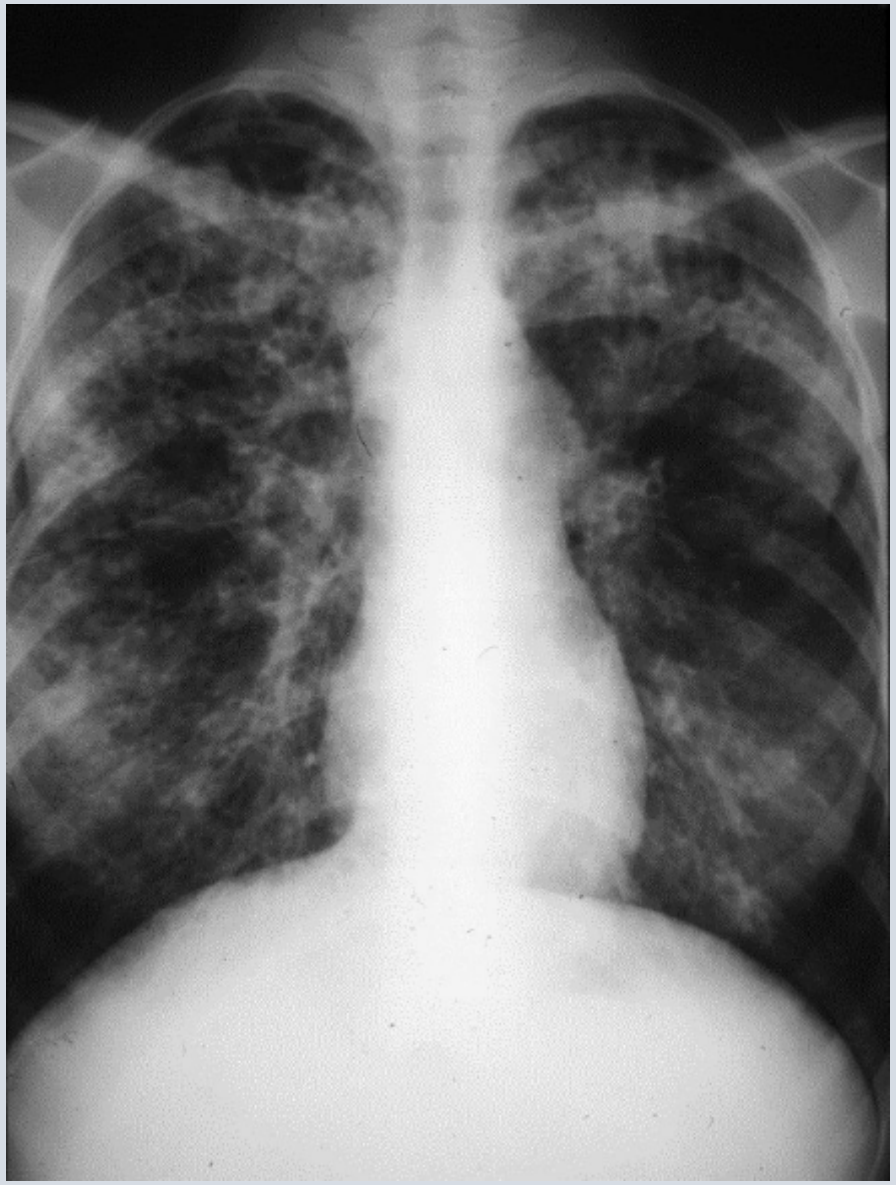


Question 1

- 60 y/o F with COPD
- O2 sats 89%
- Loud pulmonic component S2
- Arterial PO2 57 mm Hg
- Mean PAP 52 mm Hg

- Which of the following is the most appropriate treatment?
  - A. Daily prednisone
  - B. Long-term oxygen therapy
  - C. Overnight pulse oximetry
  - D. Repeat pulmonary rehabilitation

Question 1 – Answer B (long term oxygen therapy)

- Criteria for O2 in patients with COPD:
  - **Arterial PO2 ≤ 55 mm Hg or O2 sat ≤ 88%** with or without hypercapnia
  - and/or
  - Evidence of
    - **pulmonary hypertension,**
    - **peripheral edema suggesting right-sided heart failure,** or
    - **polycythemia,**
    - *** in combination with an arterial pO2 < 60 mm Hg or O2 sat < 88%

- Other answers:
  - Systemic glucocorticoids – for acute exacerbation
    - PO glucocorticoids usually do the job **(5 days)**
    - Consider IV if a severe exacerbation, non-response to oral glucocorticoids at home, who are unable to take oral medication, or who may have impaired absorption due to decreased splanchnic perfusion
  - Nocturnal oximetry – LTOT already established
  - Pulmonary rehab
    - Recommended for all symptomatic pts with COPD and an FEV1 <50% predicted and specifically those hospitalized with an acute exacerbation of COPD; can also be considered in symptomatic or exercise-limited pts with higher FEV1

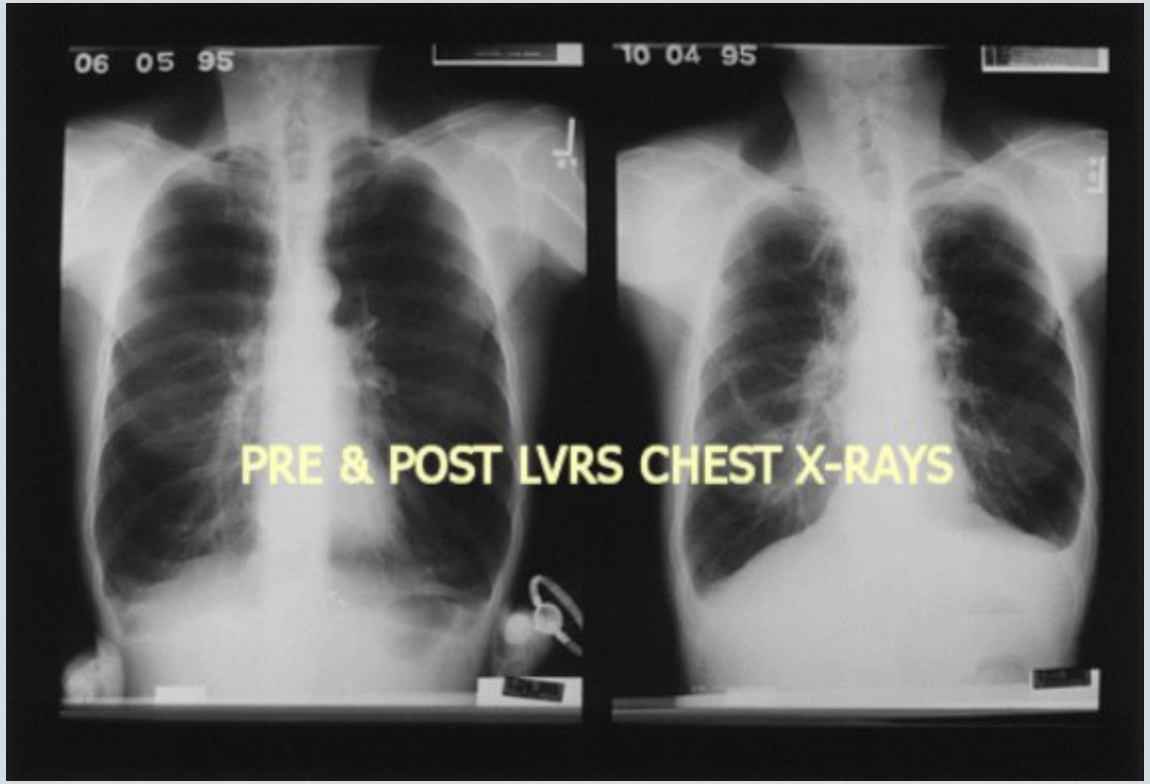
Question 2

- 55 y/o M with severe COPD
- 2 exacerbations in the past year
- Completed pulm rehab without much improvement
- Upper lobe emphysematous changes on imaging
- FEV1 40% predicted and DLCO 25% predicted
- 6 min walk 240 m

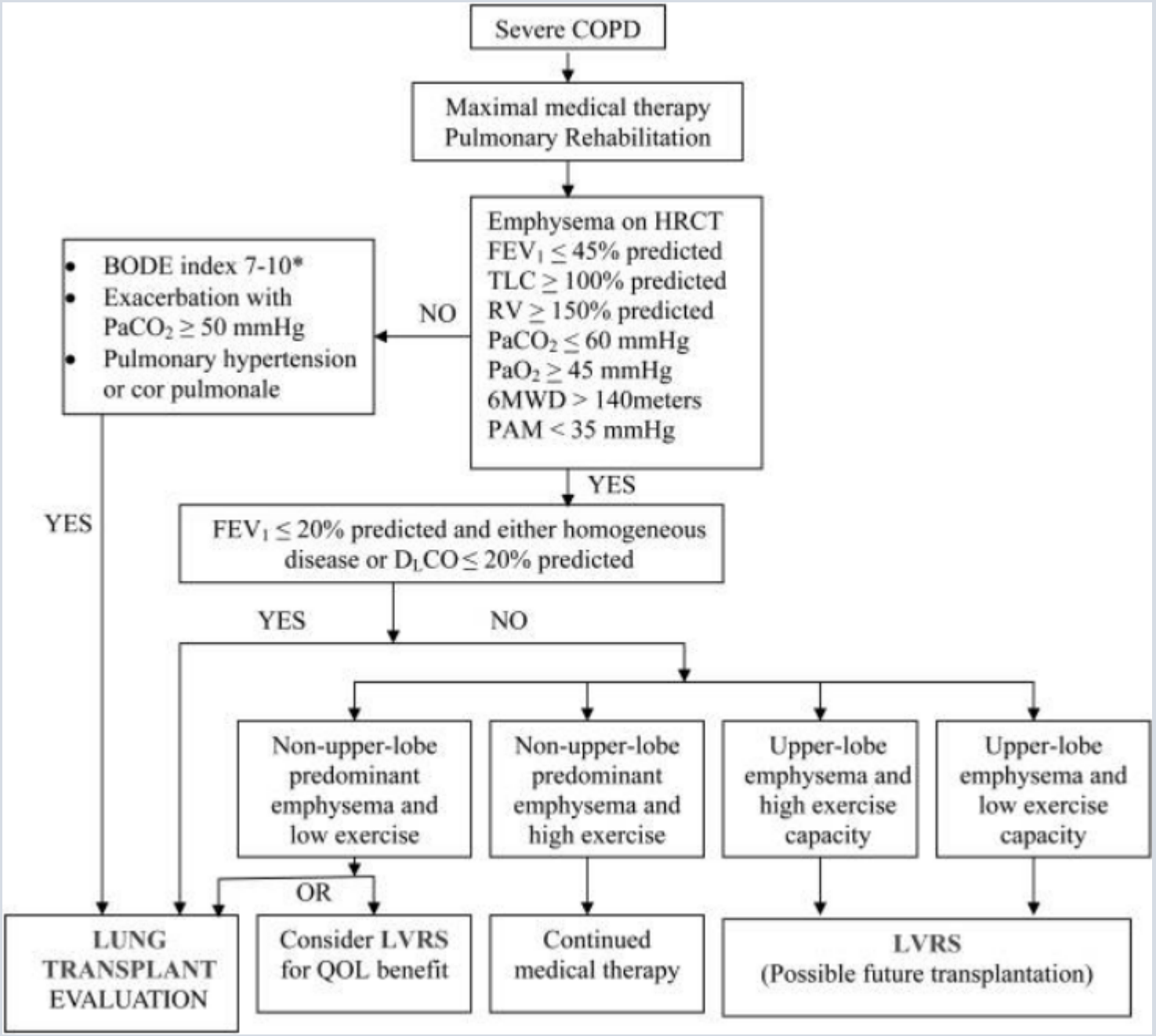
- Which of the following is most likely to benefit this patient?
  - A. Change fluticasone/salmeterol to fluticasone/vilanterol
  - B. Daily prednisone
  - C. Lung transplantation
  - D. Lung volume reduction surgery

Question 2 – Answer D (lung volume reduction surgery)

- Our patient:
  - Severe COPD, 2 exacerbations in the last year
  - Completed pulmonary rehab
  - Emphysematous changes in the b/l upper lobes
  - FEV1 40% of predicted
  - 6 min walk distance 240 m



LVRS vs. Transplant in COPD



Question 3

- 61 y/o M admitted to ICU with COPD, intubated x 5 days
- Treated for acute COPD exacerbation
- Fails spontaneous ventilation trial after 5 days with increased RR and WOB; appears fatigued but is awake/alert

- Which of the following is the most appropriate management?
  - A. Extubate now and initiate bilevel noninvasive positive pressure ventilation immediately
  - B. Extubate now and provide supplemental oxygen via nasal cannula
  - C. Recommend tracheostomy placement
  - D. Repeat spontaneous breathing trial in 1 hour

Question 3 – Answer A (extubate now and initiate bilevel noninvasive positive pressure ventilation immediately)

- In patients with COPD and hypercapnia, extubation followed by NIPPV may decrease ICU LOS and improve survival

- Other indications for NIPPV:
  - NIV decreases need for intubation in pts with moderate to severe exacerbations of COPD; intubation is indicated if no clinical response or ABG worsens after start of NIV
  - NIV reduces risk of intubation in pts with cardiogenic pulmonary edema
  - NIV can be used to support pts with ARF due to OHS
  - Hypoxemic respiratory failure, such as with severe PNA or ARDS, use is controversial

Contraindications to NIPPV

Contraindications to noninvasive positive pressure ventilation

Cardiac or respiratory arrest
Nonrespiratory organ failure that is acutely life-threatening
Severe encephalopathy (eg, GCS <10)
Severe upper gastrointestinal bleeding
Hemodynamic instability or unstable cardiac arrhythmia
Facial or neurological surgery, trauma, or deformity
Upper airway obstruction
Inability to cooperate/protect airway
Inability to clear secretions
High risk for aspiration

Question 4

- 72 y/o M evaluated for fever (38.3) after 1 week of being on mechanical ventilation for AHRF due to influenza
- Sputum production has increased and has needed increase in FiO2 to maintain oxygenation
- Increase in leukocytosis
- CXR with worsening pulmonary infiltrates

- Which of the following is the most appropriate next step in management?
  - A. Begin empiric ceftriaxone and azithromycin
  - B. Chest physiotherapy
  - C. Deep sampling of the lower respiratory tract
  - D. Substitute zanamivir for oseltamivir

Question 4 – Answer C (deep sampling of the lower respiratory tract)

- Ventilator-associated pneumonia (VAP)
  - Onset 48 hours after endotracheal intubation
  - Consider with:
    - Fever
    - Leukocytosis/ leukopenia
    - Increased purulent secretions
    - New or progressive pulmonary infiltrates
    - Worsening vent parameters

- IDSA Guidelines (2016):
- III. In Patients With Suspected HAP (Non-VAP), Should Treatment Be Guided by the Results of Microbiologic Studies Performed on Respiratory Samples, or Should Treatment Be Empiric?
- Recommendation
- 1. We suggest that patients with suspected HAP (non-VAP) be treated according to the results of microbiologic studies per- formed on respiratory samples obtained noninvasively, rath- er than being treated empirically

Other choices:

- Azithro/ceftriaxone – good choice for CAP, but pts has been hospitalized and on mechanical ventilation for 7 days and at risk for MDR organisms
- Chest physiotherapy – useful for assisting with removal of secretions in pts with COPD, CF, ciliary dyskinesia, but will not help with diagnosis
- Zanamivir - Initially improved with oseltamivir, so switching to different antiviral unlikely to help, and clinical picture more c/w VAP than oseltamivir resistance

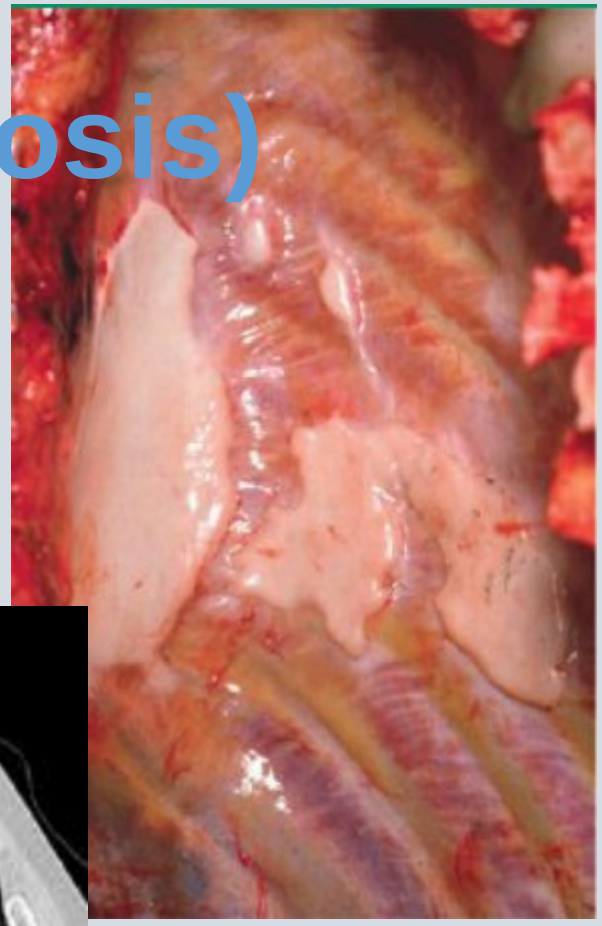
Question 5

- 72 y/o M with 2 year hx of cough and 1 year of increasing dyspnea, cough
- 15-pack-year smoking hx, quit 40 years ago
- Construction worker x 40 years
- Imaging with calcified pleural plaques b/l and basilar honeycombing
- PFTs with restrictive pattern

- Which of the following is the most likely diagnosis?
  - A. Asbestosis
  - B. COPD
  - C. Hypersensitivity pneumonitis
  - D. Idiopathic pulmonary fibrosis

Question 5 – Answer A (asbestosis)

- Occupational exposures:
  - Construction, automotive servicing, ship building
  - Inhalation of asbestos fibers
- Latency period (15-35 years)
- PFTs – restriction
- Pleural disease:
  - Benign asbestos pleural effusion
  - Pleural plaques and diffuse pleural thickening, malignant mesothelioma



Job risk for asbestos

Occupations and industries associated with asbestosis

Occupations	Industries
Plumbers	Construction
Pipefitters	Shipbuilding and repairing
Steamfitters	Chemicals and other manufacturing
Electricians	Nonmetallic mineral stone products
Insulation workers	Railways
Carpenters	Yarn, thread, and fabric mills
Laborers	Trucking
Boilermakers	Plastic and rubber manufacture
Welders and cutters	
Janitors	

Question 6

- 75 y/o M with hx of IPF x 5 years, admitted with 2 weeks of increased SOB
- 2 mos ago, required increase in supplemental O2 to 5L/min O2
- Treated with broad spectrum abx and IV steroids
- PE ruled out
- ABG with pH 7.32, PCO2 55 mm Hg and PO2 50 mm Hg
- High res CT shows progression of fibrosis

- Which of the following is the most appropriate treatment?
  - A. Additional intravenous methylprednisolone
  - B. Hospice care
  - C. Intubation and mechanical ventilation
  - D. Lung transplantation

Question 6 – Answer B (hospice care)

- The American Thoracic Society recommends palliation of symptoms rather than intubation and mechanical ventilation for patients with respiratory failure due to idiopathic pulmonary fibrosis
- Mean survival in pts with IPF is 3-5 years
- Our patient:
  - High baseline supplemental O2 component now with acute hypercapnia and hypoxia refractory to high flow O2
  - Advanced age, comorbidities and advanced lung disease make him a poor candidate for lung transplant

Question 7

- 57 y/o F with 6 mos of exertional dyspnea
- BP 144/88
- Prominent S2
- CXR with increased pulm vascular markings
- ECHO with DD and RVSP 52 mm Hg

- Which of the following is the most appropriate initial management?
  - A. Epoprostenol
  - B. Lisinopril
  - C. Right heart catheterization
  - D. Sildenafil

Question 7 – Answer B (lisinopril)

Table 1. Clinical Classification of Pulmonary Hypertension

Classification	*Targeted treatment available?*
Group 1*: Pulmonary arterial hypertension	Yes
Including idiopathic, heritable, and HIV-associated; systemic sclerosis and other connective tissue disease; congenital heart disease; schistosomiasis; drug- and toxin-induced	
Group 2: Pulmonary hypertension due to left heart disease	No
Including systolic and diastolic dysfunction and valvular heart disease	
Group 3: Pulmonary hypertension due to lung diseases and/or hypoxia	No
Including chronic obstructive pulmonary disease, sleep-disordered breathing, and interstitial lung disease	
Group 4: Chronic thromboembolic pulmonary hypertension	Yes
Group 5: Multifactorial pulmonary hypertension	No
Including metabolic, systemic, and hematologic disorders (sickle cell disease), and others	

HIV = human immunodeficiency virus.

**—Also includes 1′ (pulmonary venoocclusive disease and/or pulmonary capillary hemangiomatosis) and 1″ (persistent pulmonary hypertension of the newborn).*

Information from references 3, 4, and 6.

Pulmonary HTN – AFP 2016

SORT: KEY RECOMMENDATIONS FOR PRACTICE

Clinical recommendation	*Evidence rating*	*References*
Echocardiography is the recommended initial test in the evaluation of patients with suspected pulmonary hypertension.	C	1-3, 22
Results from invasive hemodynamic testing with right heart catheterization, preferably at an expert center, should be obtained before initiating treatment with vasodilator therapy in patients with pulmonary arterial hypertension.	C	1-3
In patients with pulmonary hypertension due to lung disease or left heart disease, treatment should focus on optimizing comorbid conditions.	C	3, 9-11
In patients with pulmonary hypertension and hypoxia, supplemental oxygen should be administered to maintain saturation above 90%.	C	11, 28, 29
Use of vasodilator therapies in patients with pulmonary hypertension due to lung disease or left heart disease is potentially harmful and not recommended.	C	10, 11, 26
Patients with chronic thromboembolic pulmonary hypertension should receive lifelong anticoagulation in the absence of contraindications.	C	25
Patients with pulmonary hypertension should receive seasonal influenza vaccination and age-appropriate pneumococcal vaccination, unless contraindicated.	A	1, 31, 32
Perioperative assessment of patients with pulmonary hypertension should include echocardiographic assessment of right ventricular function.	C	33

A = consistent, good-quality patient-oriented evidence; B = inconsistent or limited-quality patient-oriented evidence; C = consensus, disease-oriented evidence, usual practice, expert opinion, or case series. For information about the SORT evidence rating system, go to http://www.aafp.org/afpsort.

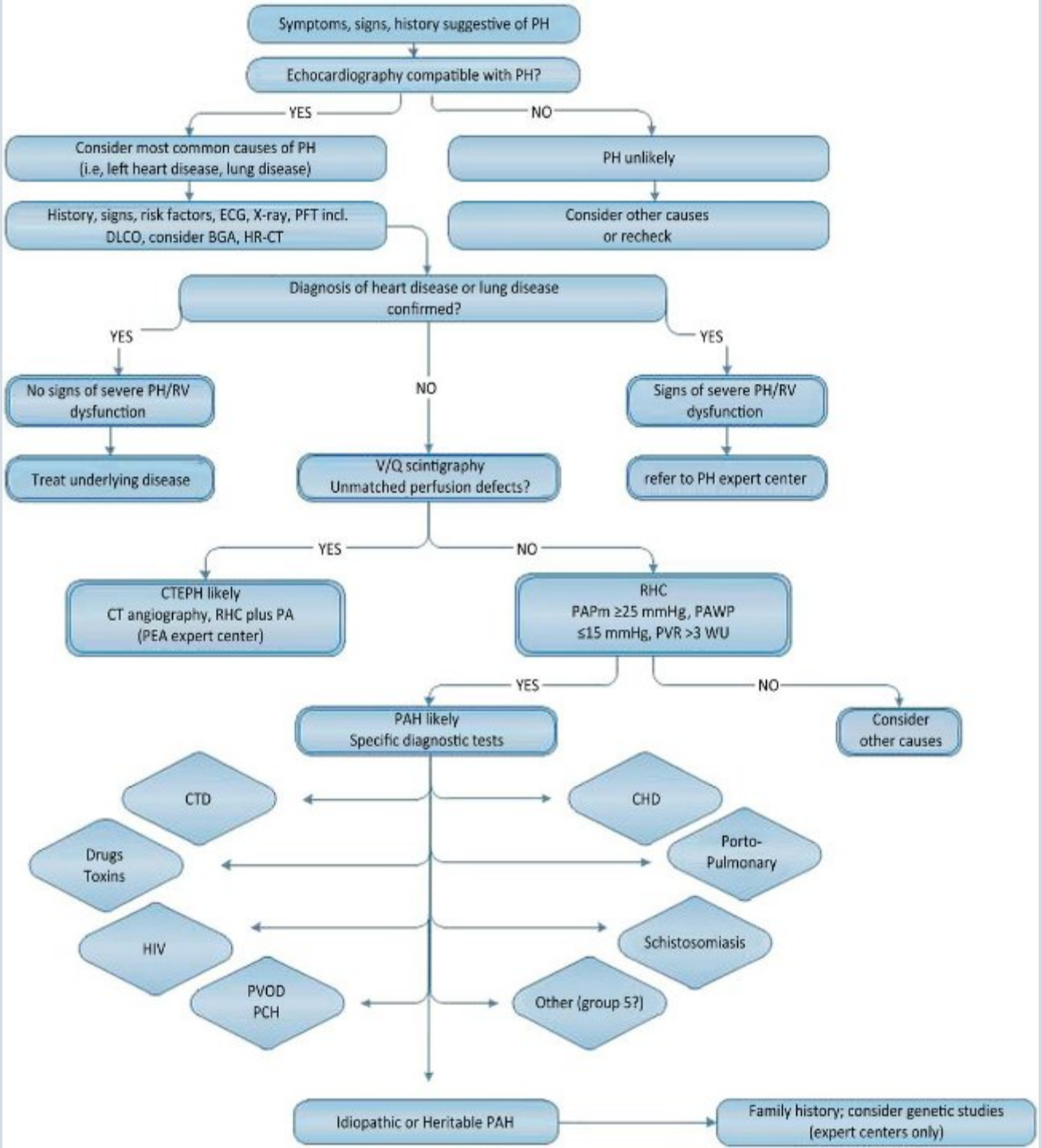
Question 8

- 53 y/o F with 3 mos of progressively worsening exertional dyspnea
- 18 mos ago had RLE DVT attributed to HRT
- Completed 6 mos of warfarin
- O2 sats 89%
- Single loud S2
- Nml CXR
- TTE shows RVSP 52 mm Hg

- Which of the following is the most appropriate diagnostic test to perform next?
  - A. CT angiography of the chest
  - B. Polysomnography
  - C. Pulmonary angiography
  - D. Ventilation-perfusion scan

Question 8 – Answer D (ventilation-perfusion scan)

- Chronic thromboembolic pulmonary hypertension (CTEPH) – group 4 pulmonary HTN
  - V/Q scan (96% sensitivity) > CTA (51% sensitivity) for dx of CTEPH
  - Will need f/u RHC and pulmonary angiography

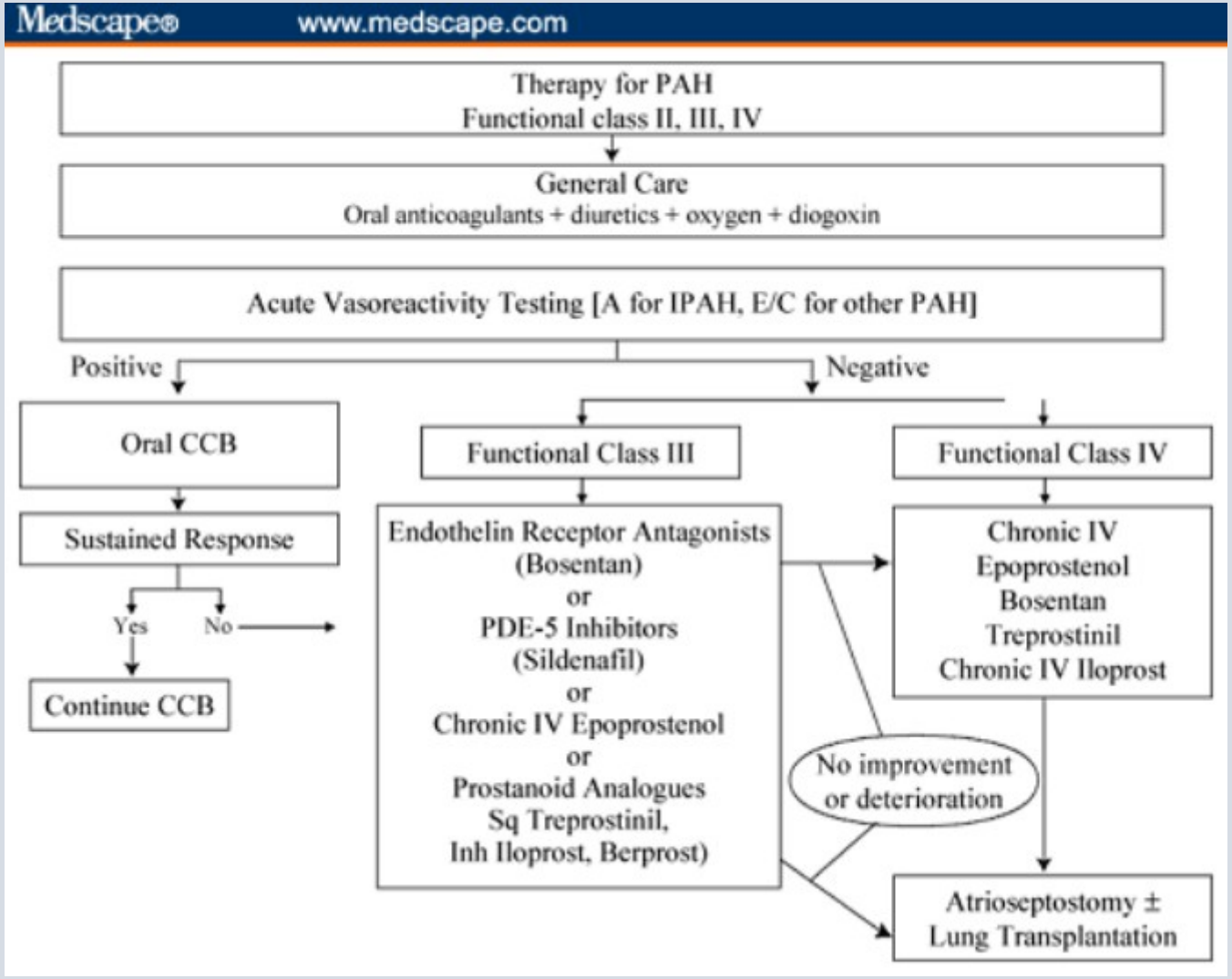

Symptoms, signs, history suggestive of PH
Echocardiography compatible with PH?
YES
NO
Consider most common causes of PH (i.e, left heart disease, lung disease)
PH unlikely
History, signs, risk factors, ECG, X-ray, PFT incl. DLCO, consider BGA, HR-CT
Consider other causes or recheck
Diagnosis of heart disease or lung disease confirmed?
YES
NO
YES
No signs of severe PH/RV dysfunction
Signs of severe PH/RV dysfunction
Treat underlying disease
V/Q scintigraphy Unmatched perfusion defects?
refer to PH expert center
YES
NO
CTEPH likely CT angiography, RHC plus PA (PEA expert center)
RHC PAPm ≥25 mmHg, PAWP ≤15 mmHg, PVR >3 WU
YES
NO
PAH likely Specific diagnostic tests
Consider other causes
CTD
CHD
Drugs Toxins
Porto-Pulmonary
HIV
Schistosomiasis
PVOD PCH
Other (group 5?)
Idiopathic or Heritable PAH
Family history; consider genetic studies (expert centers only)

Question 9

- 35 y/o F with 6 mos of exertional dyspnea (climbing a flight of stairs or walking 2 blocks)
- Prominent S2
- RHC shows mean PAP 35 mm Hg and PCWP 10 mm Hg
- No change in mean PAP with vasoreactivity testing

- Which of the following is the most appropriate management?
  - A. Epoprostenol
  - B. Nifedipine
  - C. Restriction of physical activity
  - D. Sildenafil

Question 9 – Answer D (sildenafil)



Vasoreactivity testing

- Administration of a short-acting vasodilator (inhaled nitric oxide) followed by measurement of hemodynamic response; positive response is a decrease in mean pulmonary artery pressure of 10 mm Hg
- Identifies a minority of pts (<10%) in whom PAH is due primarily to increased pulmonary vascular tone as opposed to pulmonary vascular remodeling
- More favorable prognosis and respond well to treatment with high dose CCBs

Question 10

- 58 y/o F with daily wheezing and breathlessness during allergy season
- Limits activities several times per week
- FEV1 74% or predicted with 18% bronchodilator response

- In addition to a short-acting β_2-agonist, which of the following is the most appropriate treatment?
  - A. Add a leukotriene antagonist
  - B. Add a low-dose inhaled glucocorticoid
  - C. Add a low-dose inhaled glucocorticoid and long-acting β_2-agonist
  - D. Recommend daily oral antihistamine use

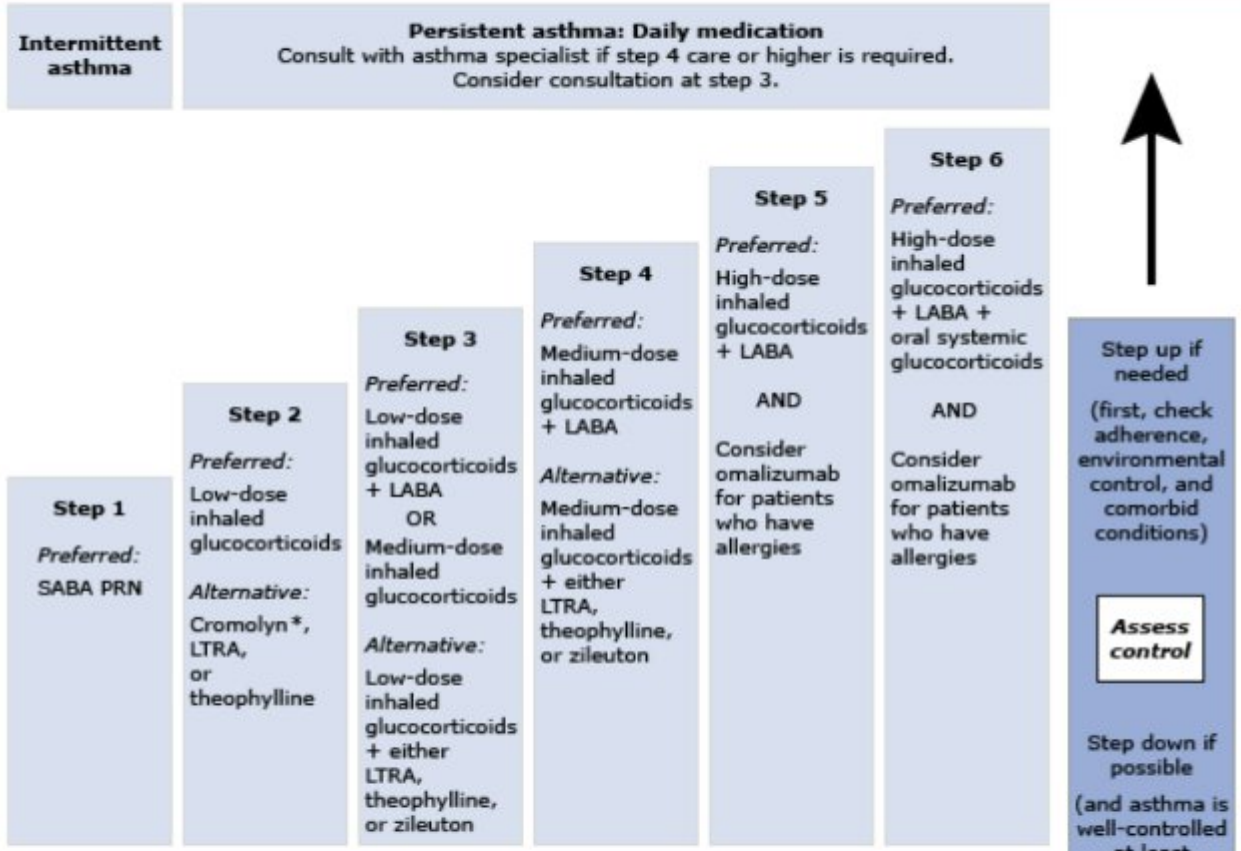
Question 10 – Answer C (add a low-dose inhaled glucocorticoid and long-acting β_2-agonist)

- Our patient:
  - Daily wheezing and breathlessness
  - Limitation in activities several times per week
  - FEV1 74% predicted with bronchodilator response

Assessing asthma control in youths greater than or equal to 12 years of age and adults

Components of control		Classification of asthma control (youths ≥12 years of age and adults)		
		Well controlled	**Not well controlled**	**Very poorly controlled**
Impairment	Symptoms	≤2 days/week	>2 days/week	Throughout the day
	Nighttime awakenings	≤2x/month	1 to 3x/week	≥4x/week
	Interference with normal activity	None	Some limitation	Extremely limited
	Short-acting beta$_2$-agonist use for symptom control (not prevention of EIB)	≤2 days/week	>2 days/week	Several times per day
	FEV_1 or peak flow	>80 percent predicted/personal best	60 to 80 percent predicted/personal best	<60 percent predicted/personal best
	Validated questionnaires			
	ATAQ	0	1 to 2	3 to 4
	ACQ	≤0.75*	≥1.5	N/A
	ACT	≥20	16 to 19	≤15
Risk	**Exacerbations**	**0 to 1/year**	**≥2/year (see footnote)**	
		Consider severity and interval since last exacerbation		
	Progressive loss of lung function	Evaluation requires long-term follow-up care		
	Treatment-related adverse effects	Medication side effects can vary in intensity from none to very troublesome and worrisome. The level of intensity does not correlate to specific levels of control but should be considered		

Stepwise approach for managing asthma in youths ≥12 years of age and adults



Question 11

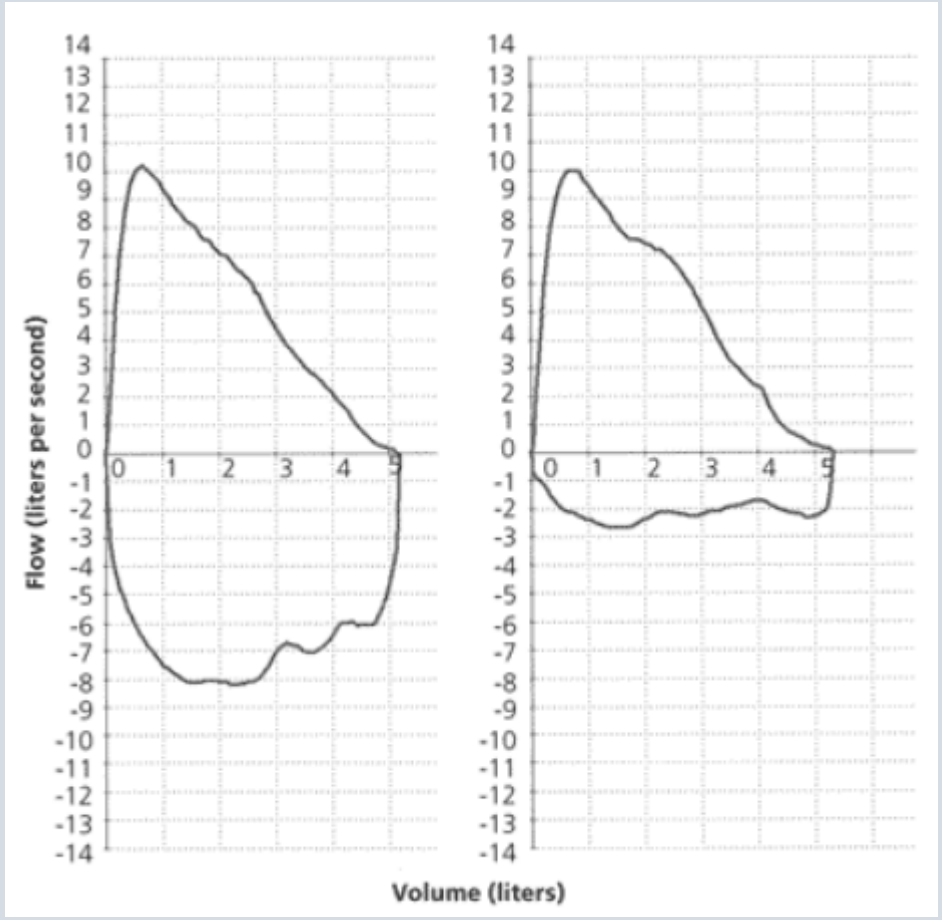
- 18 y/o M with 6 mos of chest and throat tightness; acute episodes of barking cough and prolonged wheeze
- Both with exertion and occasionally at rest
- Hx of moderate persistent asthma, which had been well controlled
- SABA not controlling his sxs

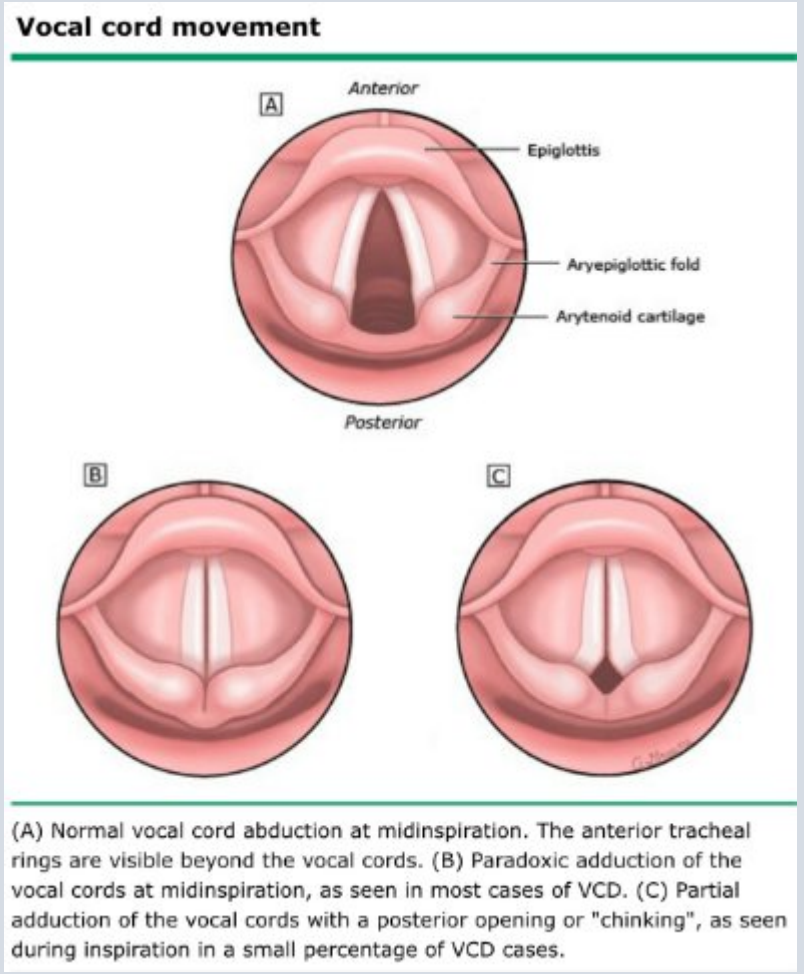
- Which of the following is the most appropriate next step in management?
  - A. Allergen immunotherapy
  - B. Echocardiography
  - C. Otolaryngology evaluation
  - D. Switch to a medium-dose inhaled glucocorticoid

Question 11 – Answer C (otolaryngology evaluation)

- Consider paradoxical vocal cord motion (VCD) when pts describe:
  - Mid-chest tightness with exposure to particular triggers such as strong irritants or emotions,
  - Difficulty breathing in, and
  - Symptoms that only partially respond to asthma medications
  - May also have: cough, dysphonia, stridor
- Often mis-diagnosed as asthma

Vocal cord dysfunction diagnosis





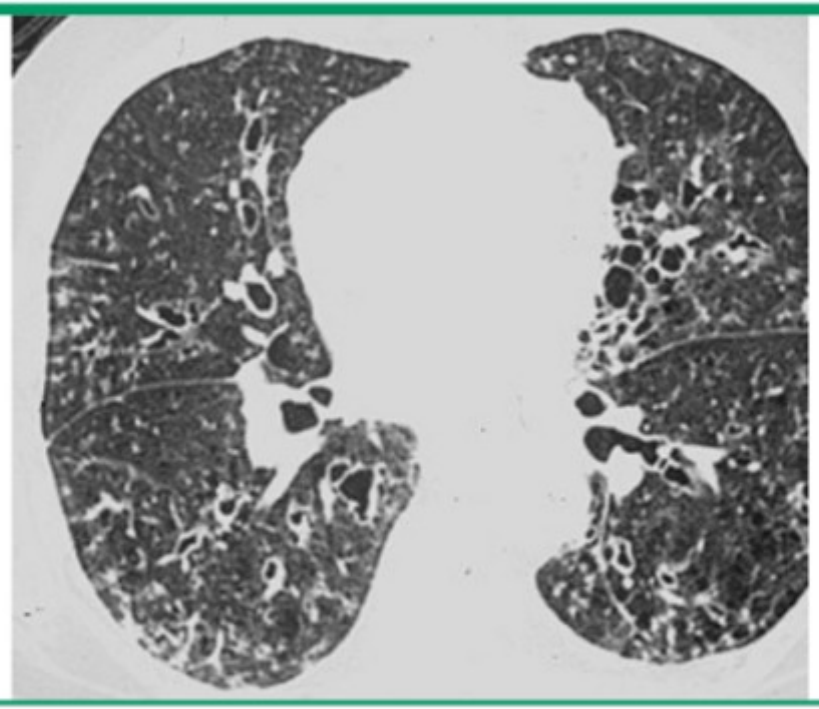
Question 12

- 35 y/o F seen in f/u for worsening asthma
- Asthma had previously been well controlled though with worsening sxs over the past year with increased wheezing and cough productive of dark-colored mucous
- Diminished airflow across upper right lung field
- WBC 10,500 with 15% eosinophils
- CXR with RUL infiltrate

- Which of the following is the most likely diagnosis?
  - A. Allergic bronchopulmonary aspergillosis
  - B. Cystic fibrosis
  - C. Eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome)
  - D. Hypersensitivity pneumonitis

Question 12 – Answer A (allergic bronchopulmonary aspergillosis)

Central bronchiectasis in allergic bronchopulmonary aspergillosis



Central bronchiectasis in a patient with allergic bronchopulmonary aspergillosis. Multiple dilated third and fourth generation bronchi are seen. Smaller peripheral bronchi filled with mucus account for the branching linear opacities in the distal lung parenchyma.

International Society for Human and Animal Mycology (ISHAM) working group diagnostic criteria for allergic bronchopulmonary aspergillosis

Predisposing conditions (one must be present):
Asthma
Cystic fibrosis
Obligatory criteria (both must be present):
Aspergillus skin test positivity or elevated IgE levels against *Aspergillus fumigatus*
Elevated total IgE concentration (typically >1000 IU/mL, but if the patient meets all other criteria, an IgE value <1000 IU/mL may be acceptable)
Other criteria (at least two must be present):
Precipitating serum antibodies to *A. fumigatus* or elevated serum *Aspergillus* IgG by immunoassay
Radiographic pulmonary opacities consistent with ABPA
Total eosinophil count >500 cells/microL in glucocorticoid-naïve patients (may be historical)

IgE: immunoglobulin E; ABPA: Allergic bronchopulmonary aspergillosis.

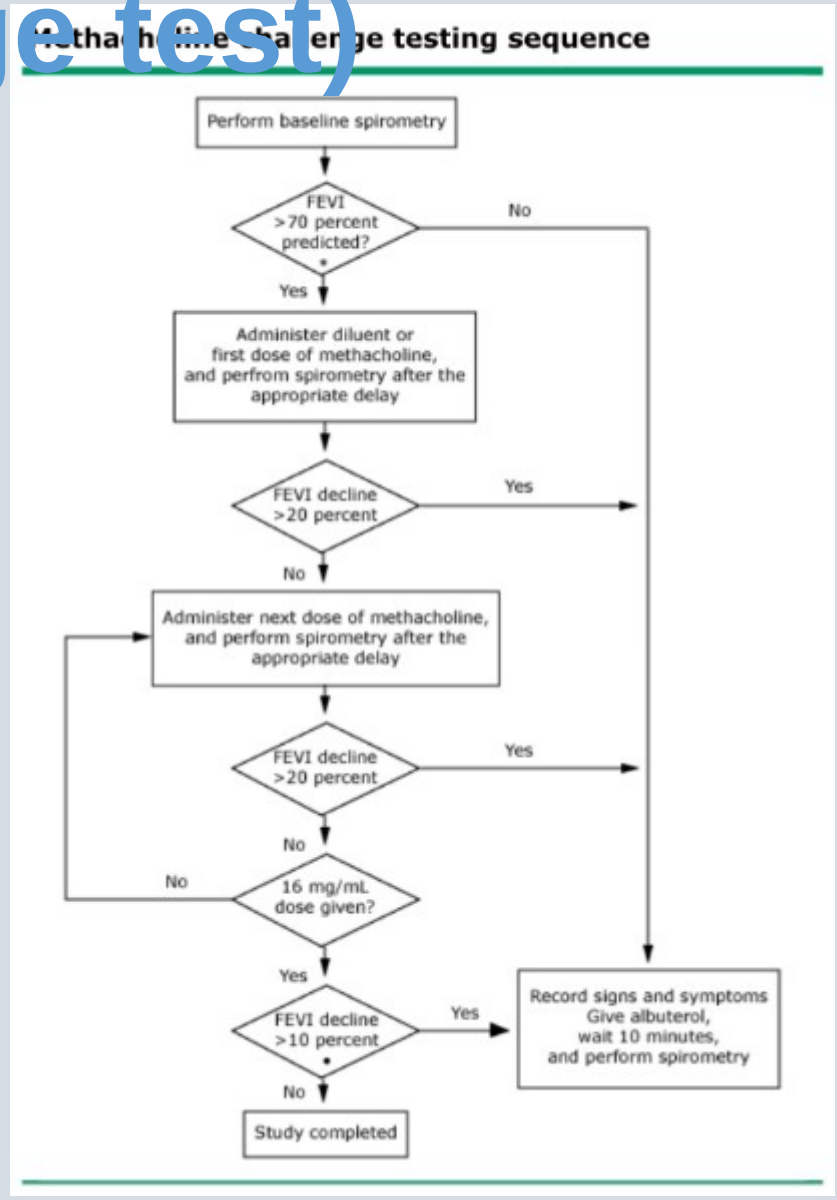
Question 13

- 24 y/o M with shortness of breath and occasional wheeze occurring several times per week
- Significant shortness of breath with exercise and cold weather
- Cough with exercise and at night
- Hx of seasonal allergies
- CXR nml
- Spirometry nml

- Which of the following is the most appropriate next step in management?
  - A. Methacholine challenge test
  - B. Nasal glucocorticoid
  - C. Short-acting β_2-agonist inhaler as needed
  - D. Clinical observation

Question 13 – Answer A (methacholine challenge test)

- Spirometry is often normal in between episodes and a methacholine challenge test can be performed if diagnosis of asthma is in question
- Indications:
  - Diagnosis of asthma
  - Assessment of response to therapy
  - Identification of specific asthma triggers



Contraindications to methacholine challenge:

TABLE 1

Contraindications to methacholine challenge testing: American Thoracic Society guidelines

Absolute contraindications

Severe airflow limitation: forced expiratory volume in 1 second (FEV_1) < 50% of predicted or < 1.0 L
Heart attack or stroke in last 3 months
Uncontrolled hypertension: systolic blood pressure > 200 mm Hg or diastolic pressure > 100 mm Hg
Known aortic aneurysm

Relative contraindications*

Moderate airflow limitation: FEV_1 < 60% of predicted or < 1.5 L
Inability to perform spirometry of acceptable quality
Pregnancy
Nursing mothers
Current use of cholinesterase inhibitor medication (for myasthenia gravis)

*Authors' additional relative contraindications: cerebral aneurysms; failure to withhold medications (may affect the test results) and upper- or lower-respiratory-tract infection within previous 2 to 6 weeks

CRAPO RO, CASABURI R, COATES AL, ET AL. GUIDELINES FOR METHACHOLINE AND EXERCISE CHALLENGE TESTING—1999. THIS OFFICIAL STATEMENT OF THE AMERICAN THORACIC SOCIETY WAS ADOPTED BY THE ATS BOARD OF DIRECTORS, JULY 1999. AM J RESPIR CRIT CARE MED 2000; 161:309–329.

CCJM, 2008

Question 14

- 20 y/o F seen in the ED for acute exacerbation of asthma
- She is in respiratory distress and ABG with acute acidosis
- She is intubated
- Peak inspiratory pressure rises to 45 cm H20, BP drops to 80/40, HR 115 and O2 sats 80%

- Which of the following is the most appropriate immediate next step in management?
  - A. Decrease the set respiration rate to 12/min
  - B. Decrease the tidal volume to 300 mL
  - C. Increase the inspiratory flow rate
  - D. Increase the sedative infusion rate and prepare for therapeutic paralysis
  - E. Temporarily disconnect the endotracheal tube from the ventilator

Question 14 – Answer E (temporarily disconnect the endotracheal tube from the ventilator)

- Auto-PEEP
  - Breath stacking on the ventilator in the setting of obstructive lung disease
  - Impairment of central venous return due to increased intrathoracic pressure
  - Worsening oxygenation due to low cardiac output and V/Q mismatch from hyperinflation
- Other answers:
  - ↓ RR, ↓ tidal volume and ↑ inspiratory flow rate will increase exhaled volume with each cycle
  - *** but, not appropriate interventions until after the increased intrathoracic pressure is released by temporarily disconnecting the ventilator circuit

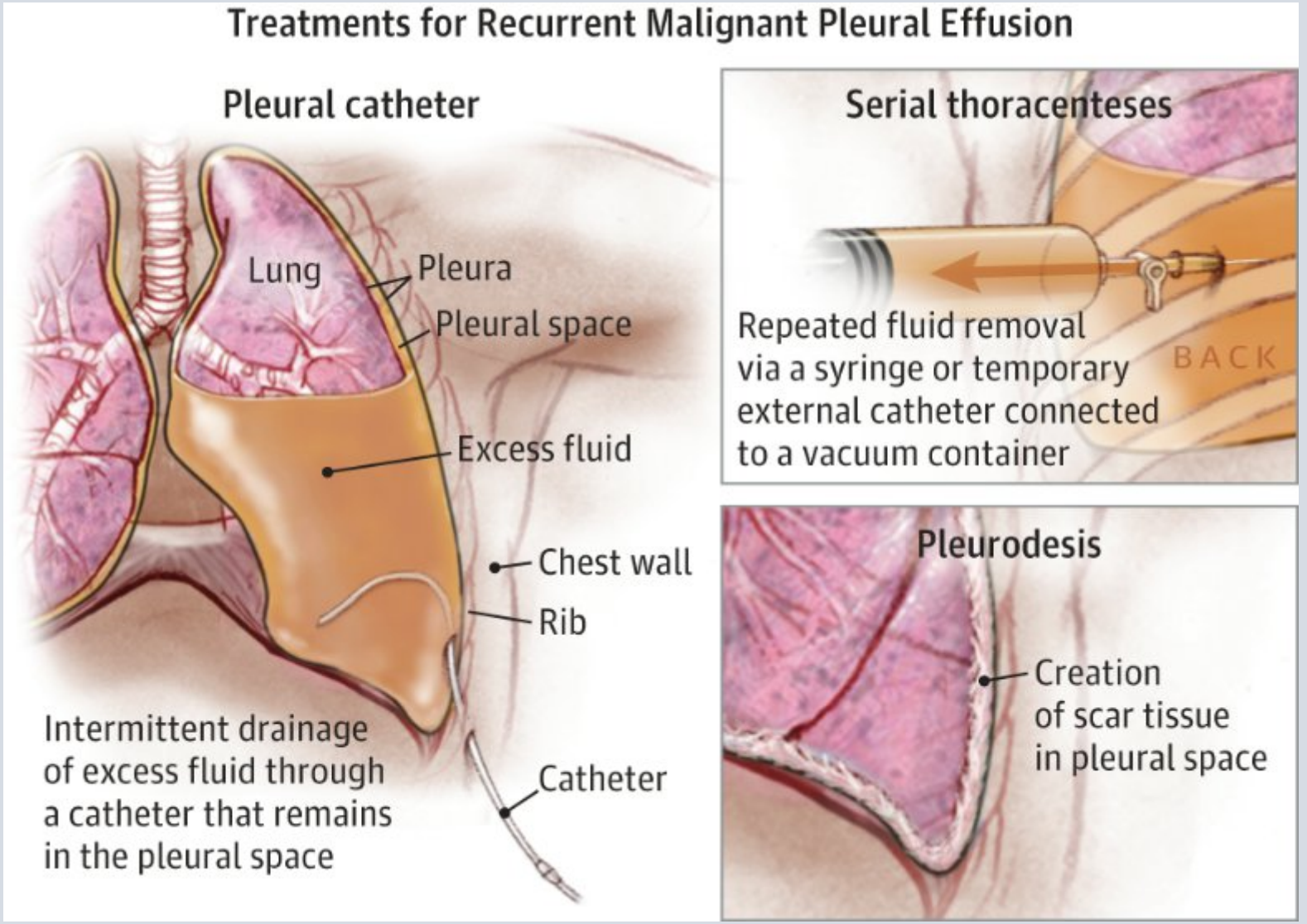
Question 15

- 66 y/o F with breast mass and 6 weeks of increasing dyspnea
- Pleuritic CP and weight loss
- Thoracentesis reveals serosanguinous fluid, exudative
- Gram stain negative
- Cytology negative

- In addition to evaluation of the breast mass, which of the following is the most appropriate management?
  - A. Closed pleural biopsy
  - B. Pleural fluid flow cytometry
  - C. Repeat thoracentesis and pleural fluid cytology
  - D. Thoracoscopic pleural biopsy

Question 15 – Answer C (repeat thoracentesis and pleural fluid cytology)

- Pleural fluid cytology 60% sensitive for diagnosis of malignant pleural effusion
  - 65% of positive results obtained on initial thoracentesis; 2nd sample provides additional 27% and 3rd, only 5%
- Closed pleural bx less sensitive than cytology for pleural malignancy
- Flow cytometry may be helpful with lymphocyte predominant effusion when lymphoma is a consideration
- Thorascopic pleural biopsy indicated for all undiagnosed exudative pleural effusions following 2 pleural fluid samplings – 90% sensitive for pleural malignancy


Treatments for Recurrent Malignant Pleural Effusion
Pleural catheter
Lung
Pleura
Pleural space
Excess fluid
Chest wall
Rib
Catheter
Intermittent drainage of excess fluid through a catheter that remains in the pleural space
Serial thoracenteses
Repeated fluid removal via a syringe or temporary external catheter connected to a vacuum container
BACK
Pleurodesis
Creation of scar tissue in pleural space

Question 16

- 18 y/o M found somnolent, emesis-covered
- Meds: sertraline
- Na 140, K 3.2, Cl 104, bicarb 6, BUN 14, glucose 180, lactate 2.8
- Plasma osmolality: 325
- ABG: pH 7.17, PCO2 17, PO2 120

- Which of the following is the most likely diagnosis?
  - A. Isopropyl alcohol poisoning
  - B. Methanol poisoning
  - C. Salicylate poisoning
  - D. Serotonin syndrome

Question 16 – Answer B (methanol poisoning)

- Encephalopathy, high AG and elevated osmolal gap consistent with methanol or ethylene glycol ingestion
  - AG = 30
  - Osmolal gap (measured – calculated)
    - Calculated: 2 x serum sodium + plasma glucose/18 + BUN/2.8
    - 2 (140) + 180/18 + 14/2.8 = 295
    - 325 – 295 = 30; nml is <10
- *** Methanol – converted to formic acid which is toxic to the retina
- *** Ethylene glycol – converted to oxalic acid which crystallizes in the renal tubules and kidney injury
- Other answers:
  - Isopropyl alcohol poisoning – elevated osmolal gap, but do not have AGMA
  - Salicylate toxicity – causes increase AGMA, but not elevation of the osmolol gap
  - Serotonin syndrome – fever, encephalopathy, agitation, muscle rigidity and hyperreflexia; not c/w anion gap or osmolal gap

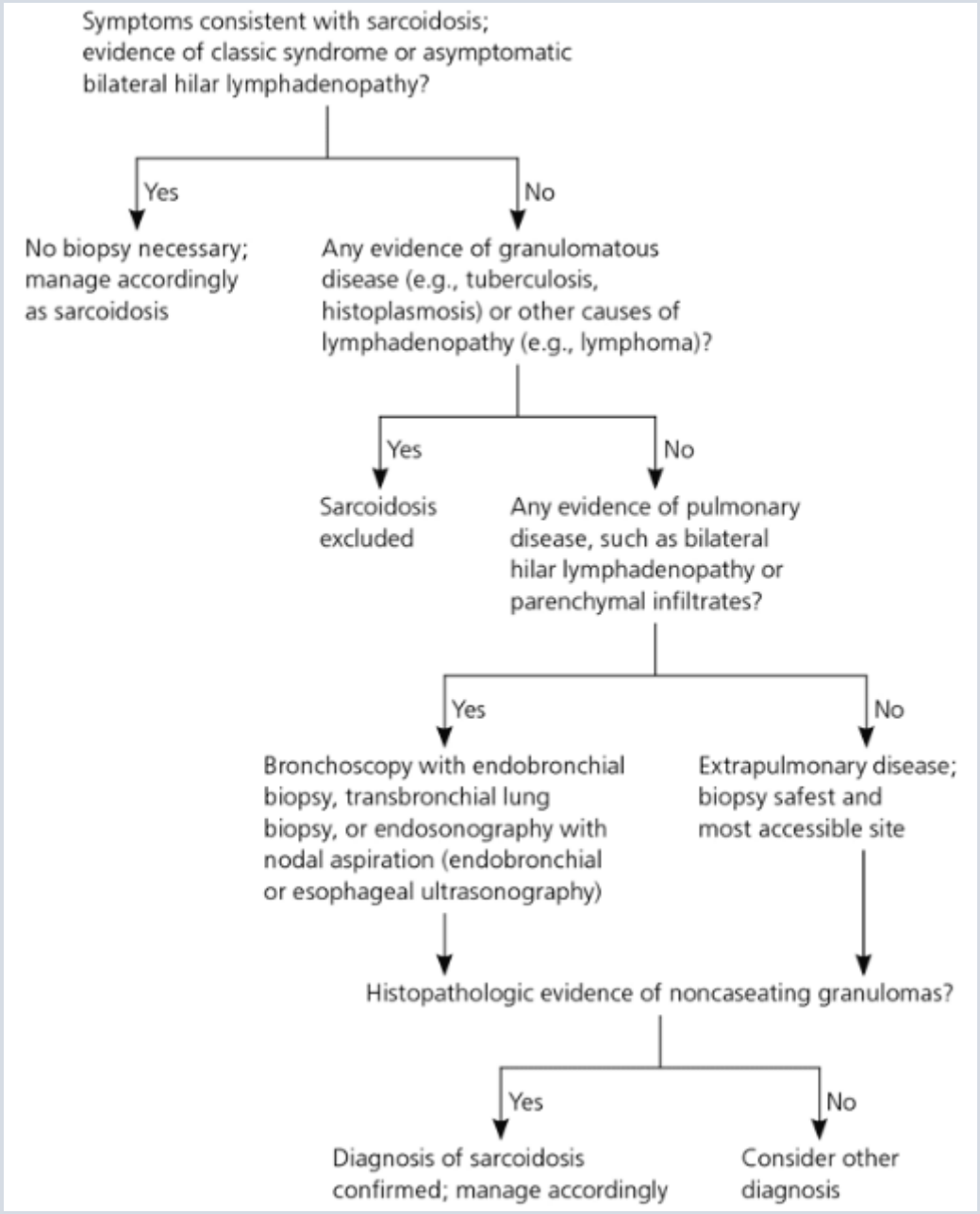
Question 17

- 28 y/o M with 6 mos of fatigue, increase in exertional dyspnea, and cough
- CXR with b/l hilar LAD and nml lung parenchyma
- CT chest with b/l hilar, mediastinal and subcarinal LAD with b/l small lung nodules with perihilar distribution
- TST negative

- Which of the following is the most appropriate next step in management?
  - A. Bronchoscopic biopsy
  - B. Empiric therapy with prednisone
  - C. Interferon-γ release assay
  - D. Measurement of angiotensin-converting enzyme level

Question 17 – Answer A (bronchoscopic biopsy)

- Sarcoid - multisystem granulomatous disease; 90% have lung involvement
  - Tissue bx usually required for diagnosis, except…
  - Treat with glucocorticoids, but only if clinical sxs from organ dysfunction, as there is a high rate of spontaneous remission

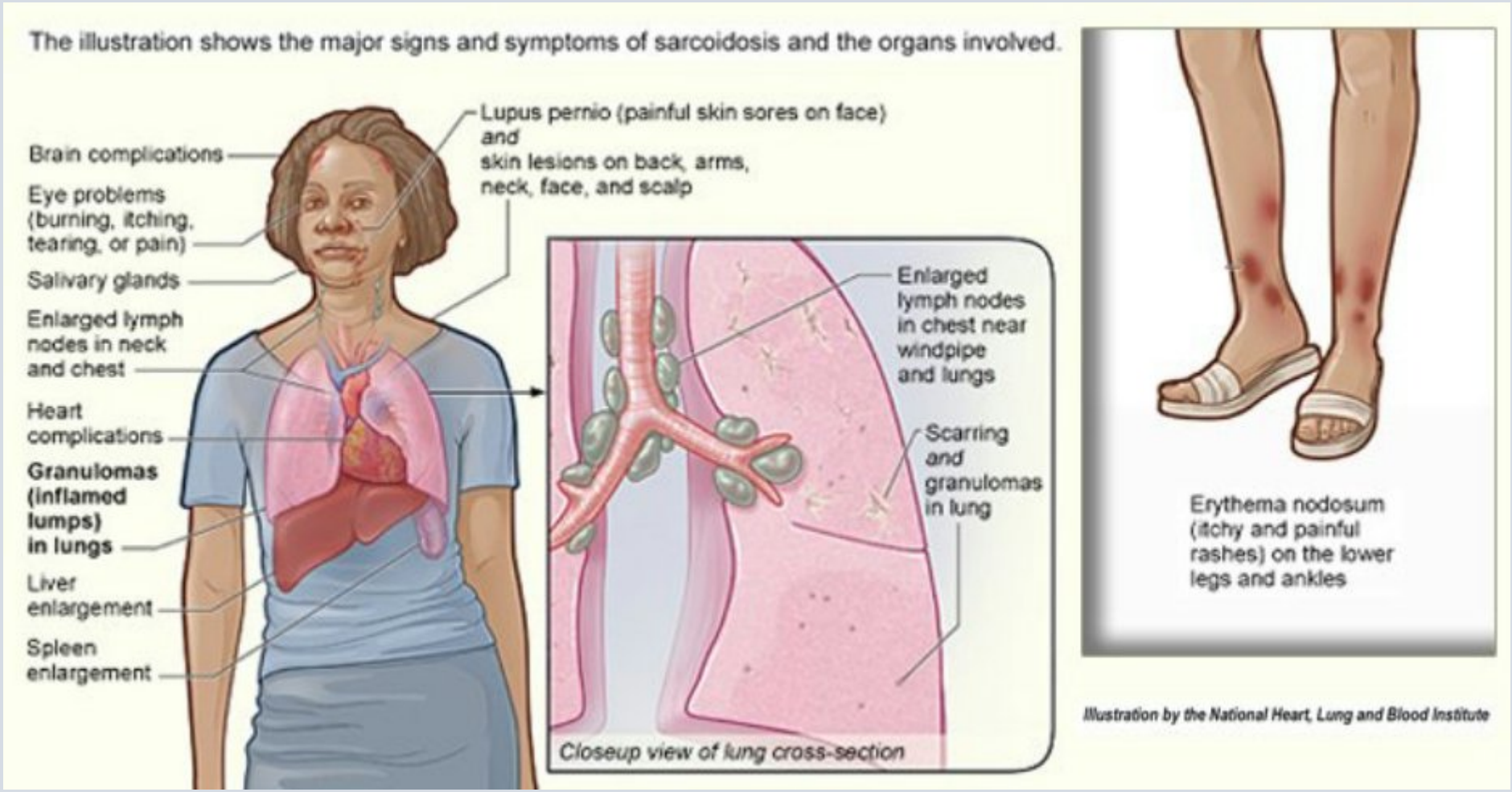


No biopsy needed if asx or classic syndrome:

Syndrome/Sign	Comments
Asymptomatic bilateral hilar lymphadenopathy	No evidence of fevers, malaise, or night sweats to suggest a malignancy
Löfgren syndrome	Bilateral hilar lymphadenopathy, migratory polyarthralgia, erythema nodosum, and fevers
Heerfordt syndrome	Anterior uveitis, parotiditis, fevers (uveoparotid fever), and facial nerve palsy

Diagnosis and Management of Sarcoidosis - AAFP

Multisystem granulomatous disease



Question 18

- 25 y/o M with hx of asthma and chronic productive cough, no dyspnea
- Recurrent sinus infections
- Meds: ICS and prn albuterol
- Polyps seen in nasal passages, lung exam with coarse breath sounds with occasional exp wheezing
- PFTs with obstruction

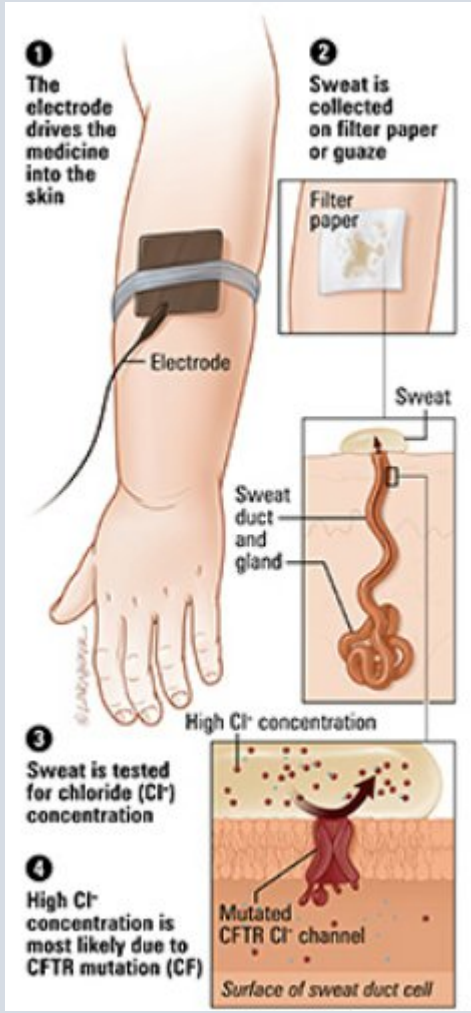
- Which of the following is the most appropriate diagnostic test to perform next?
  - A. α_1-Antitrypsin measurement
  - B. Antineutrophil cytoplasmic antibody assay
  - C. Sinus radiographs
  - D. Sweat chloride testing

Question 18 – Answer D (sweat chloride testing)

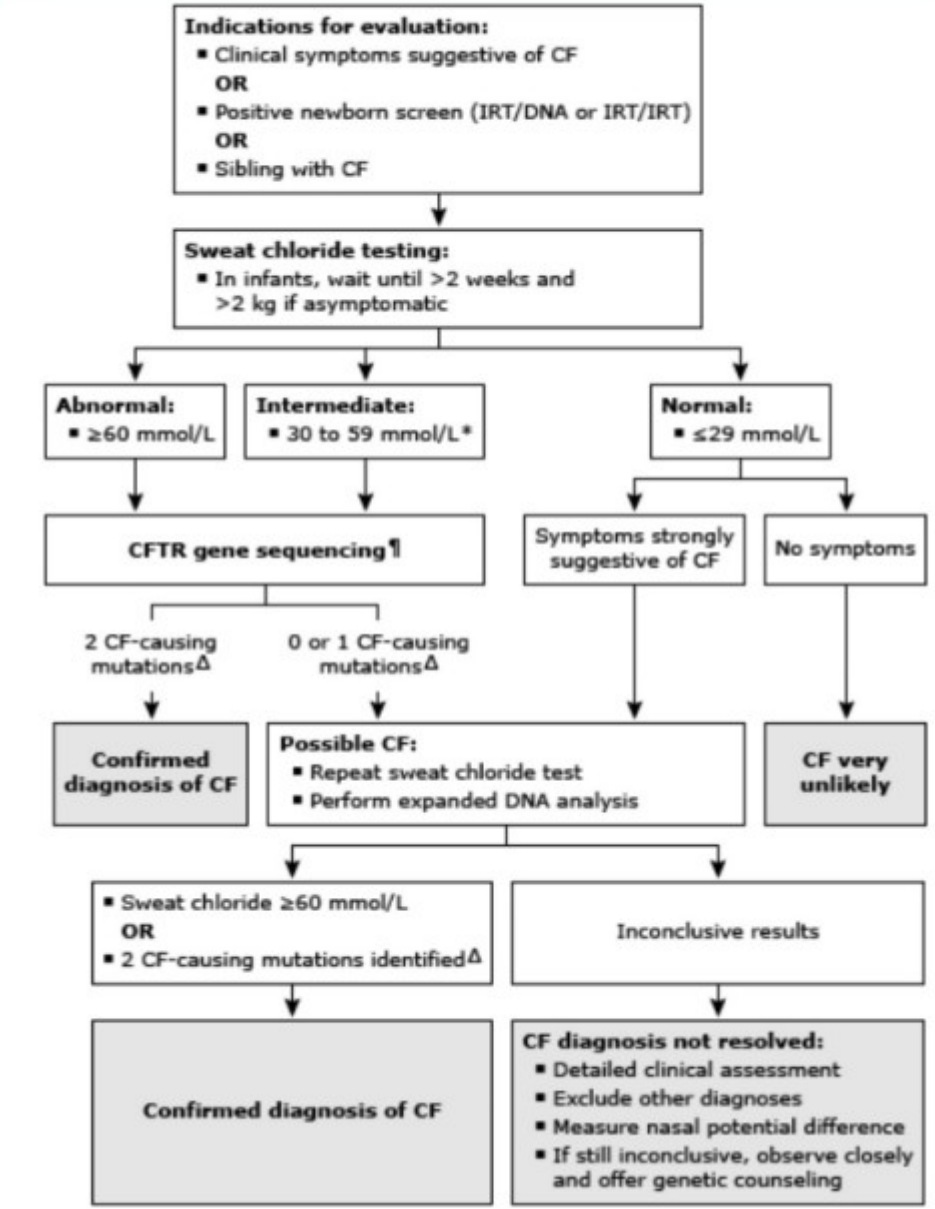
- Consider CF in young adults with
  - chronic asthma-like sxs
  - chronic sinusitis
  - nasal polyps
  - recurrent pancreatitis
  - male infertility
  - non-TB mycobacterial infection
  - ABPA, bronchiectasis
  - positive sputum cx for burkholderia cepacia and/or pseudomonas aeruginosa
- Diagnose CF with
  - CF-compatible clinical findings

  +
  - Sweat chloride testing, nasal potential difference

  Or
  - CFTR mutations



Algorithm for the diagnosis of cystic fibrosis



Other answers:

- **?1-antitrypsin measurement** – deficiency leads to accelerated emphysema and liver disease; bullous changes at the bases; liver and lung disease
- **antineutrophil cytoplasmic antibody assay** – useful in diagnosing GPA or eGPA; nml u/a suggests absence of kidney disease; bronchiectasis not characteristic of GPA, but rather nodues, diffuse opacities, transient pulm infiltrates and hilar LAD
- **sinus radiographs** – may help define extent of polyposis, but not underlying cause; furthermore, CT is the preferred imaging modality for sinus disease

Question 19

- 56 y/o M with hx of COPD p/w episode of hemoptysis, half-cup of bright red blood
- Intermittent non-productive cough x several weeks
- 55-pack-year smoking hx, continues to smoke
- CXR with mild hyperinflation and hazy infiltrate in the RLL
- No PE on chest CTA

- Which of the following is the most appropriate next step in management?
  - A. Bronchoscopy
  - B. Ceftriaxone and azithromycin
  - C. High-resolution chest CT
  - D. Upper endoscopy

Question 19 – Answer A (bronchoscopy)

- Massive hemoptysis = 100 mL-600 mL of blood
- Etiology:

Table. BATTLE CAMP in the Differential Diagnosis of Hemoptysis
B – Bronchitis, Bronchiectasis including cystic fibrosis
A – Abscess, Aspergilloma, Arteriovenous malformations, Anticoagulants
T – Tumor, Trauma causing lung contusion
T – Tuberculosis including bronchial artery aneurysms (Rasmussen's aneurysm)
L – Lung abscess, Lung cancer, Lupus pneumonitis
E – Embolism with lung infarction, Endometriosis of the lung (rare)
C – Coagulopathy, inherited, acquired, drug-induced, Cryptogenic hemoptysis from Dieulafoy disease, Carcinoid tumor
A – Alveolar hemorrhage from autoimmune disorders (eg, Goodpasture syndrome, systemic lupus erythematosus)
M – Mycetoma (eg, aspergilloma), Mitral stenosis (very rare today)
P – Pneumonia, acute or chronic with infectious pathogens including bacteria and fungi
AVM: arterio-venous malformations, SLE: systemic lupus erythematosus

Indications for bronchoscopy

- (1) evaluation of ned respiratory sxs associated with airway pathology (hemoptysis, stridor)
- (2) pulmonary infections, esp if progression despite appropriate empiric therapy or in immunocompromised hosts
- (3) diagnosis or staging of primary or metastatic cancer of the lung
- (4) diagnosis of an abnormal imaging finding such as pulm nodule, persistent infiltrate or atelectasis
- (5) diffuse parenchymal lung disease of unknown cause
- (6) therapeutic use (airway stenosis, foreign body aspiration, mucous plug, or local treatment of a lesion)

Other answers:

- Ceftriaxone and azithromycin
  - No sxs of infection as etiology of hemoptysis, so abx is not the right choice, furthermore, pulm parenchymal infections unlikely to cause massive hemoptysis
- High-resolution chest CT
  - HRCT useful to eval pulm parenchyma, ie. ILD eval, though does not adequately define endobronchial lesions
- Upper endoscopy
  - Sometimes hard to differentiate b/w hemoptysis and hematemesis, though if GI source were suspected, endoscopy would appropriate

Question 20

- 29 y/o F with shortness of breath while mountain climbing
- Elevation 3200 m (10,500 ft)
- O2 sat 86%, moderate resp distress
- Few bibasilar crackles on lung exam
- Receives supplemental O2 and arrangements made to descend

- Which of the following is the most appropriate adjunctive treatment for this patient?
  - A. Acetazolamide
  - B. Dexamethasone
  - C. Ibuprofen
  - D. Nifedipine

Question 20 – Answer D (nifedipine)

- 21% FiO2, though increase in altitude and decreasing barometric pressure reduces the amount of O2 available for gas exchange
- High altitude illness (HAI) more common at elevations ≥ 8200 feet
- RFs for HAI include destination altitude and rate of ascent

- HAPB (high altitude periodic breathing)
- Acute mountain sickness
  - HA, fatigue, N/V and disturbed sleep
  - Mild sxs: ASA, NSAIDs, antiemetics
- HACE (high-altitude cerebral edema)
  - Confusion, irritability, ataxic gait, coma
  - Immediate descent
- HAPE (high altitude pulmonary edema)
  - Cough, dyspnea, exertional intolerance
  - Supplemental O2 and rest
  - Vasodilators

High Altitude Illness Therapy

Table 3. Medications for the Prevention and Treatment of Altitude Illness

Medication	*Indication*	*Typical dosage*
Acetazolamide	Prevention and treatment of acute mountain sickness (first-line therapy)*	125 mg twice daily (prophylaxis) 250 mg twice daily (treatment)
Dexamethasone	Prevention of acute mountain sickness†	4 mg every 6 to 12 hours
	Treatment of acute mountain sickness†	4 mg every 6 hours
	Prevention of high-altitude cerebral edema†	4 mg every 6 hours
	Treatment of high-altitude cerebral edema†‡	8 mg, then 4 mg every 6 hours
Nifedipine (Procardia)	Prevention and treatment of high-altitude pulmonary edema†	20 mg every 8 to 12 hours
Salmeterol (Serevent)	Prevention and treatment of high-altitude pulmonary edema†	125 mcg every 12 hours
Sildenafil (Viagra)	Prevention and treatment of high-altitude pulmonary edema†	20 mg every 6 to 8 hours
Tadalafil (Cialis)	Prevention and treatment of high-altitude pulmonary edema†	10 mg every 12 hours

*—U.S. Food and Drug Administration (FDA)-approved indication.
†—Not an FDA-approved indication.
‡—Descent is mandatory.

Information from references 3, 7, and 16 through 29.

Bonus: air travel in pulmonary disease

- sPO2 < 92% indicates likely need supplemental O2 in flight
- 92% - 95% consider hypoxia altitude simulation testing
- If already on long-term supplemental O2, doubling the flow rate is typically adequate

- Pneumothorax
  - Current PTX is a contraindication to air travel
  - May consider air travel 2 weeks after resolution in pts with relatively nml lung parenchyma
  - With severe underlying lung disease (bullous emphysema, limited cardiopulmonary reserve, prior spontaneous PTX, air travel may be contraindicated