Pulmonology Case Presentation

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UNC
SCHOOL OF MEDICINE

Focused patient history and workup

- 72-year-old thin (BMI 22) African American female with lifelong symptoms of cough, sputum production, and intermittent episodes of hemoptysis. Patient was found to have "idiopathic bronchiectasis" and a RUL cavitary lesion(w/ negative TB work up) on admission to outside hospital 7 years ago.
- Patient transferred to UNC with new onset hemoptysis. Sputum cultured a mucoid *Pseudomonas aeruginosa* and *Mycobacterium avium complex.*

Differential Diagnosis for Bronchiectasis

- Idiopathic (commonest in adults)
- Pulmonary Infections ("post infectious")
- Airway Obstruction
- Tracheobronchomalacia and tracheobronchomegaly
- Defective host defenses (Immune deficiency)
- Cystic Fibrosis
- Young Syndrome
- Rheumatic and other systemic disease
- Primary Ciliary Dyskinesia (PCD)
- Alpha-1 Antitrypsin Deficiency
- Allergic bronchopulmonary aspergillosis
- Inflammatory Bowel Disease
- Asthma
- COPD (overlap syndrome)[3,4,5]

Differential Diagnosis for Hemoptysis

- Airway disease (bronchitis, bronchiectasis, neoplasm, foreign bodies, trauma, fistulas, dieulafoy lesion)
- Primary parenchymal disease (infection, rheumatic or immune disorder, genetic connective tissue disorders, coagulopathies, iatrogenic)
- Miscellaneous(cocaine-induced hemorrhage, catamenial hemoptysis, bevacizumab treatment)
- Pulmonary vascular disorders (Pulmonary embolism, pulmonary AV malformation, elevated pulm capillary pressures, Pulmonary artery pseudoaneurysms)
- Cryptogenic[3,4,5]

Imaging Studies

What studies should be ordered?

Imaging Studies

- CXR
- CTA Chest (given initial concern for pulmonary embolism)
- CT Chest w/o contrast

ACR Appropriateness Criteria

Clinical Condition: Hemoptysis

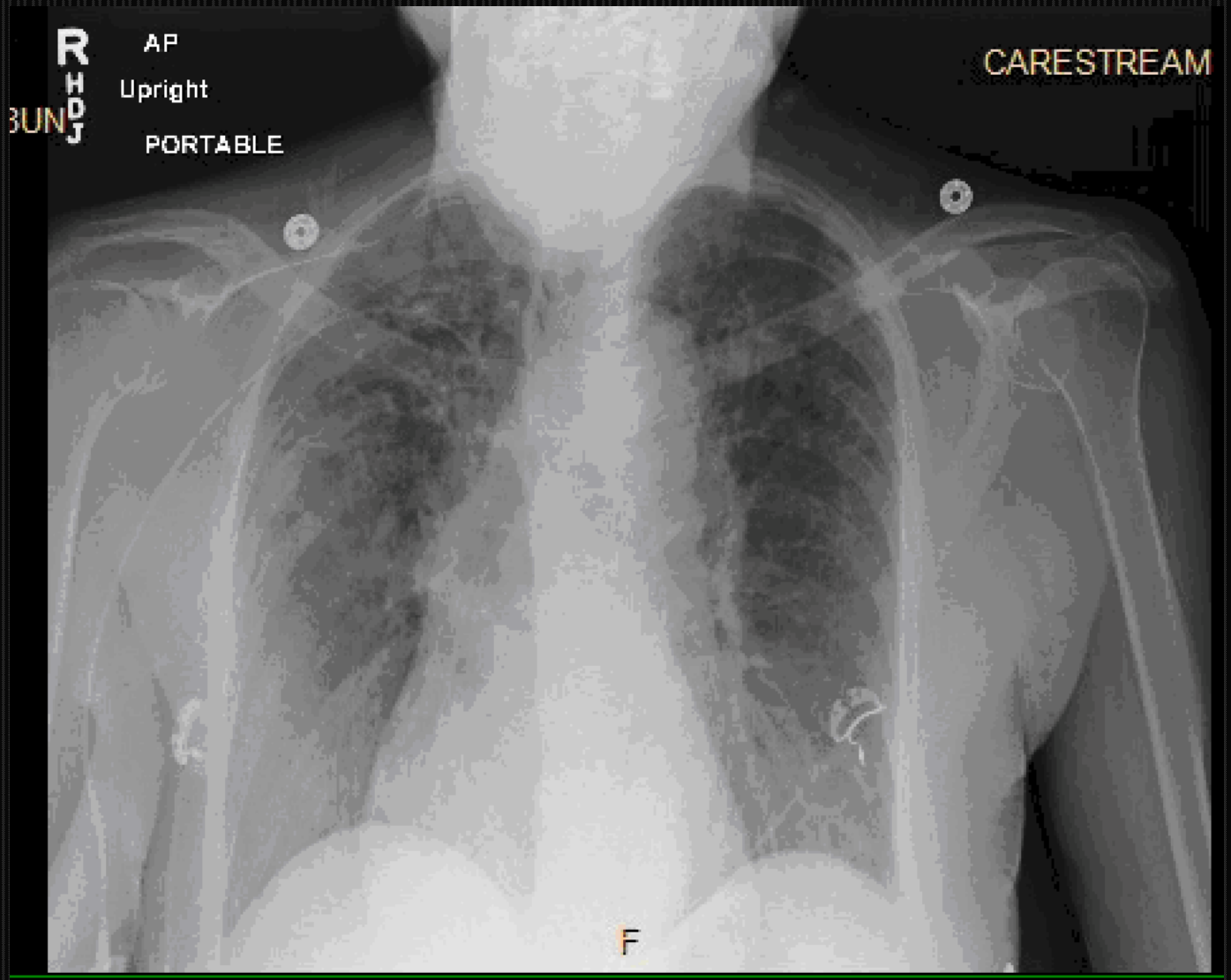
Variant 1: Hemoptysis ≥30 cc OR 2 risk factors (>40 years old and >30 pack-year history).

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		☢
CTA chest with IV contrast	8		☢☢☢
CT chest without IV contrast	6	Consider this procedure if there is a contraindication to iodinated contrast.	☢☢☢
Arteriography bronchial with or without embolization	5	For patients with a preprocedure diagnosis that carries a high risk for recurrent hemorrhage.	Varies
Arteriography pulmonary	2	Consider this procedure for therapy.	☢☢☢☢
Rating Scale: 1,2,3 = Usually not appropriate; 4,5,6 = May be appropriate; 7,8,9 = Usually appropriate			*Relative Radiation Level

Variant 2: Persistent/recurrent hemoptysis (<30 cc) and one risk factor (>40 years old, >30 pack-year history).

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		☢
CTA chest with IV contrast	8		☢☢☢
CT chest without IV contrast	6	Consider this procedure if there is a contraindication to iodinated contrast.	☢☢☢
Arteriography pulmonary	2		☢☢☢☢
Rating Scale: 1,2,3 = Usually not appropriate; 4,5,6 = May be appropriate; 7,8,9 = Usually appropriate			*Relative Radiation Level

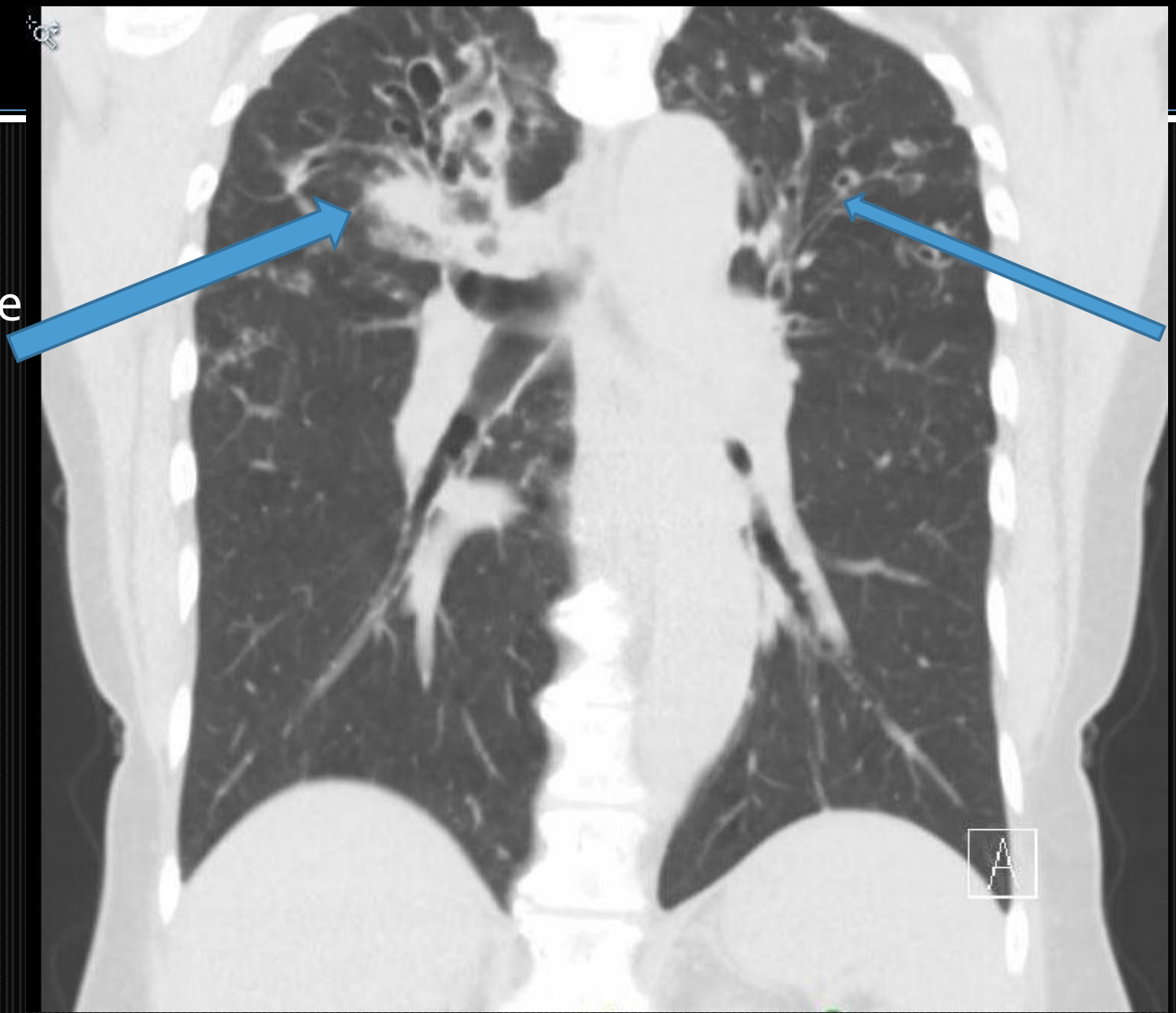
AP Chest XR



A/P CXR: Diffuse heterogeneous airspace disease likely related to patient's diagnosis of bronchiectasis

Chest CT

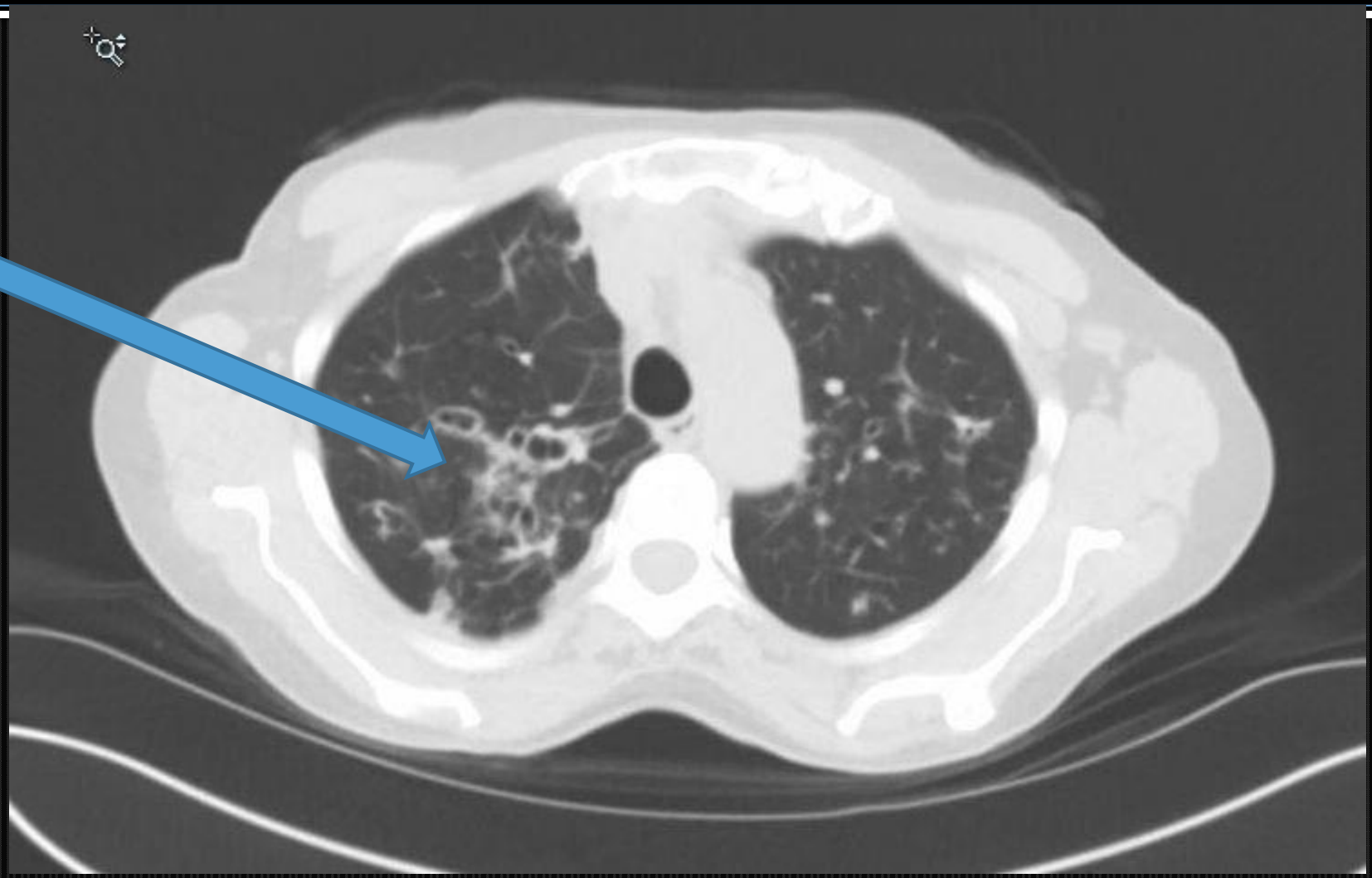
- Upper lobe predominant disease
- RUL consolidation



Airway dilation with bronchial wall thickening

Chest CT

Dilated and thickened airways with peribronchial cysts



Imaging discussion- CT

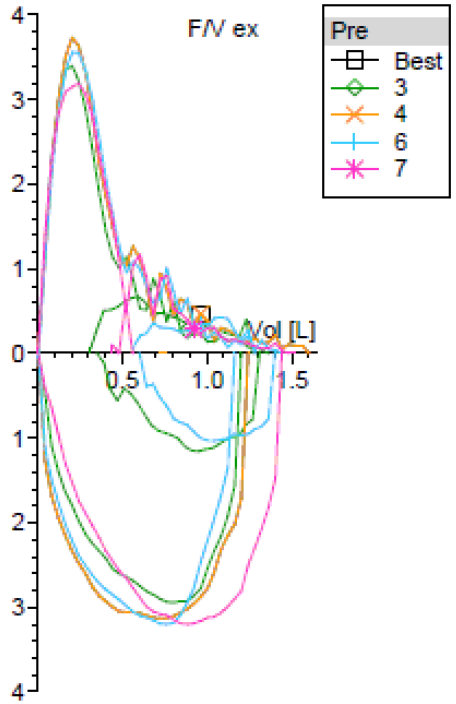
- High resolution CT(HRCT) or multidetector helical CT(MDCT) are the "gold standards" for bronchiectasis
- Features of bronchiectasis on CT:
  - Airway dilation without tapering
  - Bronchial wall thickening
  - Post-obstructive air trapping with "tree-in-bud" pattern
    - Peripheral short linear branching
  - Cysts off bronchial walls, may appear clustered
  - Distribution can help narrow diagnosis (perihilar consistent with ABPA, upper lobe predominant suggests CF, middle and lower lobe consistent with PCD, lower lobe predominant with post-viral disease)[1]

Imaging discussion continued

- CXR can be suggestive but not diagnostic of bronchiectasis
- Findings consistent with bronchiectasis include
  - Linear atelectasis
  - Dilated & thickened airways
  - Irregular peripheral opacities[1,2,4]

Pulmonary Function Tests

- Patient underwent PFTs that revealed severe obstructive impairment with FEV1 43.8%, FVC 57% and FEV1/FVC ratio 77%.



PULMONARY FUNCTION ANALYSIS

		Ref	LLN	Pre	Pre % Ref
FVC	L	2.81	2.01	1.61	57.2
FEV 1	L	2.19	1.50	0.96	43.8
FEV1/FVC	%	77	67	60	77.3
FEV 6	L	2.72	1.92	1.52	56.1
FEV1/FEV6	%	80	70	63	78.6
FEF 25-75%	L/s	1.96	0.40	0.44	22.4
ISOFEF 25-75%	L/s			0.44	
FEF 50%	L/s			0.52	
PEF	L/s	5.45	3.18	3.77	69.1
FET 100%	sec			8.60	
FVC IN	L	2.81	2.01	1.29	45.7
FIF 50%	L/s	2.97	1.54	3.13	105.3
FEF 50 % MIF 50	%			16.80	
PIF	L/s	3.44	3.44	3.13	91.0
V backextrapolation ex	L			0.03	

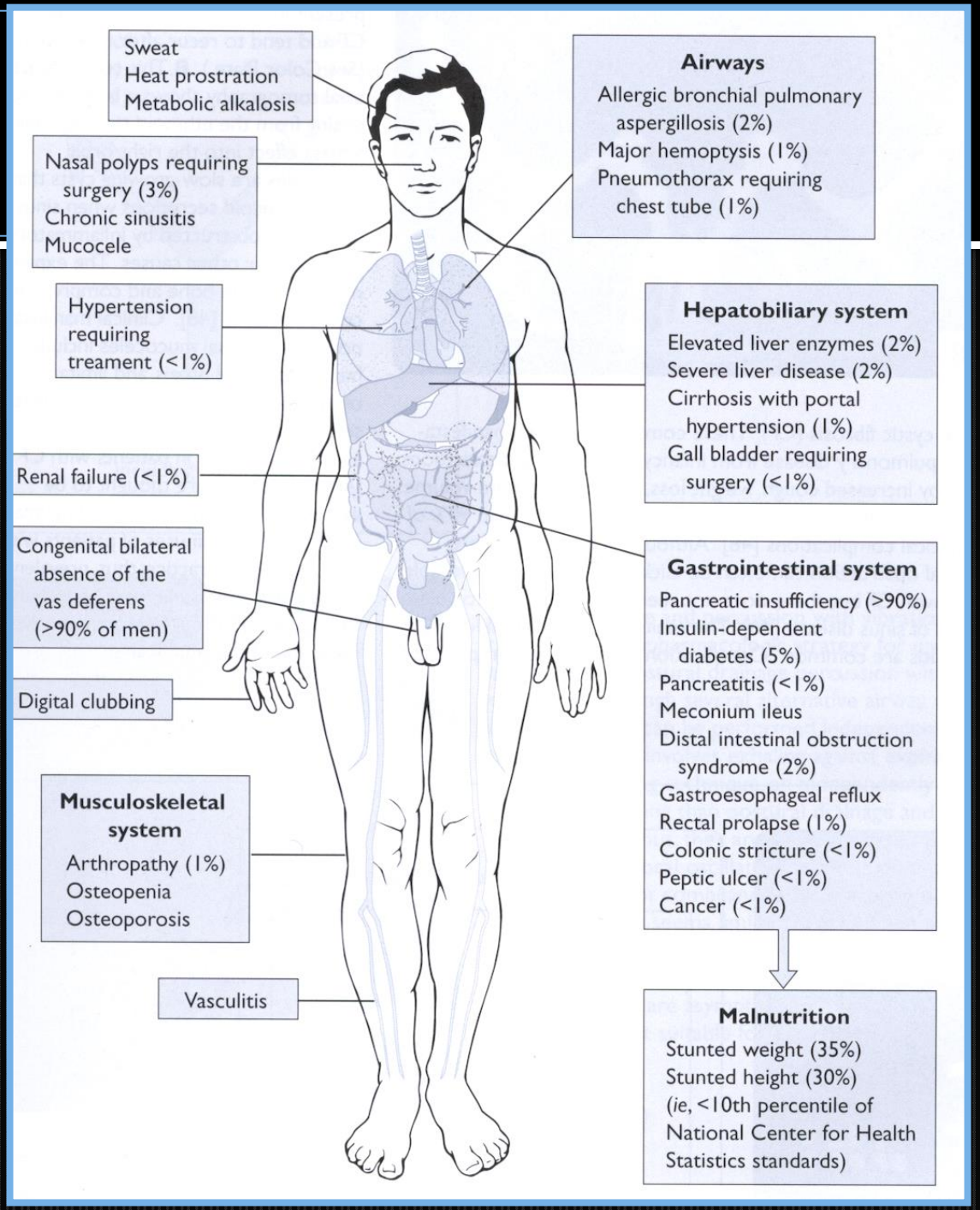
Additional Workup for "Idiopathic Bronchiectasis"

- Given upper-lobe predominance on CT and sputum culture, there was suspicion for **non-classic CF**: Sweat chloride test was positive at 88 mmol/L
- A standard CF genetics panel was negative for CFTR mutation
- Still - the combination of the clinical phenotype , the imaging abnormalities, and the abnormal sweat test support the hypothesis that the patient's lung disease is caused by defective CFTR, likely with some residual function, leading to "non-classic CF" with a late adult diagnosis.

Cystic Fibrosis: Quick Review

Clinical manifestations and complications

(https://www.nejm.org/doi/full/10.1056/NEJMe020070)



Patient treatment

- Hemoptysis felt to be secondary to an acute exacerbation of underlying CF bronchiectasis.
- Patient received the following antibiotic course: vancomycin, meropenem , tobramycin and ceftazidime
- Discharged home on an airway clearance regimen

Acknowledgments

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References

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