Pulmonary ABIM Certification Exam Review Course

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Relative Value?

- Medical Content
  - CV 14%
  - Pulmonary 10%
  - ID 9%; GI 9%

- Cross Content
  - Critical Care 10%
  - Geriatrics 10%
  - Prevention 6%; Women’s Health 6%
Relative Value?

- Pulmonary: ½ is
  - Obstructive disease
  - Occupational and Environmental
  - Restrictive & Interstitial
  - Pulmonary vascular disease

Lecture Outline

- PFTs
- Cough
- Asthma
- Solitary Pulmonary Nodule
- PVD
- ILDs
- TB
- Etc.
A 65 year-old woman with known COPD has spirometry. Which values can be obtained from the following graph?

A. FEV1
B. FEV1/FVC
C. FEF 25%-75%
D. Maximal flow rate at 25% of FVC
E. Maximal flow rate at 50% of FVC
All you need to know is:
FEV1
FEV1/FVC
(FEV1%)
TLC
(and DLCO)
Question 2

Which of the following diseases typically has a normal DLCO?

A. Asthma
B. Emphysema
C. Idiopathic pulmonary arterial hypertension
D. Idiopathic pulmonary fibrosis
E. Pneumocystis jiroveci pneumonia

DLCO: integrity of the alveolar–capillary membrane

Destroy alveoli or capillaries →
Low DLCO
Just narrow airways → Normal DLCO

DLCO: integrity of the alveolar–capillary membrane

**Question 2**

Which of the following diseases typically has a normal DLCO?

A. Asthma
B. Emphysema
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D. Idiopathic pulmonary fibrosis
E. Pneumocystis jiroveci pneumonia

Air “sac” problem

Capillary problem
PFTs

Obstructive Disease: Low FEV1%
- normal DLCO: asthma and chronic bronchitis
- low DLCO: emphysema

Restrictive Disease: Low TLC
- normal or mildly decreased DLCO: obesity
- low DLCO: ILD

Question 3

A 72 year-old man with 5 years of progressive DOE has the following CXR
And has the following PFTs:
FVC 2.4L (52%)
FEV1 1.02L (38%)
FEV1/FVC 41%
TLC 5.0L (77%)
RV 2.6L (120%)
DLCO 12 (48%)

You should order which of the following?

A. Body box plethysmography
B. CT Angiogram
C. HRCT
D. Negative inspiratory pressure
E. Pulmonary exercise testing
Pulmonary Function Tests

- Spirometry
- Flow-volume loop
- Lung volumes
- Diffusing capacity

Diagnose obstruction if FEV1/FVC < 70%
Diagnose restriction if TLC < 80%

Non-specific, but sensitive for alveolar capillary wall integrity
Low in emphysema, ILDs, PVD

TLC measurements in COPD

Can be underestimated by gas dilution technique

Standard
\[ C_1V_1 = C_2(V_1 + V_2) \]

Body box plethysmography estimates entire thoracic cage

Key: CXR was hyperinflated, didn’t make sense that TLC was low.

Large blebs don’t equilibrate
In US, only common disease with RISING mortality


http://www.goldcopd.org
COPD Pathogenesis: Aging + Genes + Noxious Stimuli

In non-smokers, environmental exposure is primary risk factor

Especially in low- and medium-income countries

Indoor smoke from biomass solid fuels → Contribute up to 35% of COPD
A 27 year-old woman has intermittent SOB & wheezing. She has a history of asthma on beta-agonists, high dose ICS, and leukotriene modifiers. 2 prior hospitalizations; 1 requiring intubation for respiratory distress. On exam, she is comfortable but anxious. O2 saturation on room air is 99%. Her lungs have a faint inspiratory wheeze throughout.

You send her for pulmonary function testing with flow volume loop and a chest radiograph. The chest radiograph is normal.
Question 4

The clinical picture most likely represents:

A. Allergic pulmonary aspergillosis
B. Poor adherence to medications
C. Tracheal stenosis
D. Worsening of her underlying asthma
E. Vocal cord dysfunction

Flow-volume Loops

- Normal
- Restriction
- Obstruction
- Severe Obstruction
Flow-volume Loops
Upper Airway Obstruction

- Fixed Large Airway
- Variable Extrathoracic
- Variable Intrathoracic

Variable Extrathoracic Obstruction from Vocal Cord Dysfunction

- Psychogenic
- Most commonly in women, ages 20 - 40
- May present with respiratory distress and dramatic inspiratory stridor
- Loudest noise above throat
- Normal ABGs and A-a gradient
- Resolves when asleep
- Minimal response to aggressive asthma treatment
- Really hard when co-exists with asthma
- Diagnosis by endoscopy
The clinical picture most likely represents:

A. Allergic pulmonary aspergillosis
B. Poor adherence to medications
C. Tracheal stenosis
D. Worsening of her underlying asthma
E. Vocal cord dysfunction

A 38 year-old woman has a nonproductive cough for 6 months. She “clears her throat” frequently. She does not have heartburn or other medical problems including asthma or allergies. She takes no medications. + 5 pack-year history of smoking but quit ten years ago. Her mother had asthma. A chest x-ray is normal.
Question 5
Which of the following would be the most appropriate next step?

A. Obtain radiographs of her sinuses
B. Begin empiric trial of antihistamines
C. Begin empiric trial of inhaled bronchodilators
D. Obtain pulmonary function tests
E. Perform esophageal pH monitoring

Chronic Cough
Non-smoker, not on ACE-i
Most Common Causes

- Post-nasal drip
- Gastroesophageal reflux
- Asthma
- Chronic bronchitis
- Bronchiectasis
- Other

Most cost effective to try empiric Rx, esp. if PND
Evaluation for Chronic Cough that’s not improving

- Post-nasal drip
  - Sinus CT
- Gastroesophageal reflux
  - 24 hour esophageal pH monitoring
- Asthma
  - Spirometry
  - Methacholine challenge
- Other stuff
  - HRCT for occult ILD, bronchiectasis (MAC)

Try to confirm most likely dx, then consider that more than one may be causing, consider others

Question 5

Which of the following would be the most appropriate next step?

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B. Begin empiric trial of antihistamines
C. Begin empiric trial of inhaled bronchodilators
D. Obtain pulmonary function tests
E. Perform esophageal pH monitoring

Second line after best guess with empiric Rx

Good choice if hear wheezing or if exercise induced
A 23 year-old woman is seen for increasingly frequent asthma exacerbations. She has asthma symptoms approximately 3x a week and is awakened at night about 3x a month. The patient is taking a short-acting inhaled beta 2-agonist for symptomatic relief.

On exam, afebrile, BP 120/75, RR 16/min. Lungs: no wheezes (normal).

Her peak flow is 400 liters per minute (her best value is 450 liters per minute).

Which of the following asthma medications would be the most appropriate addition to the treatment regimen at this time?

A. Oral corticosteroids  
B. Oral theophylline  
C. Low-dose inhaled corticosteroid  
D. Long-acting beta 2-agonist  
E. Leukotriene modifier
### Management of Asthma

#### A Step-Wise Approach

<table>
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<tr>
<th>Step</th>
<th>Symptoms</th>
<th>Nocturnal</th>
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<tbody>
<tr>
<td>4, 5, 6</td>
<td>Continual symptoms</td>
<td>Frequent</td>
</tr>
<tr>
<td>Severe persistent</td>
<td>Limited physical activity</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Frequent exacerbations</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Daily symptoms</td>
<td>&gt; one/wk</td>
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<tr>
<td>Moderate persistent</td>
<td>Daily use of inhaler</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Exacerbations affect activity</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Exacerbations ≥ 2 times/wk</td>
<td></td>
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<tr>
<td>2</td>
<td>Symptoms &gt;2 times/wk, &lt;1/day</td>
<td>&gt;2/mo</td>
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<tr>
<td>Mild persistent</td>
<td>Exacerbations may affect activity</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Symptoms ≤ 2 times/wk</td>
<td>≤2/mo</td>
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<tr>
<td>Mild intermittent</td>
<td>Asymptomatic between exacerbations</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Exacerbations brief</td>
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</tbody>
</table>
Management of Asthma
A Step-Wise Approach

Quick-relief

Long-term

STEP 1
Mild Intermittent

Low-dose* inhaled steroids

Low-med dose* inhaled steroids

Med-dose inhaled steroids

High-dose Inhaled steroids

Consider Anti-IgE

STEP 5 & 6

STEP 4
Severe Persist.

STEP 3 Moderate Persistent

STEP 2 Mild Persistent

STEP 1 Mild Intermittent

Patient education & environmental control at each step

* Alternatives: leukotriene modifiers, cromolyn, nedocromil, theophylline

Take home points

- Only step to know for boards is from intermittent → mild persistent; if patient is using b-agonist > few times per week for rescue, add controller medication (best for most = an inhaled corticosteroid)
- If poor control on ICS: increase ICS or add long acting b-agonist (deals with concerns about safety of LABA)
- Long acting b-agonist without a controller medication is always the wrong answer
- For emergency rescue, b-agonist always the right answer
LABA

**Pharmacokinetics**
- Salmeterol & Formoterol
  - Effect lasts 12 hours
  - *Formoterol* – quick onset, so some have used as quick relief medication
  - Combo of ICS + Formoterol used for exacerbations (plus action plan!)

**Concerns @ safety of LABAs?**
- Genetic polymorphisms of beta-receptor

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**LABA**

- Beta-receptor → Substitution of glycine for arginine
- Homozygous for Arg/Arg → reduced response to b-agonists
  - Usual care +/- Salmeterol
  - Asthma related deaths 4.4 x more likely in salmeterol group → Black Box warning by FDA
  - Related to LABA, subset with Arg/Arg, or non-compliance with ICS?
Not all patients with mild persistent asthma use their ICS everyday!

Boushey H et al. Daily versus As-Needed Corticosteroids for Mild Persistent Asthma NEJM 2005; 352:1519

- Some “step–down”
- Okay IF patients have symptom based action plan, using ICS for mini-exacerbations

Anti-IgE or Omalizumab

- Consider for Steps 5 & 6
- Binds IgE → complex cleared
- Rx: fewer exacerbations & less steroid needed; no change in baseline FEV1
Anti-IgE or Omalizumab

- Need to get IgE to extremely low levels for it to work (very low levels trigger mast cell degranulation)
- Baseline serum IgE should be between 30 and 700 IU/mL
- + Skin test or RAST to a perennial aeroallergen (e.g., dust mite, animal danders, cockroach, molds)
- Sq each 2-4 weeks
- Anaphylaxis 1:1,000
- Minimum dose $12,000/year

Question 6

Which of the following asthma medications would be the most appropriate addition to regimen at this time (only on short acting b-agonists)?

A. Oral corticosteroids
B. Oral theophylline
C. Low-dose inhaled corticosteroid
D. Long-acting beta 2-agonist
E. Leukotriene modifier

Works, but too big a gun

All acceptable by guidelines, in practice like ICS; Caveat: some young people w/ exercise induced asthma do well on LT agents. Smokers have blunted response to ICS.
30 year-old woman was exposed to chlorine gas 2 months ago at work & now has a persistent cough & mild SOB. At exposure, she noted irritation of her eyes and mucus membranes. Immediately after the exposure, she developed a cough. A chest x-ray was normal. No treatment was given. The patient has no history of asthma, but since this, has been wheezing at night.

Exam is unremarkable with clear lungs.
Spirometry:  FVC of 89% of predicted  
            FEV₁ of 84% of predicted  
            FEV₁/FVC 73%
Methacholine challenge + for bronchial hyperresponsiveness

Which of the following is the most likely diagnosis?

A. Bronchiolitis obliterans  
B. Hypersensitivity pneumonitis  
C. Post nasal drip  
D. Reactive airways dysfunction syndrome
Occupational Asthma

Occupational asthma

Irritant-induced asthma

Reactive airways dysfunction

Acquired sensitization in the workplace

Multiple exposures to irritant

Single big exposure to irritant

Non-immunologic

Reactive Airways Dysfunction

Diagnostic Criteria

- Exposure to irritant in high concentration
- Symptoms of asthma
- Onset of symptoms after single exposure within 24 hrs; persist for at least 3 months
- PFTs +/- airflow obstruction, but Methacholine test positive

Brooks SM et al. Chest 1985;88:376
RADS

Take home points
- “Big Bang” → big exposure, symptoms right away
- Can last for YEARS!!!
- Rx like asthma, though typically harder to control
- CHLORINE! (Including mixing household cleaners)
- Gulf War → sulfur mustard gas

Question 7
Typically slower onset; HRCT scan +
Which of the following is the most likely diagnosis?
- A. Bronchiolitis obliterans
- B. Hypersensitivity pneumonitis
- C. Post-nasal drip
- D. Reactive airways dysfunction syndrome

Can exacerbate asthma, but doesn’t CAUSE airway hypereactivity
An asymptomatic 62 year old smoker has a pre-op film for unrelated problem which reveals a 1.8 cm nodule:

CT scan: no evidence of calcification or metastases
Spirometry: normal
PPD: negative

In retrospect, it was present on a CXR 1 year ago and was .8 cm in diameter.
**Question 8**

You recommend:

A. Repeat chest x-ray in one year
B. Bronchoscopy with transbronchial biopsy
C. Mediastinoscopy
D. Sputum cytology x 3
E. Surgical resection

---

**Solitary Pulmonary Nodule**

- **Is it cancer?** Then, is it resectable…?  

If no prior chest x-ray, calculate risk based on size, age, amount of smoking (can also use upper lobe (bad), prior cancer (bad), non-smooth edge (bad)

This patient = @59%

Compare to 35 y.o. non-smoker with a .8 cm nodule < 1%

- If probability of cancer very **high** (70 %+), immediate **surgery**: slightly longer average life expectancy.
- If probability of cancer **intermediate** (3-69%), **biopsy** strategies: narrow advantage.
- If probability of cancer **very low** (< 3%), **observation**: slightly longer average life expectancies.
**Question 8**

A. Repeat chest x-ray in one year

B. Bronchoscopy with transbronchial biopsy

C. Mediastinoscopy

D. Sputum cytology x 3

E. Surgical resection

Not if CT scan neg for LNs

Won’t change need for resection and $

Unless central mass – yield of bronch low.

Yield of FNA high (80% but with 20% risk of PTX)

This lesion is growing at a rate suggestive of a cancer! Take out!

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**SPN – Is it resectable?**

60 yo woman smoker with COPD, SPN 2 cm in LLL, not seen on film 3 years ago. CT no evidence of mets. PFTs: FVC 2.2 L (79%) FEV1 1.2 L (58%)

Because of surgical risk higher for resection, FNA performed and showed adenocarcinoma.

Spilt perfusion scan:

<table>
<thead>
<tr>
<th></th>
<th>Right lung</th>
<th>Left lung</th>
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<tbody>
<tr>
<td>Upper</td>
<td>20%</td>
<td>16%</td>
</tr>
<tr>
<td>Lower</td>
<td>36%</td>
<td>28%</td>
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Question 9

60 yo with FEV1 1.2 L (58%); Split perfusion to LLL = 36%. Which of the following is appropriate?

A. Chemotherapy first to shrink tumor then resect
B. Left lower lobectomy
C. Observation
D. XRT at high dose to cure
E. Wedge resection

Lung Cancer

Take home points
- XRT ½ cure rate of surgical resection
- Wedge resection is inferior for cure vs. full resection except if > 75
- Pre-op chemo now only indicated for Stage IIIa cancers
- If she loses her LLL, her post-op predicted FEV1 = 1.2L x 72% = .86L .... or 56% of predicted.
- Want post op FEV1 to be > 40%
Question 9

60 yo with FEV1 1.2 L (58%); Split perfusion to LLL = 36%. Which of the following is appropriate?

A. Chemotherapy first to shrink tumor then resect
B. Left lower lobectomy
C. Observation
D. XRT at high dose to cure
E. Wedge resection

IIIa only
Use % predicted for risk of surgery
Decreased cure rates

Question 10

Which of the following are used in the routine treatment of patients with primary pulmonary hypertension?

A. Calcium channel blocker
B. Digoxin
C. Epoprostenol
D. Nitric oxide
E. Warfarin
Pulmonary Hypertension

Endothelin → NO → Prostacyclin

Endothelial Cell

Smooth muscle contraction
Endothelin is also smooth muscle mitogen

Smooth muscle relaxation

ET-R

Pulmonary Hypertension - RX

Endothelin receptor-blockers
- Bosentan (Oral)
  - Hepatotoxicity
  - (warfarin trickier)

Prostanoids:
- Prostacyclin = Epoprostenol
- (Flolan) continuous IV
- Iloprost (Inhaled)
- Treprostinil (IV or SQ)

Calcium Channel Blockers
- Only 5-10% respond

Phosphodiesterase inhibitors:
- Prolong NO action:
- Sildenafil & Vardenafil
**Pulmonary Hypertension- RX**

<table>
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<tr>
<th>Drug</th>
<th>Notes</th>
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<tbody>
<tr>
<td>Calcium Channel Blockers</td>
<td>Only 5-10% respond</td>
</tr>
<tr>
<td>Epoprostenol</td>
<td>Cheap</td>
</tr>
<tr>
<td>Iloprost (Inhaled)</td>
<td>If severe, most start here</td>
</tr>
<tr>
<td>Treprostinil (IV or sq)</td>
<td>Combos with other drug classes work</td>
</tr>
<tr>
<td>Bosentan</td>
<td>NOT if hepatopulmonary HTN</td>
</tr>
<tr>
<td>Sildenafil</td>
<td>Oral, well tolerated</td>
</tr>
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</table>

**Question 10**

Which of the following are used in the routine treatment of patients with primary pulmonary hypertension?

A. Calcium channel blocker  
B. Digoxin                    
C. Epoprostenol              
D. Nitric oxide              
E. Warfarin                  

- Only 5% with sustained benefit, never do without PAC to prove efficacy
- OK if LV failure
- If severe
- No portable system yet
- Endothelial disruption: in-situ clotting, even small clots can tip a patient over
Don’t forget to correct Hypoxemia!

pH 7.38/ pCO2 44/ pO2 58/ sat’n 89%

Prescribe oxygen!

What about Pulmonary HTN related to COPD?

On going studies in COPD patients with pulmonary HTN. HUGE market. Stay-tuned but pre-lim: Hypoxemia worsens from more V/Q mismatch

Question 11

65 year old man with a history of TB has intermittent hemoptysis without fevers/chills/weight loss.

Recent spirometry: FEV1 1.0L (40%), FVC 1.5L. He now expectorates 200 mL of bright red blood.

Exam: afebrile, BP 145/82, pulse 104, RR 18, SaO₂ 93% on air.

Bronchoscopy: blood coming from the left upper lobe bronchus.
Chest X-ray

Chest CT Scan
Question 11

What is the best management option for this patient at the present time?
A. Bronchial arteriography with embolization
B. Four first-line drugs for tuberculosis
C. Intravenous Amphotericin B
D. Left upper lobe resection

Causes and Management of Massive Hemoptysis

Massive usually means ≥ 200 mL in 24hrs
Most common causes include:
1) TB (active or inactive disease)
2) Bronchiectasis
3) Lung cancer
4) Mycetoma
5) Immunologic diseases (ANCA-associated vasculitis, Goodpasture’s, SLE)
Management of Massive Hemoptysis

- First, protect the airway
- Bronchoscopy can localize; make some diagnoses
- Majority of massive bleeds have bronchial circulation → Bronchial arteriography with embolization next step. 85% successful.
- Surgery is definitive, but high M&M if done urgently

Our patient has a mycetoma, and actively bleeding → arteriography and embolization successful

Question 11

What is the best management option for this patient at the present time?

A. Bronchial arteriography with embolization
B. Four first-line drugs for tuberculosis
C. Intravenous Amphotericin B
D. Left upper lobe resection

Old cavity; no clear evidence of active disease

Doesn't penetrate fungus ball well; Itraconazole may
Question 12

52 year old man with alcoholic cirrhosis with prior variceal bleeding has progressive dyspnea on exertion for 3 months. Denies chest pain, fever, or sputum production. Has gained 5 pounds over the past month.

Meds: propranolol.

Chest X-ray
Lateral Decubitus

Right side

Question 12

What is the optimal management in this case?
A. Large volume thoracentesis
B. Chest tube insertion
C. Pleurodesis
D. Medical management with diuretics
Pleural Effusions in Patients with Liver Disease

- Hepatohydrothorax: Effusions usually when ascitic fluid is present, but not always
- Fluid passes from peritoneum to pleural space via diaphragmatic pores & possibly lymphatic channels. Negative pleural pressure draws fluid up.
- Fluid is transudative with very protein
- Typically free-flowing

Management: decrease ascites formation
- Low salt diet
- Diuretics
- TIPS if refractory
**Question 12**

What is the optimal management in this case?

A. Large volume thoracentesis
B. Chest tube insertion
C. Pleurodesis
D. Medical management with diuretics

**Just keeps draining; reserve large volume thoracentesis for acute dyspnea relief**

**Question 13**

33 year-old woman presents with intermittent fever, night sweats, migratory joint pains, and red, painful nodules on her shins.

Chest x-ray: bilateral hilar adenopathy without infiltrates or effusions.

Bronchoscopy with transbronchial biopsy: non-caseating granulomas. Stains and cultures for fungi and mycobacteria were negative.
Question 13

Which best describes the status of her lung disease in 2 years?
A. Progression to advanced obstructive lung disease
B. Progression to advanced interstitial lung disease with fibrosis
C. Progression to pulmonary hypertension
D. Normal lung function
Overview of Sarcoidosis

- Multisystem granulomatous disorder of unknown etiology characterized by non-caseating granulomas in involved organs
- Incidence varies geographically and is much more common in African-Americans (lifetime risk of 2.4%)
- Usually presents ages 10 - 40, half detected by CXR without symptoms
- Any organ can be involved, lungs most frequent (90%)

Sarcoidosis-Staging

- Stage I  Bilateral hilar adenopathy
- Stage II  Above + interstitial infiltrates (upper>lower lung zones)
- Stage III  Interstitial disease with shrinking hilar nodes
- Stage IV  Advanced fibrosis

- Extra-pulmonary disease-skin (E. nodosum, lupus pernio), eyes, liver, lymph nodes most frequent
Sarcoidosis: Diagnosis and Treatment

- Key to diagnosis remains suspicion, exclusion of infection, and histologic evidence of granulomas
- Usual indications for treatment are: worsening pulmonary symptoms, lung function, progressive radiographic changes
- Therapy is not indicated in
  - Asymptomatic stage I disease patients
  - Asymptomatic patients with stage II and mildly abnormal lung function
    - Follow first for 3-6 months and document impairment of lung function

Question 13

Which best describes the status of her lung disease in the following 2 years?

- She had Lofgren's syndrome: “Acute” sarcoid with abrupt onset with erythema nodosum, hilar adenopathy, migratory polyarthritis, and fever seen primarily in women.
  - Strongly associated with HLA-DQB1*0201
  - Good prognosis and spontaneous remission
Question 14

34 year old man has exertional dyspnea of 6 months and cough productive of yellow sputum. No fever, chills, or hemoptysis. No risk factors for HIV. Physical exam is normal. Sputum smears are negative for AFB. He has had a pet pigeon for the past 2 years.

Pulmonary function tests:
FEV1/FVC 83% predicted  ABG 7.49/30/60
TLC 68% predicted  DLCO 50% predicted

Chest X-ray
Question 14
Which of the following is the most likely diagnosis?
A. Idiopathic pulmonary fibrosis
B. Lymphangioleiomyomatosis
C. Sarcoidosis
D. Hypersensitivity pneumonitis

Interstitial Lung Diseases
- Characterized by restriction on PFTs with low diffusion capacity and desaturation with exercise (or if bad → hypoxemia at rest)
- High resolution Chest CT scan is almost always the right answer to “what to do next” if hasn’t been ordered
  • Some ILDs have classic findings
  • Shows where to biopsy
Interstital Lung Diseases

- May ask WHETHER to biopsy (yes though many patients with IPF are old with lots of comorbidities)
- NOT likely to ask if patient should have biopsy by bronch (sarcoidosis) or surgical lung biopsy (most others to get enough tissue to diagnose)

ILD: HP

Hypersensitivity pneumonitis: ground glass opacities, centrilobular nodules, and air trapping on expiratory views
Hypersensitivity Pneumonitis

Chronic granulomatous inflammation after repeated inhalation of environmental antigens

- Can present as acute, subacute or chronic syndromes
- No single test is diagnostic
- Suspect when there is a
  - History of recurrent pneumonias
  - Symptoms develop after moving to a new job or home or birds or water damage/visible mold in work/home
  - Improvement in symptoms when away from work/home

Air-trapping?

Inspiration  Expiration

If small airways are inflamed, air can’t exit well with exhalation. On CT scan, involved lung areas remain black Splotchy pattern suggestive of small airway inflammation.
HRCT What to order?
Get inspiratory and expiratory views (small airway disease)
Plus prone & supine images. Can open up atelectasis that can be confused with an ILD

ILDs: Classic HRCT Findings
Sarcoid: nodular thickening of bronchovascular bundles (lumpy-bumpy), centrilobular nodules, and adenopathy.
**ILD: Sarcoid**

Disease @ bronchovascular bundles

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**Question 14**

34 man with a bird.
Which of the following is the most likely diagnosis?

A. Idiopathic pulmonary fibrosis
B. Lymphangioleiomyomatosis
C. Sarcoidosis
**D. Hypersensitivity pneumonitis**

Lack of systemic symptoms, typical HRCT, bird exposure favor HP
Distinction important – need to remove antigen (bird)!
LAM

Classic LAM story: 35 year old woman with dyspnea and PTX or chylothorax

Question 15

46 year old woman has 4-weeks of fever, night sweats, cough, and 10-pound weight loss. She also has arthralgias, epistaxis, nasal congestion. 2 weeks of clarithromycin did not relieve her symptoms. Now has hemoptysis. Exam: 99.7, RR 24/min, crackles right chest, 1+ edema
WBC 6800/mm³ Hgb 10.3
Platelets 568,000/mm³
Creatinine 1.3 mg/dL
Urinalysis: 2+ protein, 0 WBCs, rare RBC casts
Question 15

Which of the following is the best diagnostic step?

A. Serum angiotensin-converting enzyme
B. Measure rheumatoid factor
C. Antineutrophil cytoplasmic antibodies
D. Culture of bronchoalveolar lavage fluid
E. Percutaneous needle biopsy of the lung
Pulmonary-Renal Syndromes

Systemic vasculitis
- Wegener’s granulomatosis
- Microscopic polyangiitis
- Pauci-immune GN
- Churg-Strauss (allergic angiitis and granulomatosis)
- Goodpastures syndrome
- Systemic lupus erythematosus
- Henoch-Schonlein purpura

Infection
- Post-streptococcal glomerulonephritis, endocarditis

ANCA?
- C-ANCA 80%
- P-ANCA 10%
- P-ANCA 70%
- Most P-ANCA
- ½ ANCA
- P-ANCA 10-40%
- Some +

Approach to Pulmonary-Renal Syndromes

- Serologic tests: ORDER
  - Anti-GBM Abs, anti-neutrophil cytoplasmic Abs (ANCA), ANA if SLE suspected
  - ANCAs are positive in 90% of those with generalized Wegener’s (PR3-ANCA or “C-ANCA”)

- Tissue should be obtained to provide evidence of vasculitis
  - Skin (easy), Kidney, or lung (surgical biopsy)
  - If Anti-GBM possible, kidney better to bx than lung
Question 15

Which of the following is the best diagnostic step?
A. Serum angiotensin-converting enzyme
B. Measure rheumatoid factor
C. Antineutrophil cytoplasmic antibodies
D. Culture of bronchoalveolar lavage fluid
E. Percutaneous needle biopsy of the lung

Reasonable, but fungi, TB can not explain GN
Not enough tissue to see vessel

Not pulmrenal syndromes

Question 16

72 year old man with progressive dyspnea for 2 years. No sputum, hemoptysis, weight loss, or sweats. He previously smoked 1 ppd for 25 years, quit 15 years ago. PMH: HTN and peptic ulcer disease.

Meds: omeprazole and lisinopril

Exam: afebrile, RR 16, SaO2 92% RA
   Crackles bilaterally at bases
   + Clubbing

Labs: normal CBC, Chem-20,
ABG: 7.42/28/58
Chest X-ray

HRCT

Honeycombing
HRCT

Traction bronchiectasis

Question 16

Which of the following is the most likely diagnosis?

A. Bronchiolitis obliterans organizing pneumonia
B. Chronic aspiration pneumonia
C. Sarcoidosis
D. Idiopathic pulmonary fibrosis
**Diffuse Parenchymal Lung Diseases**

- Other “Systemic” Disorders
- Connective Tissue Diseases
- Idiopathic Interstitial Pneumonias
- Occupational and Environmental Exposures
- Drug-Induced Lung Injury

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**Interstitial Lung Disease: Clues from HRCT scans**

- **Bronchiolitis obliterans organizing pneumonia** (BOOP or COP) has patchy consolidation, often subpleural
- **Chronic aspiration dependent areas**
- **Sarcoidosis** has bronchovascular predilection, “beading” or “string of pearls” on fissures, LNs
- **Idiopathic pulmonary fibrosis** shows basilar and subpleural linear opacities with bronchiectasis and honeycombing
**Reality? Dozens of ILDs!**

**Only 3 patterns for ABIM**
Honeycombing, especially subpleural in bases + traction (non-purulent) bronchiectasis = IPF

Bronchovascular thickening, “string of pearls” on fissures, adenopathy = sarcoid

AND….

**Everything Else!**

HRCT: + disease, but not diagnostic of what it is

**Biopsy!**
Question 17

30 year old man has increasing dyspnea with exercise and chronic daily productive cough since adolescence. He also reports frequent bronchial and sinus infections, treated with multiple courses of antibiotics. Twice he was admitted for pneumonia. He has a 20-pack year history of smoking. No other medical problems or prior surgeries. He works in an office.

Exam: SaO2 86%, diffuse crackles, and digital clubbing.

Chest X-ray
Which of the following should be ordered to establish the most likely diagnosis?

A. Serum IgG and IgE for Aspergillus
B. Serum IgA and IgG levels
C. Sweat chloride measurement
D. Nasal mucosal biopsy
E. Serum $\alpha_1$-antitrypsin level
**Bronchiectasis: Causes**

- Bronchopulmonary infections
  - Bacterial, fungal, mycobacterial
- Bronchial obstruction
  - Foreign-body, tumors, lymph nodes
- Immunodeficiency states
  - IgA, IgG deficiency
- Hereditary abnormalities
  - Cystic fibrosis, ciliary dyskinesia, α-1 antitrypsin deficiency
- Miscellaneous: Rheumatoid, Sjogren’s

**Bronchiectasis**

<table>
<thead>
<tr>
<th>Blood</th>
<th>Imaging</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>CBC</td>
<td>HRCT</td>
<td>Spirometry</td>
</tr>
<tr>
<td>IgA, IgE, RF</td>
<td>Sinus CT</td>
<td>Sputum c/Aspergillus</td>
</tr>
<tr>
<td>IgG subclasses</td>
<td></td>
<td>Sweat chloride</td>
</tr>
</tbody>
</table>
<pre><code>                   |               | Nasal mucosal bx           |
                   |               | Bronchoscopy               |
</code></pre>
Primary Ciliary Dyskinesia

- Chronic cough, rhinitis, and sinusitis
- Cilia do not beat normally
- Triad of situs inversus, chronic sinusitis, and bronchiectasis = Kartagener’s syndrome
- Situs inversus is present in 50% of patients with primary ciliary dyskinesia

Question 17

Which of the following should be ordered to establish the most likely diagnosis?

A. Serum IgG and IgE for Aspergillus (ABPA)
B. Serum IgA and IgG levels
C. Sweat chloride measurement
D. Nasal mucosal biopsy
E. Serum $\alpha_1$-antitrypsin level

All can cause bronchiectasis with purulent sputum and all part of a work-up, but with situs inversus, start with evaluation of cilia
32 year old woman from China with a known positive PPD has a chronic cough and night sweats for 2 months. Chest radiograph shows a right upper lobe cavity. Two of three smears are positive for acid-fast organisms. She is currently 32 weeks pregnant.

What is the most appropriate next step?
A. Await final sputum culture results
B. Begin treatment with isoniazid, rifampin, ethambutol, and pyrazinamide
C. Begin treatment with isoniazid, rifampin, and ethambutol
D. Begin treatment with isoniazid, rifampin, and pyrazinamide
E. Call the CDC and transfer her to National Jewish Hospital
**TB and Pregnancy**

- Pregnancy per se does not increase the risk of developing active TB
- Remember that a positive AFB smear = empiric treatment for active TB
- This is different than simply a positive tuberculin test during pregnancy, when treatment of latent TB infection can usually be deferred until after delivery

**TB and Pregnancy**

- Standard initial TB treatment is 4 drugs (isoniazid/rifampin/ethambutol/pyrazinamide)
- During pregnancy, it is recommended that pyrazinamide be avoided although teratogenicity has not been proven
- Mnemonic: P = No PZA in pregnancy!
- This means that treatment duration will be prolonged to 9 months!
52 year old woman non-smoker without significant past medical history has productive cough for 1 year. Sputum from 9 months ago grew *Mycobacterium avium*. She took 2 courses of antibiotics without improvement. Sputum from 2 months ago is also positive for *M. avium*. CT scan shows nodules in the lingula with mild bronchiectasis, and no cavities.

Which of the following is the next most appropriate step?

A. Clarithromycin daily
B. Clarithromycin, ciprofloxacin, and isoniazid daily
C. Rifampin, azithromycin, and ethambutol three times weekly
D. Video-assisted thoracoscopic lung biopsy
E. Prednisone daily
**Non-tuberculosis Mycobacterial Infection**

- Most common isolate in the U.S. is *Mycobacterium avium*
- Not contagious, isolation not required
- 2 classic presentations:
  - 1) male smokers with cough/upper lobe cavities
  - 2) middle-aged women with cough and lingular or right middle lobe bronchiectasis/nodules

**Mycobacterium avium Lung Disease**

**Diagnostic criteria:**

Symptoms + nodules, cavities, or bronchiectasis with:

- Positive cultures from at least 2 separate sputum samples
  OR
- Positive culture from at least 1 bronchial wash
  OR
- Transbronchial or other lung biopsy with characteristic histology and positive culture on either biopsy or sputum

*Am J Resp Crit Care Med 2007;175:367*
Treatment of *Mycobacterium avium* Lung Disease

- For nodular/bronchiectasis, therapy consists of a macrolide (clari or azithro), ethambutol, and rifamycin (rifampin or rifabutin) 3 times weekly
- For cavitary disease, therapy consists of the same drugs given daily +/- streptomycin or amikacin
- The goal of therapy is 12 months of negative sputum cultures while on therapy--total duration is often 14-18 months

*Am J Resp Crit Care Med 2007;175:367*

**Question 20**

65-year-old man who has recently moved to the United States from Mexico has a tuberculin skin test placed. He denies previous exposure to tuberculosis. He has a history of DVTs and takes coumadin. Four days later he returns to the clinic and his skin test is read as being 16 mm in induration. A chest radiograph shows apical pleural thickening but no evidence of parenchymal lung disease.
Question 20

What would be the most appropriate intervention?
A. Repeat the tuberculin skin test
B. Give 6 months of rifampin for treatment of latent tuberculosis infection
C. Give 9 months of isoniazid for treatment of latent tuberculosis infection
D. Collect three sputum specimens and start 4-drug antituberculosis therapy
E. Inform the older gentleman that he has latent tuberculosis infection and to return if he develops symptoms

Factors Associated with Increased Risk of Progression to TB

- **High Risk Conditions**
  - HIV infection
  - Transplant or other immunosuppression
  - Medical conditions
  - Injection drug users
  - Recent arrivals from endemic countries

- **Risk for New Infection**
  - Contacts to infectious cases
  - Health care workers
  - Other workers exposed to TB cases

ATS/CDC AJRCCM 2000;161:S221
### Medical Conditions that Increase the Risk of TB

- HIV infection
- Silicosis
- Diabetes mellitus
- Corticosteroids
- Immunosuppressives
- Hematologic/reticuloendothelial disorders
- ESRD
- Intestinal bypass
- Post-gastrectomy
- Malabsorption
- Carcinomas of head and neck
- < 10% below ideal body weight

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### Treatment of LTBI

#### Drug Regimens

<table>
<thead>
<tr>
<th>Regimen</th>
<th>Duration (months)</th>
<th>Interval</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isoniazid</td>
<td>9</td>
<td>Daily</td>
<td>Preferred regimen</td>
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<tr>
<td></td>
<td></td>
<td>Twice-wkly</td>
<td>DOT necessary</td>
</tr>
<tr>
<td>Isoniazid</td>
<td>6</td>
<td>Daily</td>
<td>Not for HIV+</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Twice-wkly</td>
<td>DOT necessary</td>
</tr>
<tr>
<td>Rifampin-PZA</td>
<td>2</td>
<td>Daily</td>
<td>Drug interactions</td>
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<tr>
<td></td>
<td>2-3</td>
<td>Twice-wkly</td>
<td>DOT necessary</td>
</tr>
<tr>
<td>Rifampin</td>
<td>4</td>
<td>Daily</td>
<td>For INH-Resistant</td>
</tr>
</tbody>
</table>

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ATS/CDC AJRCCM 2000;161:S221
Question 21

50 year old man presents for evaluation of a nonproductive cough and chest pain increasing for the past 3 months. He denies weight loss but notes weakness. Exam reveals a mild ptosis and is otherwise normal. Labs: mild normocytic anemia.

Chest X-ray
Question 21

What is the most likely diagnosis?
A. Bronchogenic cyst
B. Intrathoracic thyroid
C. Lymphoma
D. Teratoma
E. Thymoma

Mediastinal Masses
Differential Diagnosis

First, localize to anterior, middle, or posterior mediastinum

<table>
<thead>
<tr>
<th>Anterior Mediastinum</th>
<th>Middle</th>
<th>Posterior</th>
</tr>
</thead>
<tbody>
<tr>
<td>“The 4 Ts”</td>
<td>Bronchogenic cyst</td>
<td>Neurogenic cyst</td>
</tr>
<tr>
<td>Thymoma</td>
<td>Pericardial cyst</td>
<td>Esophagus</td>
</tr>
<tr>
<td>Thyroid</td>
<td>Lymph nodes</td>
<td></td>
</tr>
<tr>
<td>Teratoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>“Terrible” lymphoma</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Thymomas

Pearl: Disease of “35s”

- #1 anterior mediastinal mass in those > 35
- 35% are malignant
- 35% are associated with myasthenia
- 35% have a paraneoplastic syndrome
  - Pure red blood cell aplasia
  - Hypogammaglobulinemia
  - Cushing’s syndrome

Question 21

What is the most likely diagnosis?
A. Bronchogenic cyst
B. Intrathoracic thyroid
C. Lymphoma
D. Teratoma
E. Thymoma

All anterior mediastinal, but most have CT scan characteristics
The End!