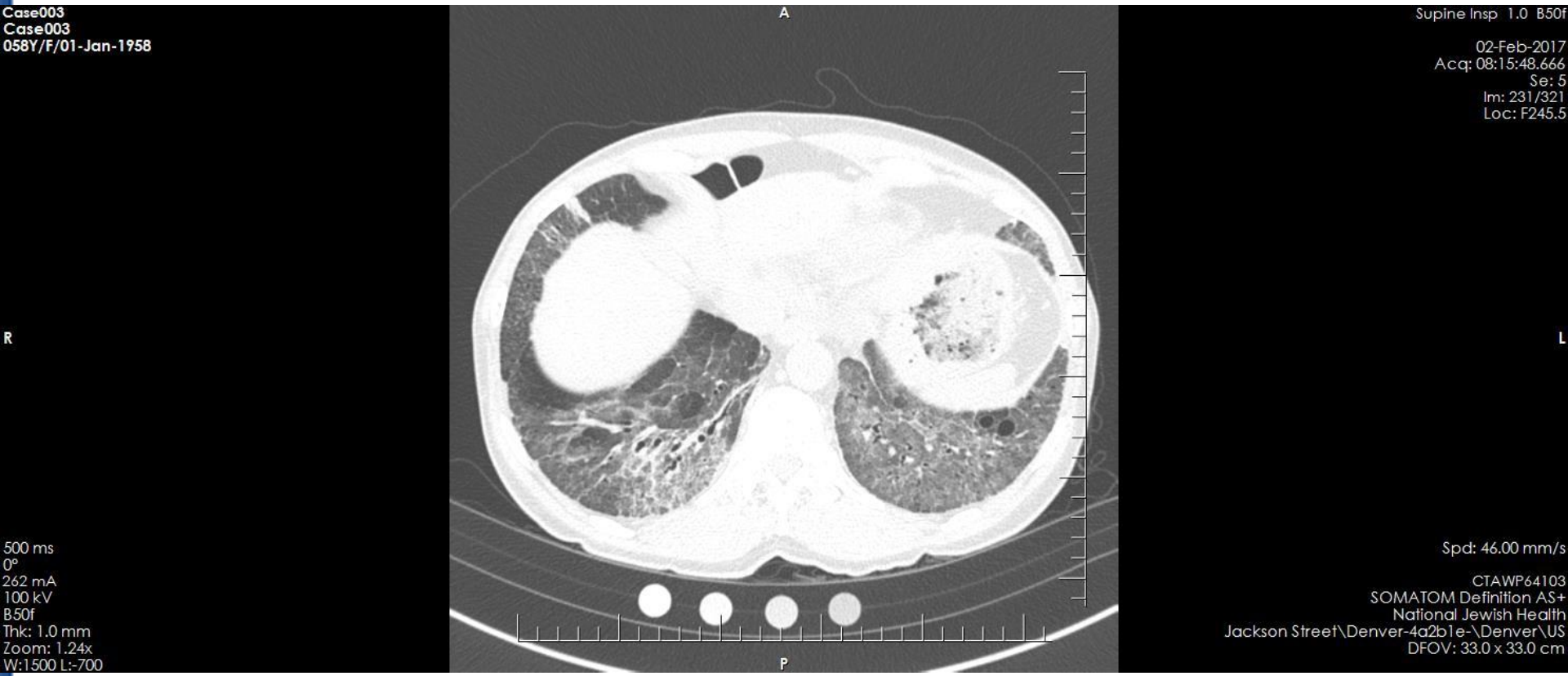
Margaret HRCTs



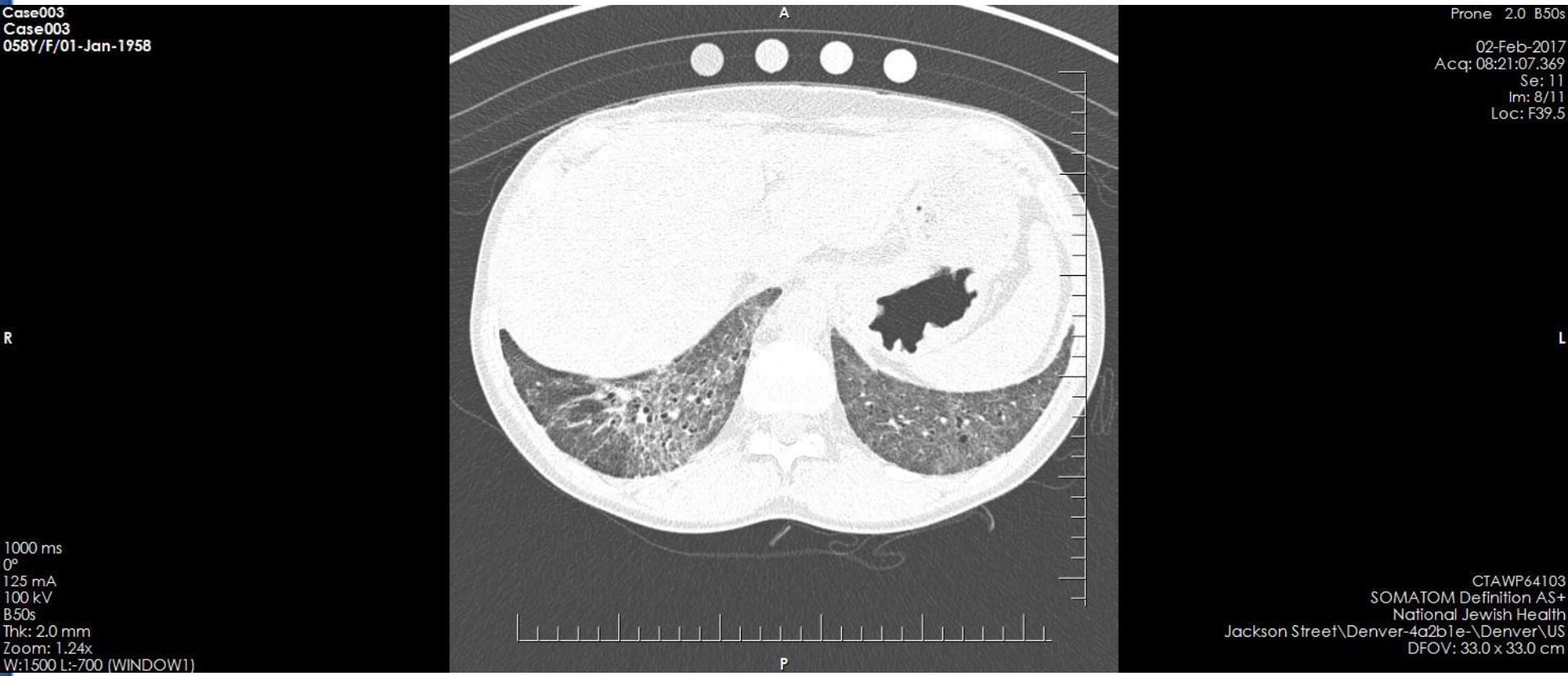
Margaret HRCTs



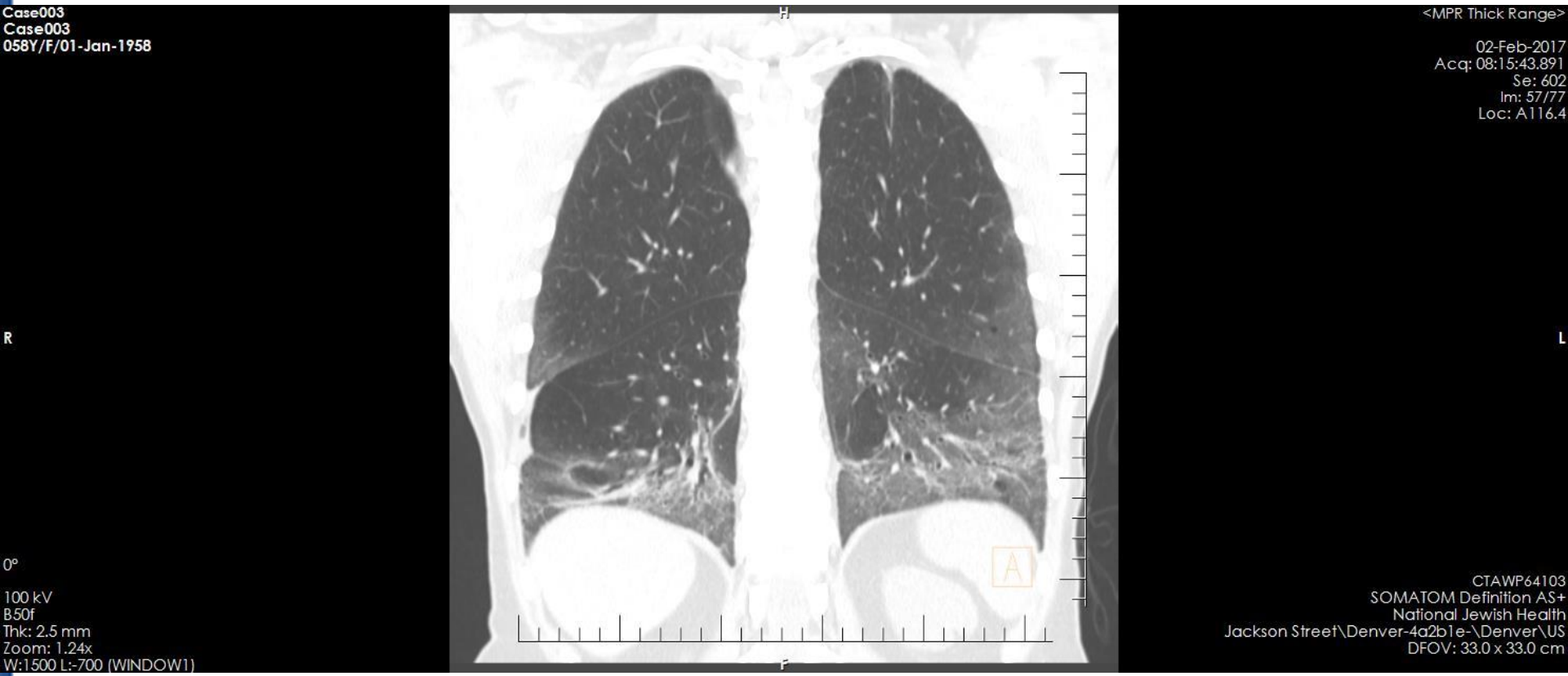
Margaret HRCTs



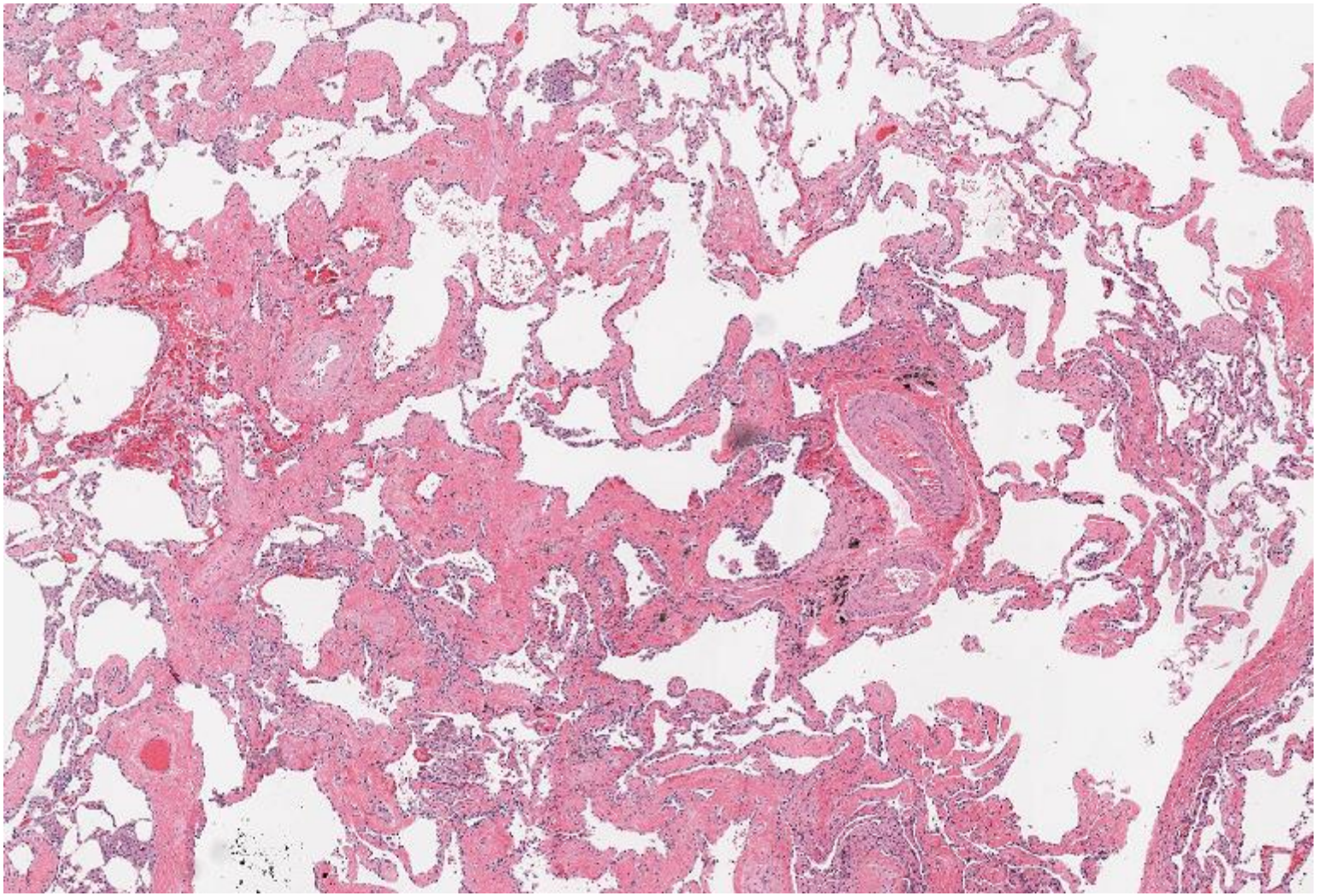
Margaret HRCTs



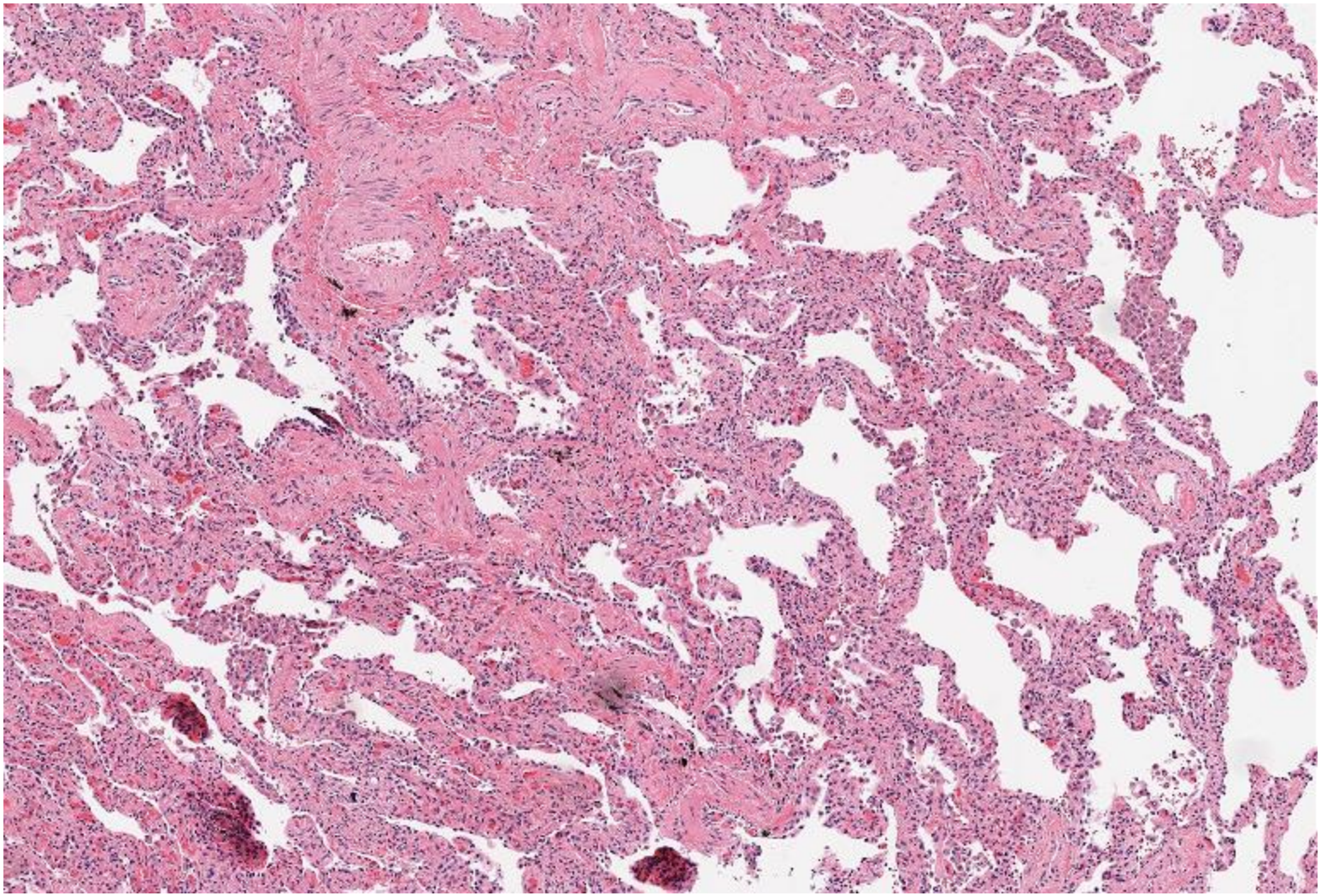
Margaret HRCTs



Margaret: Pathology



Margaret: Pathology



Case 4: Tracy

Tracy: Presentation

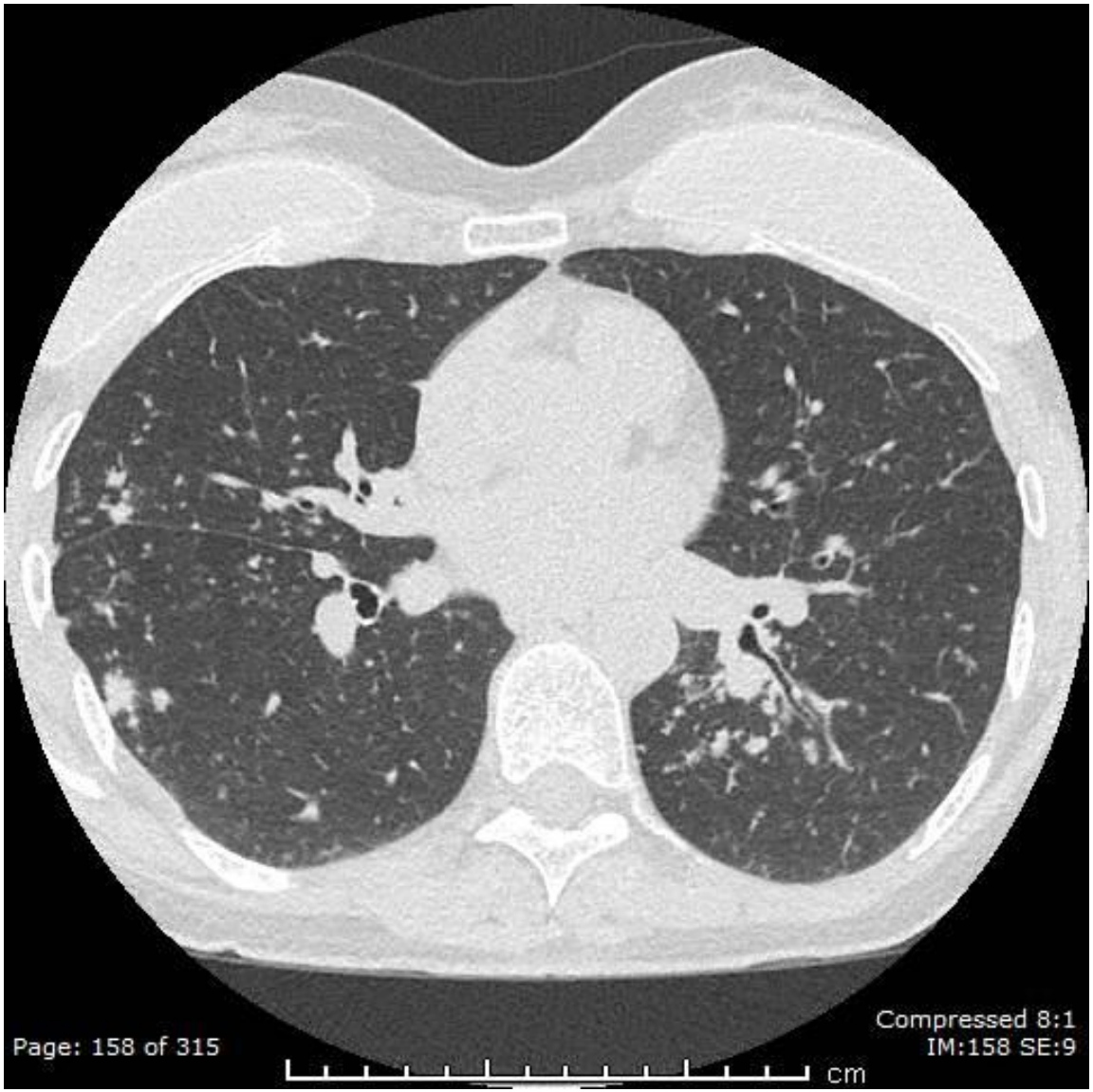
- 53-year-old female
- Presented with chest palpitations and chronic cough
- Cardiac work up negative
- Dry cough, nonproductive, does not respond to albuterol or antitussives
- Recent travel to Alaska and the Caribbean
- No pets
- Denies abdominal pain, nausea and vomiting, headache, diarrhea, weight loss
- PE: negative

Tracy: PFTs

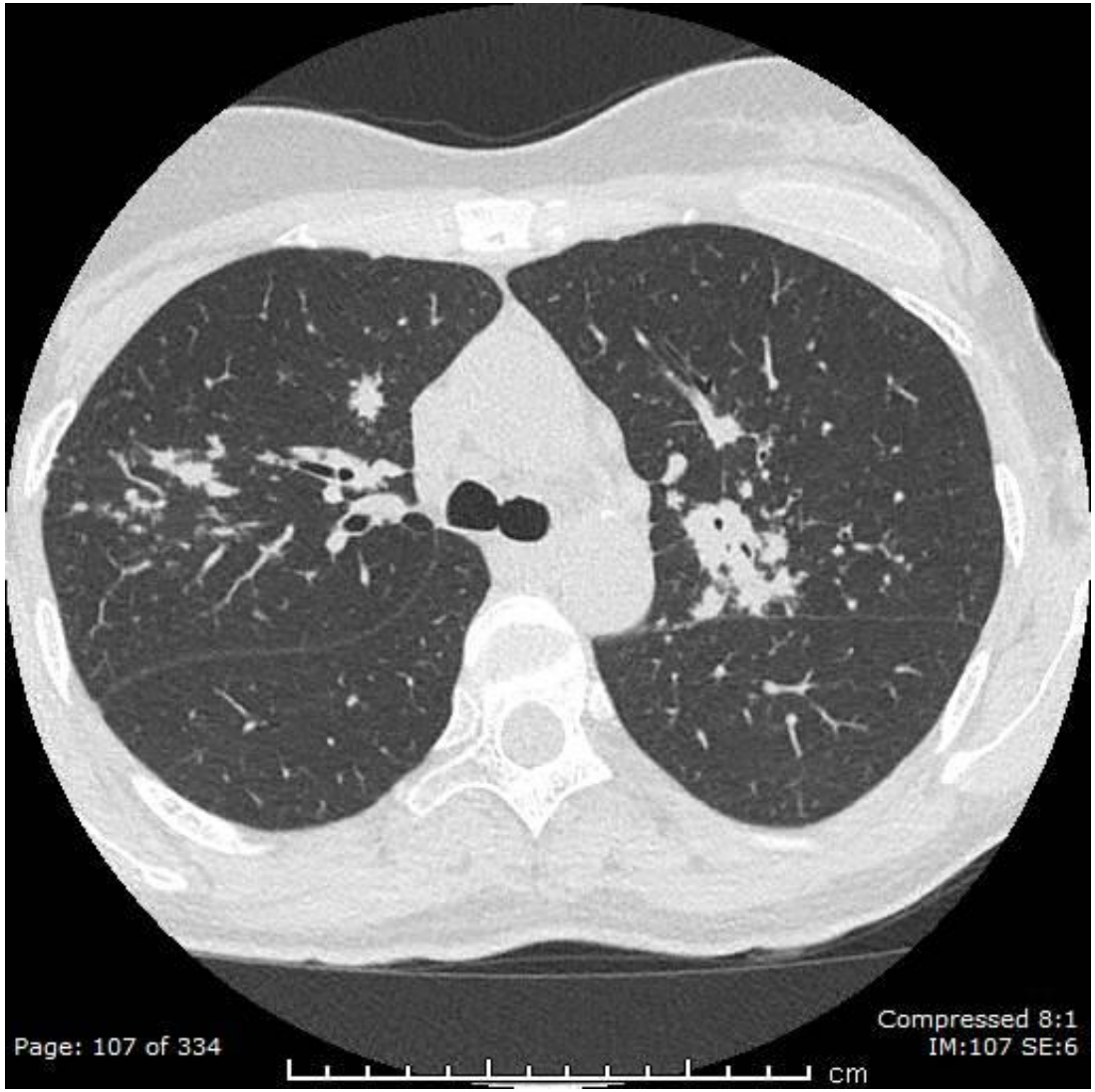
	PRE-RX		
	Pred	Actual	%Pred
SPIROMETRY			
FVC (L)	3.54	4.08	115
FEV1 (L)	2.83	3.06	108
FEV1/FVC (%)	80	75	93
FEF 25-75% (L/sec)	2.65	2.43	91
FEF Max (L/sec)	7.24	6.65	91
Expiratory Time (sec)		6.93	
FIF Max (L/sec)	3.96	5.40	136
LUNG VOLUMES			
SVC (L)	3.54	4.08	115
TLC (Pleth) (L)	5.73	6.71	117
IC (L)	2.53	2.93	115
RV (Pleth) (L)	2.06	2.62	127
RV/TLC (Pleth) (%)	37	39	105
TGV (L)	3.20	3.78	118
ERV (L)	1.01	1.16	114
DIFFUSION			
DLCOunc (ml/min/mmHg)	25.39	24.44	96
DLCOcor (ml/min/mmHg)	25.39		
DL/VA (ml/min/mmHg/L)	5.12	4.35	84
VA (L)	5.73	5.62	98
SVC (SB) (L)		3.92	
AIRWAYS RESISTANCE			
Raw (cmH2O/L/s)	1.86	0.82	43
Gaw (L/s/cmH2O)	1.03	1.25	121
sRaw (cmH2O*s)	< 4.76	4.82	
sGaw (1/cmH2O*s)	0.20	0.21	103

Tracy

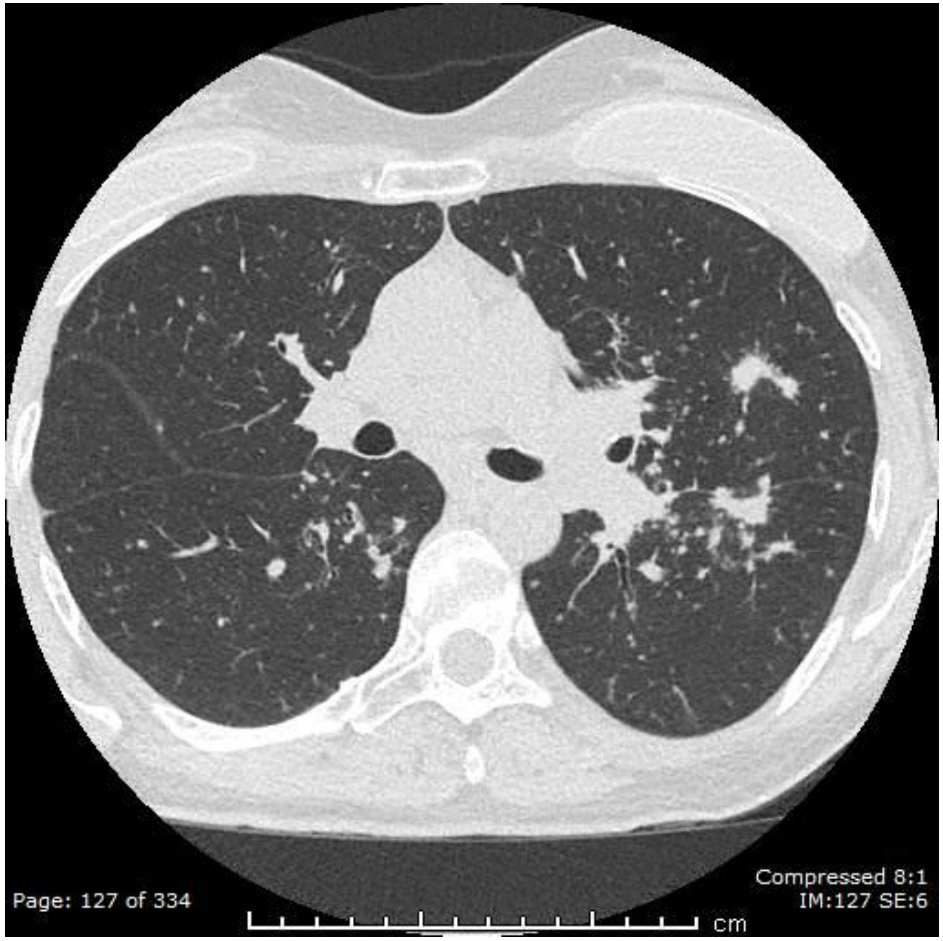
Tracy: Expiratory



Tracy



Tracy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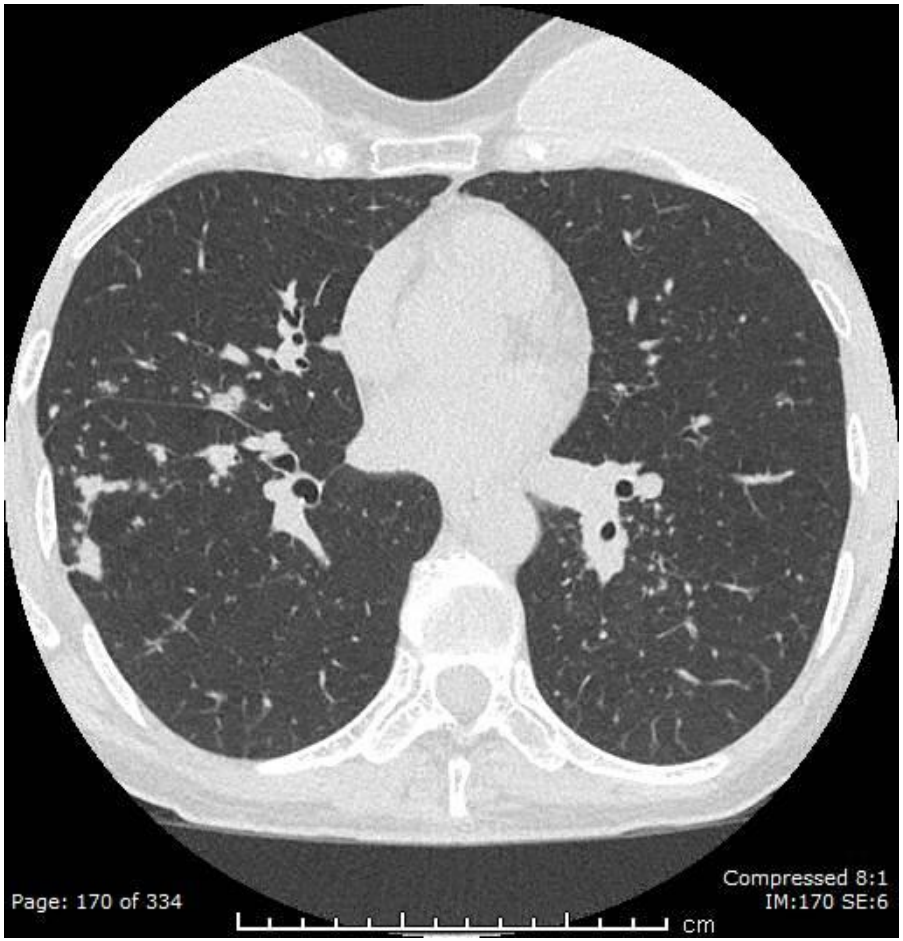
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Case 5: Rachel

Rachel: Presentation

- 49-year-old female with end-stage pulmonary fibrosis and pulmonary hypertension presenting for lung transplantation
- First presented in 2015
- Worsening progressive DOE class III-IV NYHA/WHO with dizziness and occasional wheezing
- Does not work, no alcohol, no travel, lives with children

Rachel: Medications

- Azathioprine
- Diltiazem
- Furosemide
- Prednisone
- Albuterol
- Tadalafil

Rachel

- **TEE**: LV function normal. RV enlarged. Mild tricuspid and pulmonic valve regurg. Estimated RV systolic pressure is 62 mm hg

- **Coronary**: 20% lad, 20% ramus stenosis. Right heart cath: right atrial pressure 5, RV pressure 85/18, wedge 6, CO 3.8, cardiac index 2.24

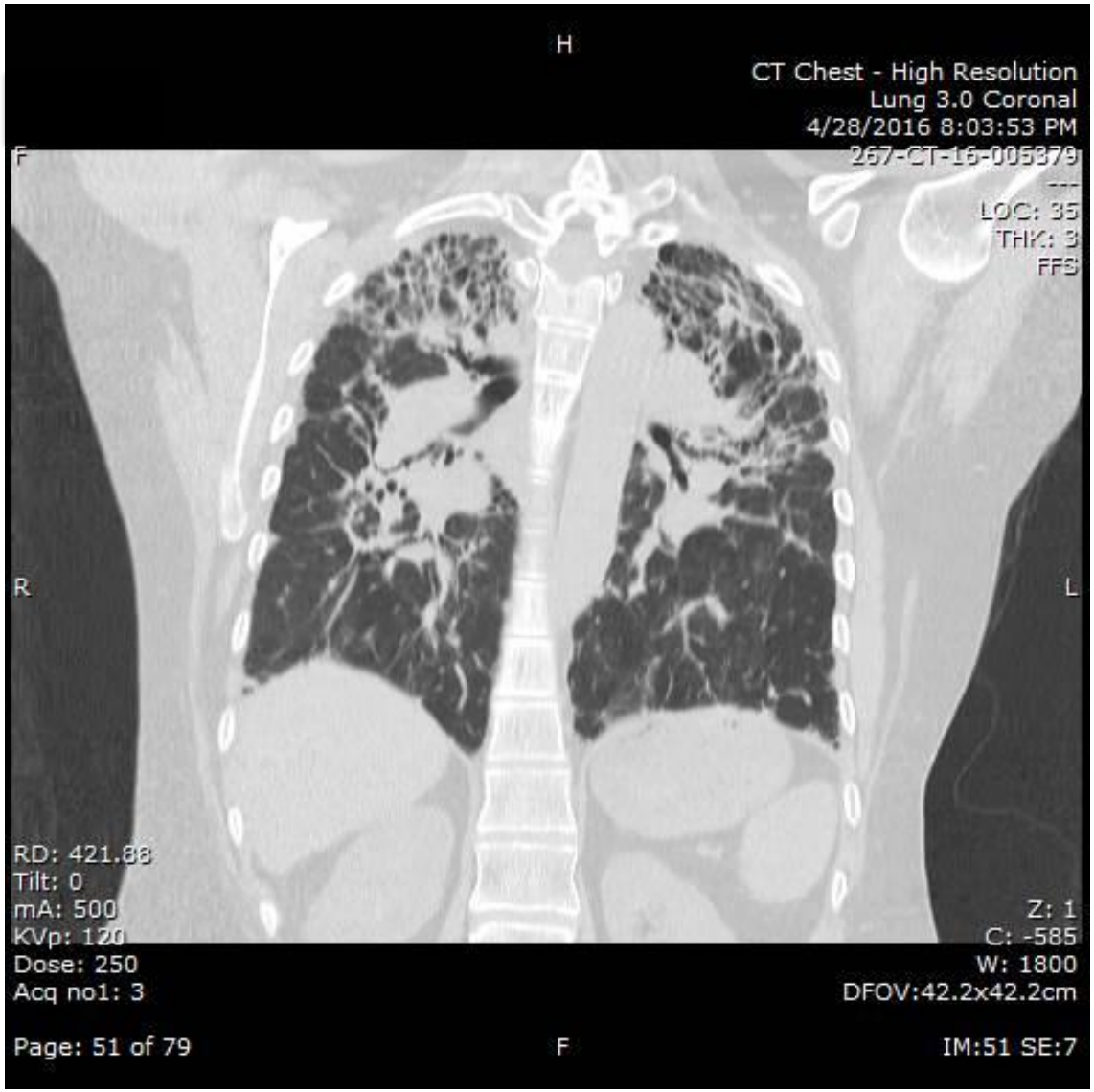
- **PE**: Positive JVD, occasional wheezes

Lung Function Studies

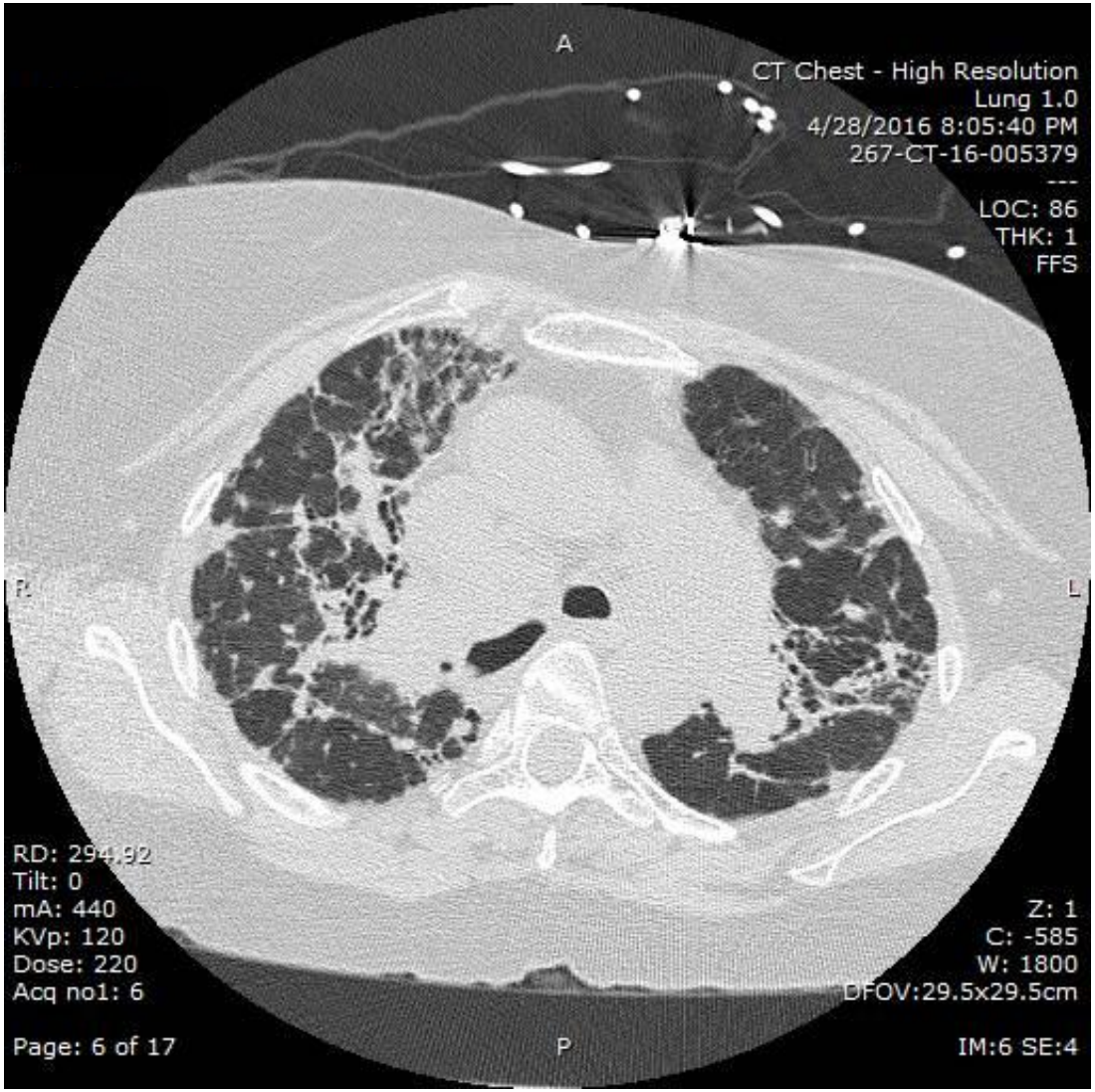
	PRE-RX			POST-RX		
	Pred	Actual	%Pred	Actual	%Pred	%Chng
SPIROMETRY						
FVC (L)	3.34	0.77	22			
FEV1 (L)	2.68	0.44	16			
FEV1/FVC (%)	81	58	71			
FEF 25-75% (L/sec)	2.68	0.20	7			
FEF Max (L/sec)	6.59	1.74	26			
Expiratory Time (sec)		9.24				
FIF Max (L/sec)	4.01	1.40	34			
BLOOD GASES						
pH	7.40	7.37				
PaCO2 (mmHg)	38-42	58.0				
PaO2 (mmHg)	87.5	48.0	54			
HCO3 (mEq/L)		33.5				
Base Excess		6.4				
SaO2 (%)		82.1				
Hgb (gm/dL)	12-18					
COHb (%)	< 1.5%					
MetHgb (%)	< 1.5%					
FIO2 (%)		21.00				

- The flow-volume curve was tiny showing extremely severe restrictive pattern
- Some airflow limitation is also noted
- BMI: 26.9

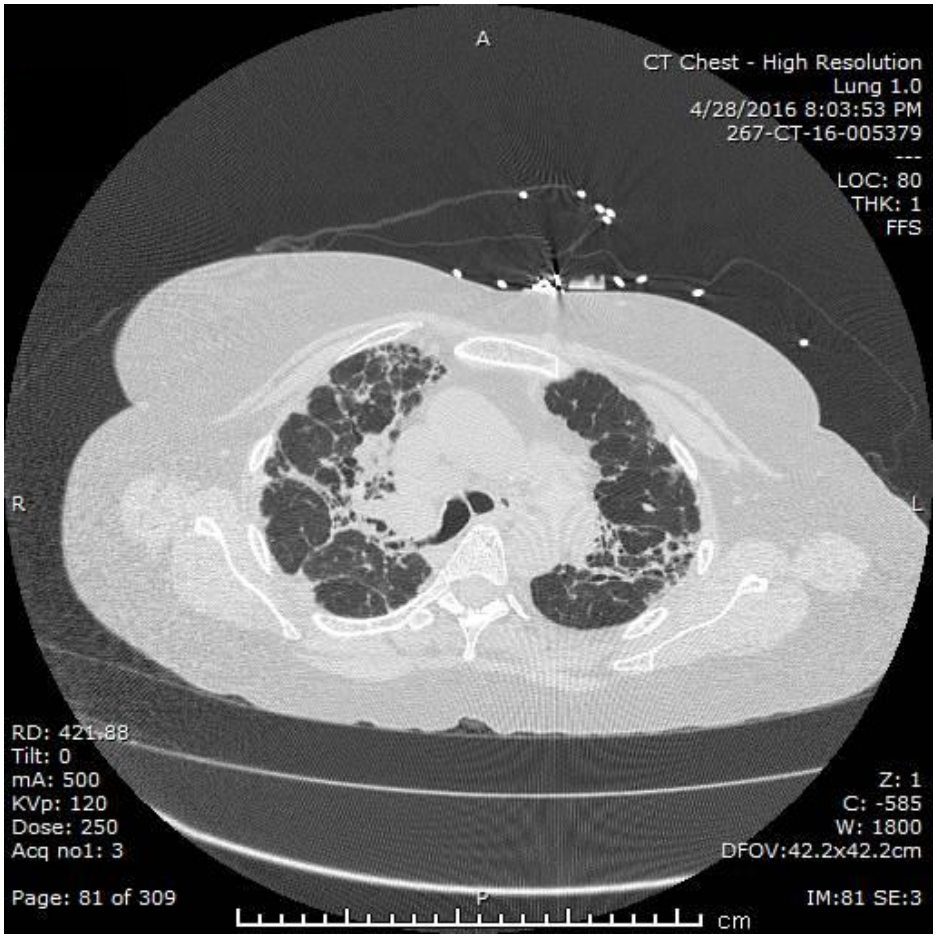
Rachel Coronal



Rachel Expiratory



Rachel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FFS
R
L
RD: 421.88
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KVp: 120
Dose: 250
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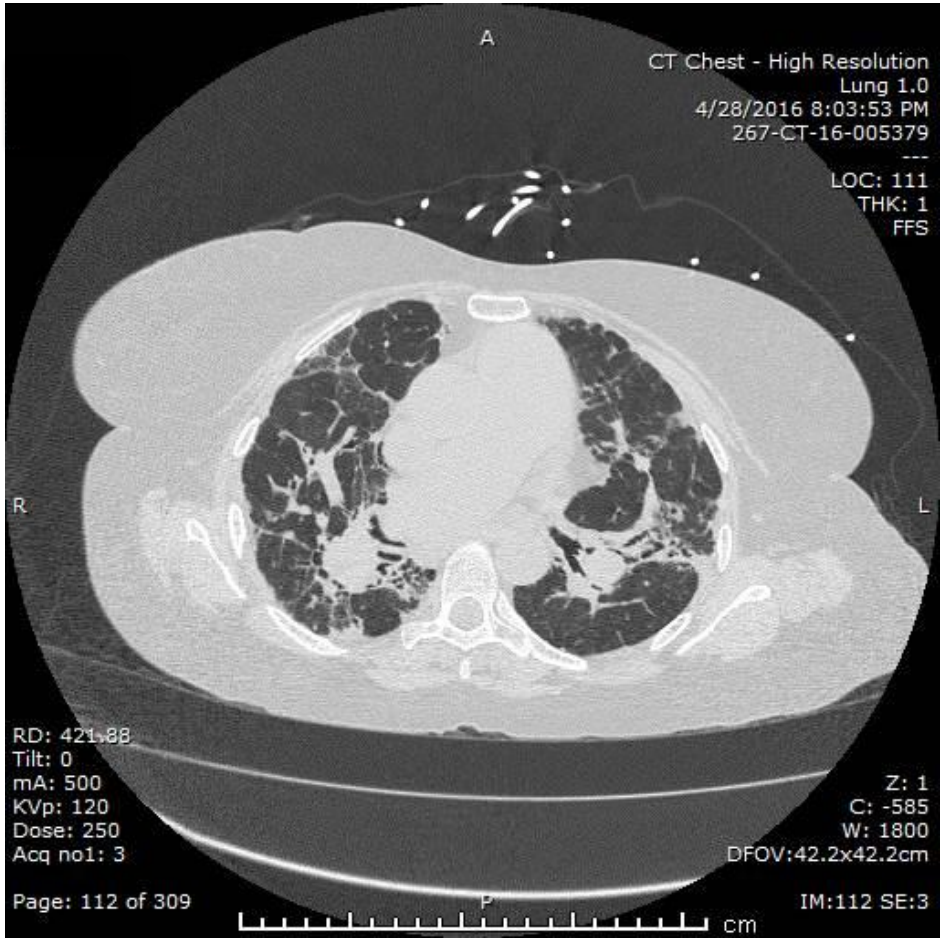
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4/28/2016 8:03:53 PM
267-CT-16-005379

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FFS
R
L
RD: 421.88
Tilt: 0
mA: 500
KVp: 120
Dose: 250
Acq no1: 3
Z: 1
C: -585
W: 1800
DFOV:42.2x42.2cm
Page: 112 of 309
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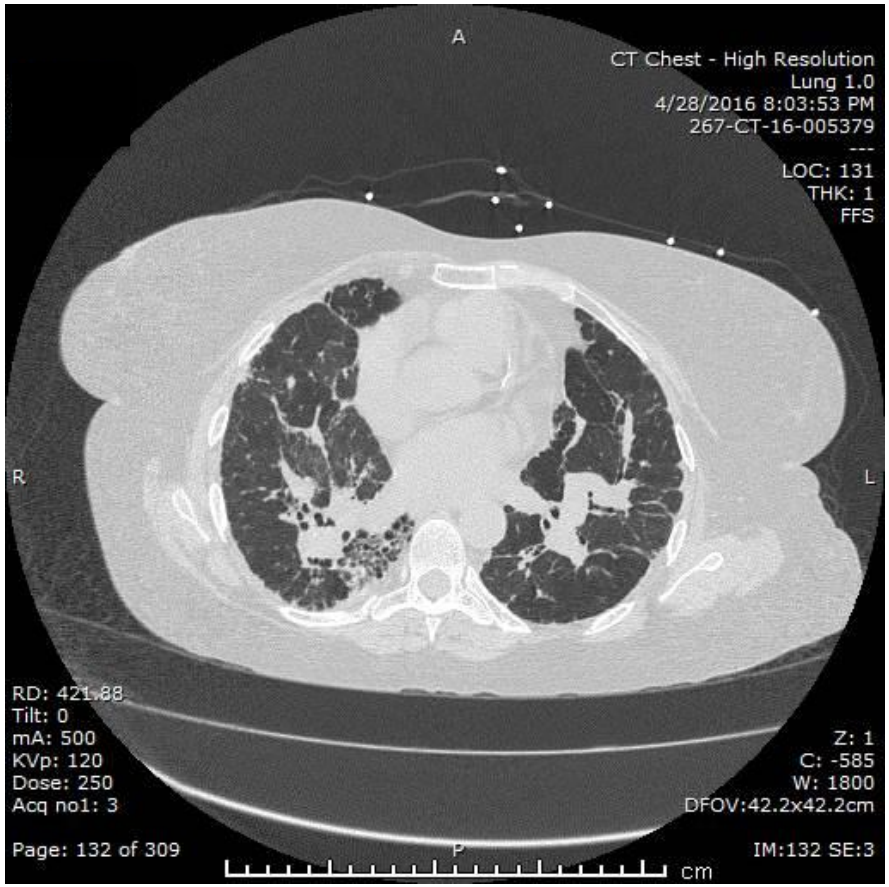
P
cm

Rachel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FFS
R
L
RD: 421.88
Tilt: 0
mA: 500
KVp: 120
Dose: 250
Acq no1: 3
Z: 1
C: -585
W: 1800
DFOV:42.2x42.2cm
Page: 132 of 309
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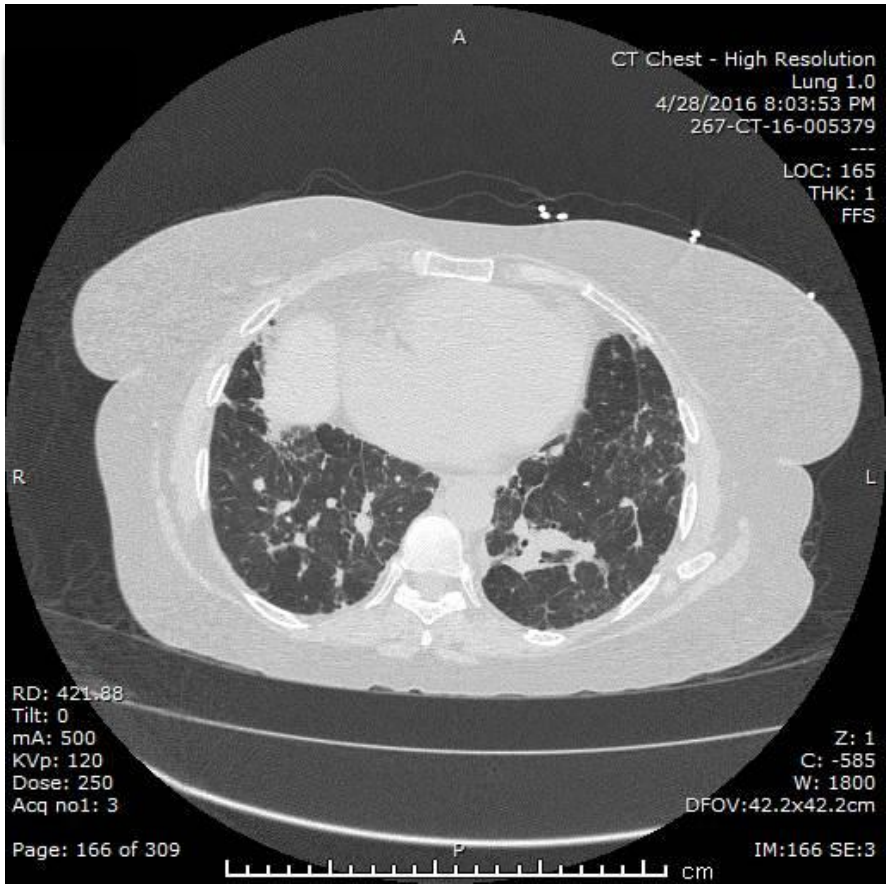
cm
IM:132 SE:3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CT Chest - High Resolution
Lung 1.0
4/28/2016 8:03:53 PM
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LOC: 165
THK: 1
FFS
R
L
RD: 421.88
Tilt: 0
mA: 500
KVp: 120
Dose: 250
Acq no1: 3
Z: 1
C: -585
W: 1800
DFOV:42.2x42.2cm
Page: 166 of 309
P
cm
IM:166 SE:3

Case 6: Sandra

- 53 year old Hispanic woman
- PMH of obesity, dm, htn history of DVT in 2017
- After a return trip in 8/2018 from El Salvador, she had symptoms of worsening SOB and worsening left lower leg swelling.
- Her previous DVT was treated with apixaban
- She reports one pregnancy miscarriage; and two kids without issue
- No history of rheumatic disease, no drug use, no liver disease. No history of lupus or family history of coagulopathy

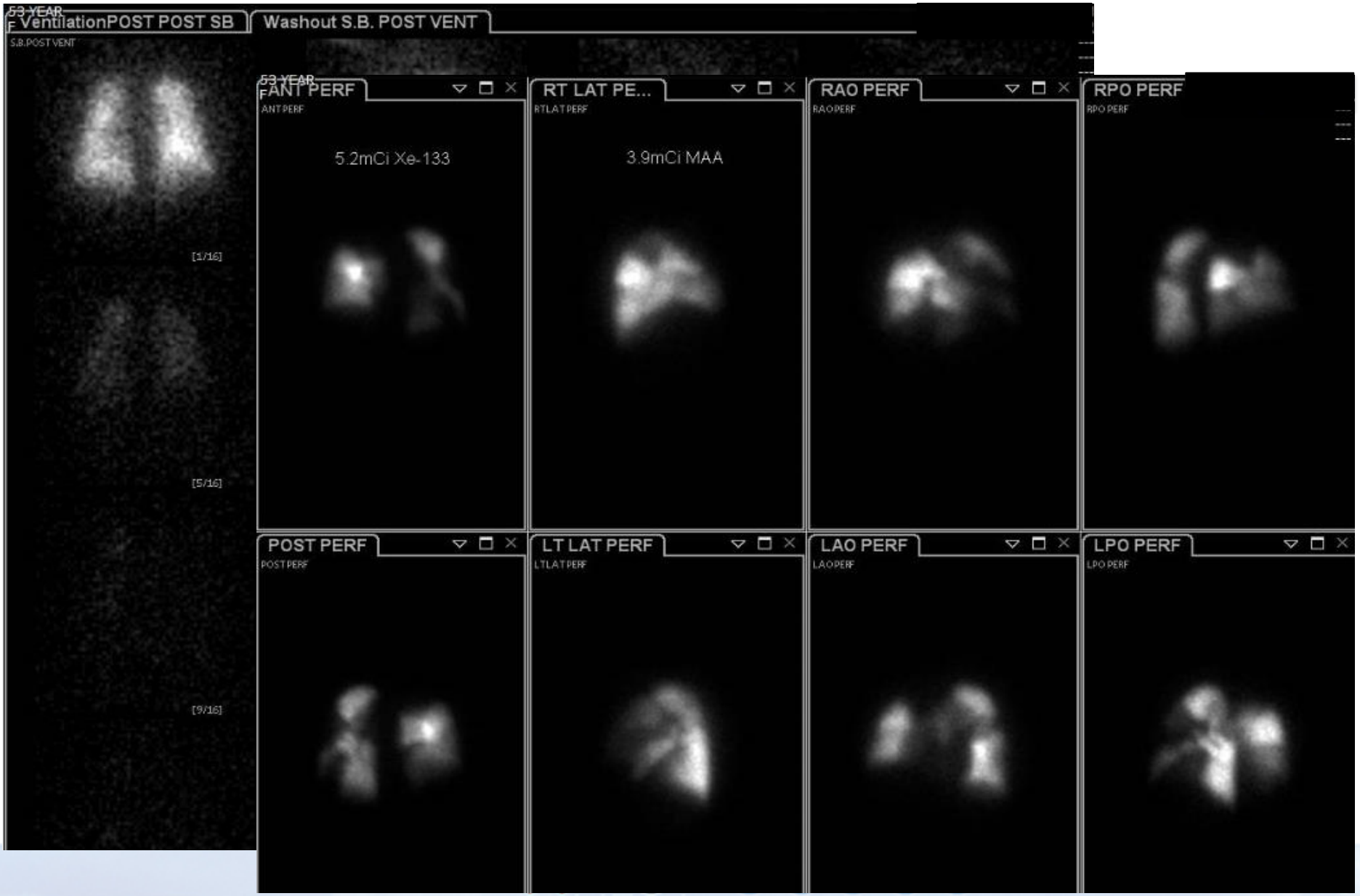
Case 6 Continued

- TTE: LV normal size and function; ejection fraction 61%
- Mild diastolic dysfunction, interventricular septal flattening during systole c/w RV pressure overload
- RV severely dilated with moderate depressed RV function
- RA severely dilated, severe tricuspid valve regurgitation with ESPAP 84 mm Hg
- RA 20, PAEDP 30 mm Hg

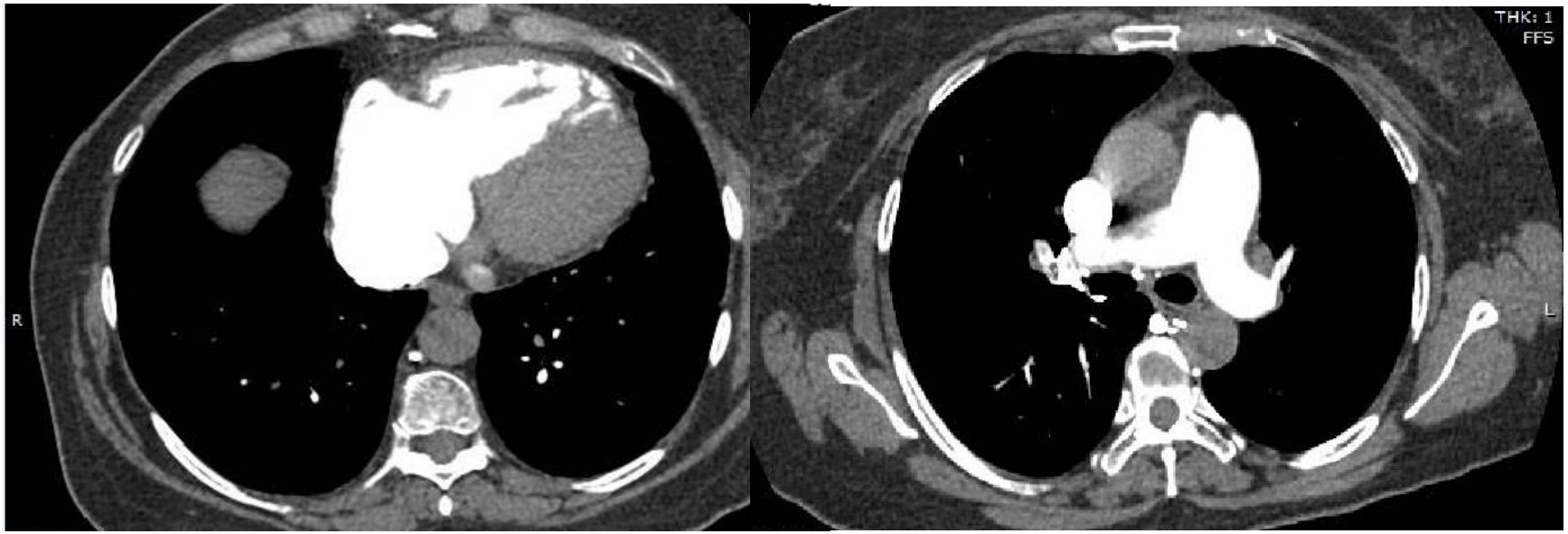
Lung Function

- PFT mild restrictive interstitial disorder, mild air trapping, moderate decrease DLCO 66%
  - FVC 2.36 (77)
  - FEV1 1.93 (78)
  - FEV1/FVC 82 (100)
  - TLC 4.2 (90)
  - DLCO 13.62 (66)

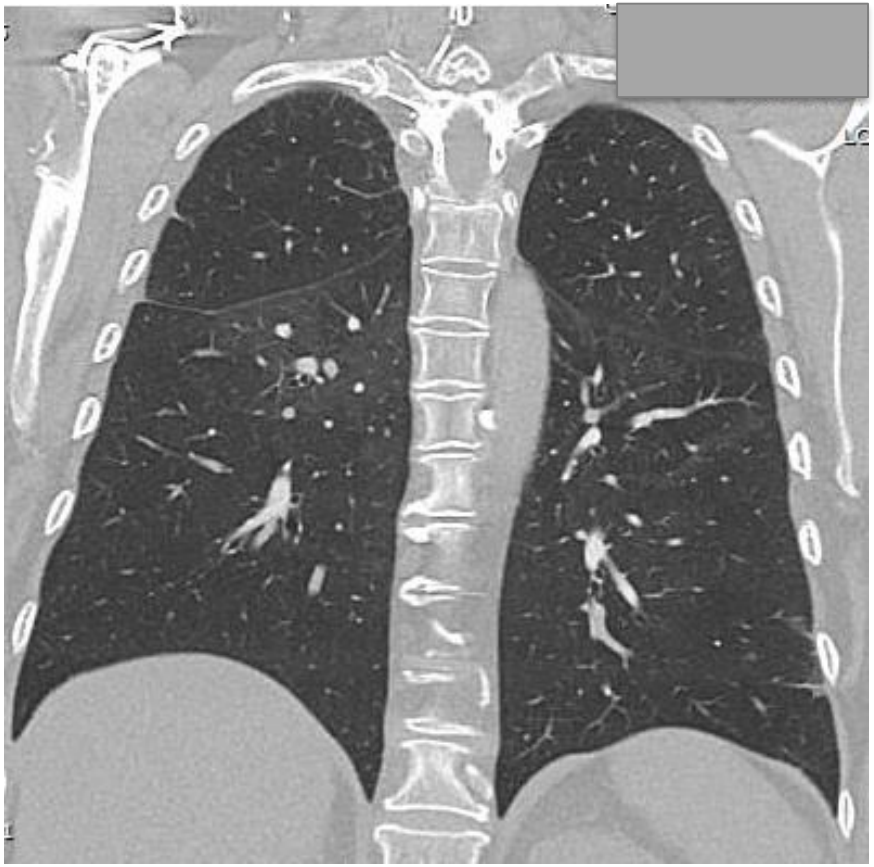
Ventilation and Perfusion



CT Findings



CT Imaging



CT Imaging