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Pulmonary Testing Chapter 1

A 74-year-old man is evaluated in the ICU for sepsis and hypotension. A central venous catheter has just been inserted into the left internal jugular vein under ultrasound visualization to start vasopressor therapy. As the catheter line is being secured, the patient suddenly becomes more hypotensive. Medications are propofol, piperacillin-tazobactam, and vancomycin.

Temperature is 37.8 °C (100.0 °F), blood pressure is 74/55 mm Hg, pulse rate is 118/min, and respiration rate is 18/min on mechanical ventilation. Oxygen saturation is 91% on Fio₂ of 1.00. Breath sounds are diminished bilaterally. Heart sounds are diminished.

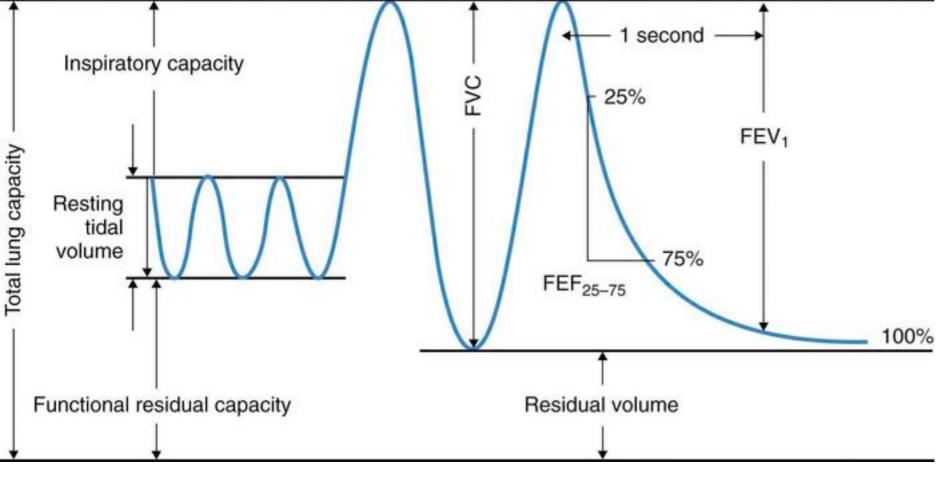
Which of the following is the most appropriate next management step?

A Chest CT

B Chest ultrasonography

C Needle thoracostomy

D Removal of central venous catheter



Obstructive lung disease: FEV1/FVC < 70%

Restrictive lung disease: Low FEV1, FVC, TLC (< 80%)

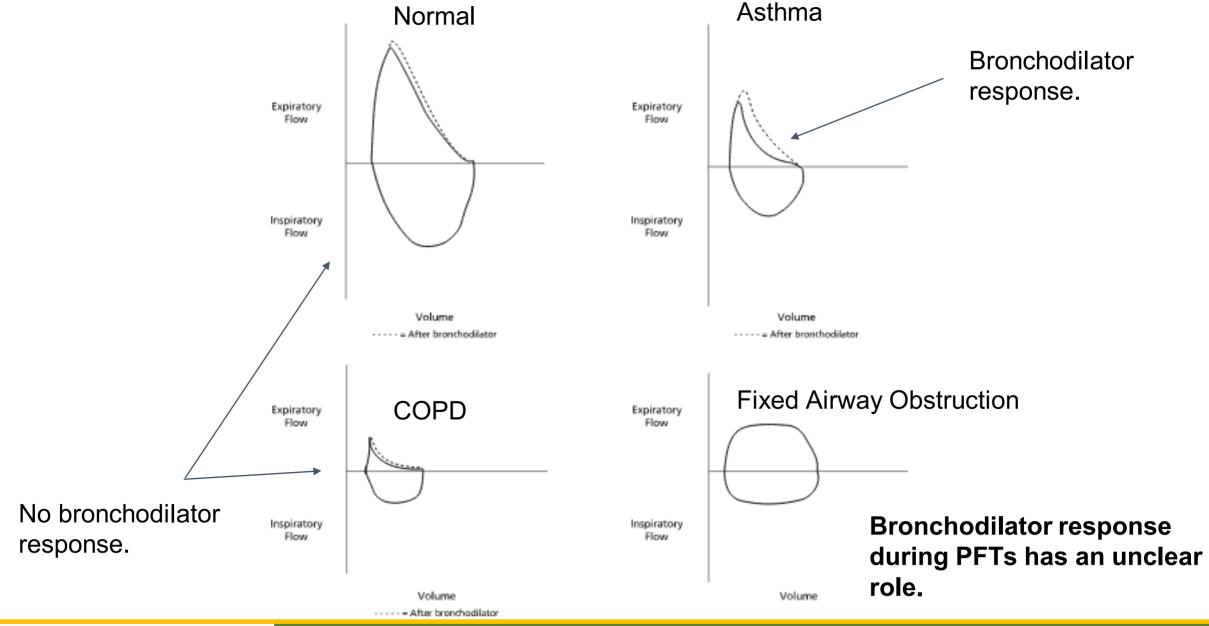
DLCO tests alveolar gas exchange. Normal in chest wall restriction, neuromuscular disease, reduced in alveolar and pulmonary vascular disease.



Methacholine / cold air / exercise challenge testing.

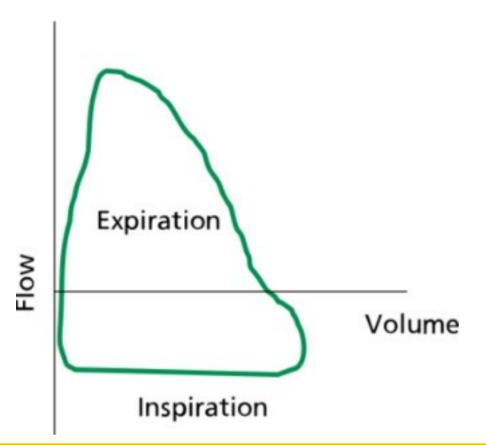
- Used to detect bronchospasm/asthma in a patient with normal baseline PFTs.
- FEV1 should fall by ~20%.



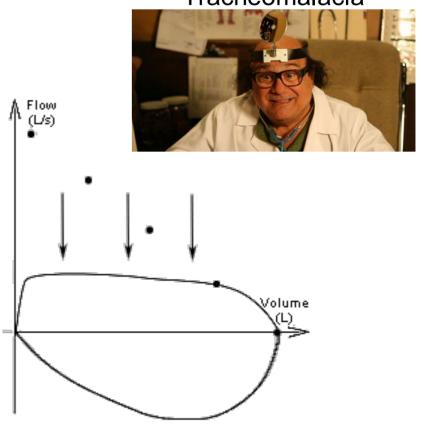




Variable Extrathoracic Obstruction [VETO] Vocal Cord Dysfunction



Variable Intrathoracic Obstruction [VITO] Tracheomalacia

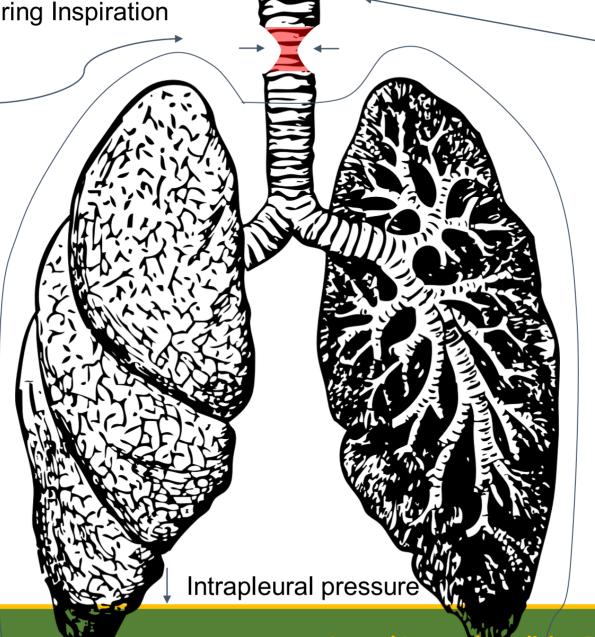


VETO: Collapse Occurs During Inspiration

Extrathoracic tracheal collapse from the environmental - tracheal luminal pressure differential.

Pleura (intrathoracic cavity)

Thoracic volume



Extrathoracic space (environment)

Tracheal luminal pressure

Diaphragm motion

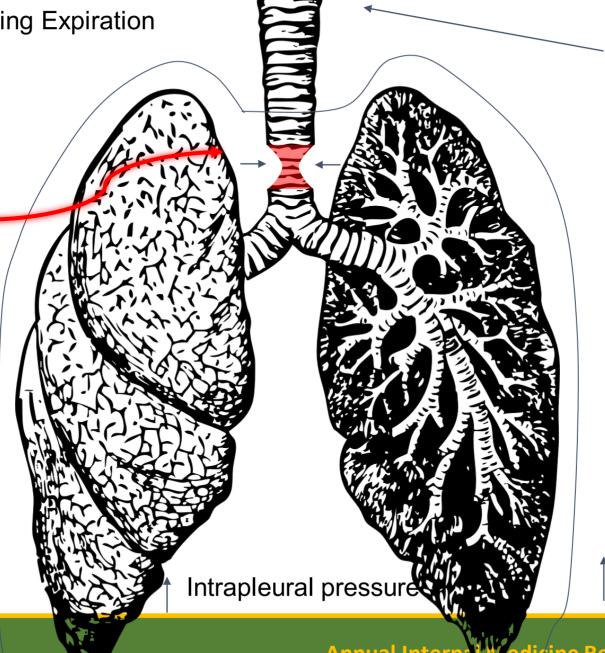


VITO: Collapse Occurs During Expiration

Intrathoracic tracheal collapse from the pleural - tracheal luminal pressure differential.

Pleura (intrathoracic cavity)

Thoracic volume



Extrathoracic space (environment)

Diaphragm motion





Pulse oximetry. Dependent on adequate digital perfusion.

Doesn't detect carbon monoxide poisoning / methemoglobinemia / etc.

Indications for home oxygen:

- a) Saturation < 89% at rest.
- b) Saturation < 90% at rest with pHTN, right CHF, or polycythemia.
- c) Saturation < 88% on exertion (weak evidence).





Six-Minute Walk Test

Not a diagnostic test, just a measure of exercise capacity.

Fractional Excretion of Nitric Oxide [FeNO]

- Correlates with the degree of eosinophilic inflammation in asthmatics.
- Correlates with glucocorticoid responsiveness.
- Unclear clinical role.



Imaging

CXR

Ultrasound - Effusions, pneumothorax, consolidations.

CTA Chest - Pulmonary Vessels

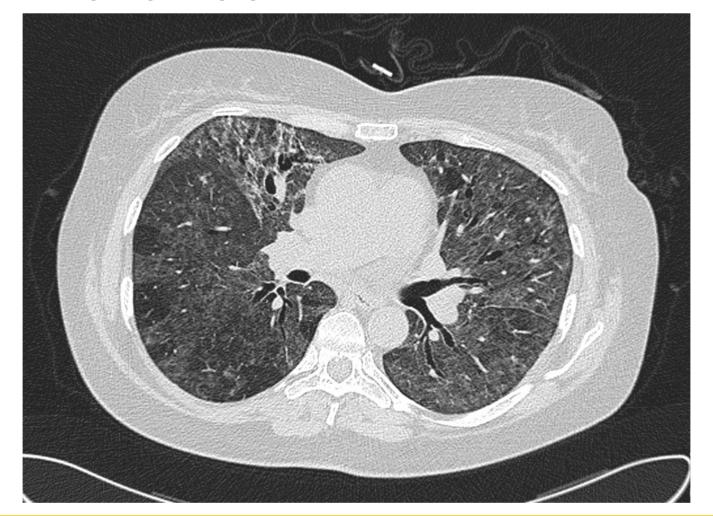
• IV contrast better defines nodules, masses, abscesses, vasculature.

PET/CT - Metabolic Activity measured by FDG [flurodeoxyglucose] uptake.

Bronchoscopy (Brushing, BAL, Biopsy, EBUS, Navigational)



EBUS and Biopsy Approach

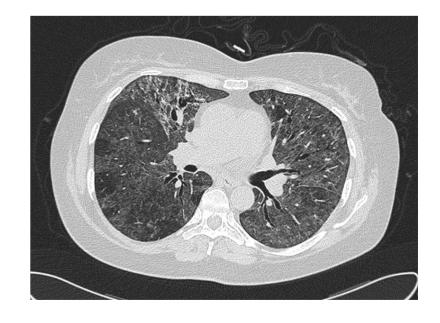


CT Scan Slice Thickness and Interval



Slice Thickness

Slice Interval



Axial Slice

Coronal Plane

Imaging - CT scans

Low-Dose CT [Screening: 50-80 years old, 20 pack-years, have smoked within the past 15 years]

- Normal Resolution, 2mm slices. Small slice interval.
- Sensitive for pulmonary nodules (not parenchyma).

Routine CT Chest

- Normal Resolution, 2mm slices. Small slice interval.
- Sensitive for pulmonary nodules (not parenchyma).

High Resolution CT Chest

- 1mm slices. <u>Large slice interval.</u>
- Sensitive for pulmonary parenchymal disease (<u>not nodules</u>).



Airway Disease Chapter 2

A 50-year-old man is referred for poorly controlled asthma. Triggers include exercise and exposure to dust, pollen, and fumes. He has allergic rhinitis. He has been treated with several courses of glucocorticoids, but symptoms recurred after he stopped treatment despite regular use of his fluticasone-salmeterol and tiotropium inhalers. His only other medication is albuterol. He has good inhaler technique.

On physical examination, vital signs are normal. BMI is 23. Pulmonary examination reveals few expiratory wheezes. The remainder of the examination is unremarkable.

Laboratory studies reveal a normal total IgE level and complete blood count.

Chest radiograph is normal. Spirometry demonstrates moderate airflow obstruction that improves with bronchodilators.

Which of the following is the most appropriate diagnostic test to perform next?

- A Absolute blood eosinophil count
- B α_1 -Antitrypsin level
- C Aspergillus-specific IgE level
- D Measurement of common allergen-specific IgE levels



Airway Disease - Asthma

Mast cell degranulation vs viral/bacterial/chemical mediated inflammation.

Can have a second phase (8 hours following first exacerbation).

Chronic asthma causes pulmonary remodeling (ie. fibrosis) over time.

Variants: Exercise, Cough, Occupational [distinguished from HSP, which has infiltrates on CXR]

Suggestive features: Sputum eosinophils, IgE Ab, increased exhaled NO.

DDx: Vocal cord dysfunction ("difficulty during inspiration"), ABPA (infectious ILD, dx with aspergillus serologies), CF, Churg-Strauss



ASA-Sensitive Asthma (AERD)

ASA-sensitivity
Severe persistent asthma
Nasal polyps (eosinophilic sinusitis)

*Can do desensitization if ASA needed.



"Reactive Airway Disease"

Describes short-lived symptoms.

Contrast to **reactive airway dysfunction syndrome**: Formal diagnosis requires 3 months of symptoms after an inciting irritant.

Consider managing other asthma associations: Sinus Disease, OSA, Obesity



Chronic Therapy

Pregnancy safe:

Prednisone/ICS/

SABA/LABA/LTRA

Intermittent

Persistent asthma in persons age ≥12 years

Consult with asthma specialist if step 4 care or higher is needed

Step 4

Preferred:

Daily and

as-needed

combination

medium-dose

ICS-formateral

Alternative:

Daily

medium-dose

ICS-LABA

ICS-LAMA.

and SABA

as needed

Step 2
Preferred:
Daily low-dose
ICS and SABA
as needed
OR
Concomitant
ICS and SABA
as needed
SABA as needed
as needed

Alternative:

Daily LTRA +

SABA as needed

Step 3

Preferred: Daily and as-needed combination low-dose ICS-formoterol

Alternative:
Daily
medium-dose
ICS + SABA
as needed
OR
Daily low-dose
ICS-LABA
or ICS-LAMA,
and SABA

as needed

Step 5

Preferred:
Daily mediumto high-dose
ICS-LABA + LAMA
and SABA
as needed

Daily medium- to high-dose ICS-LABA and SABA as needed

Alternative:

Consider add-on anti-IgE, anti-IL-5, anti-IL-4/IL13 Step 6

Preferred:
Daily high-dose
ICS-LABA +
oral systemic
glucocorticoid +
SABA as needed

Consider add-on anti-IgE, anti-IL-5, anti-IL-4/IL13

Step up if needed

(first check adherence, inhaler technique, environmental factors, and comorbidities)

> Assess control

Step down if possible

(and asthma is well controlled x 3 months)

1

Ensure good inhaler technique, compliance, consider a spacer.

* Ipratropium / tiotropium not well studied.

* Bronchial Thermoplasty

Each step: Patient education, environmental control, and management of comorbidities

For steps 2-4: Consider subcutaneous allergen immunotherapy for patients whose asthma is controlled at initiation, buildup, and maintenance phases

Quick relief for all patients:

- . SABA as needed for symptoms: up to 3 treatments at 20-minute intervals as needed
- In steps 3 and 4, use of ICS-formoterol 1-2 puffs as needed up to a maximum total daily dosage of 12 puffs (54 µg)
- . Use of SABA >2 days a week (not prevention of EIB) indicates inadequate control and the need to step up therapy



A 27-year-old woman is evaluated for a cough and chest tightness that occur during and after exercise. She has been training for her first marathon, but she has been unable to increase her training intensity because of these symptoms. She denies cough or chest tightness at any other time. She reports no stridor, throat tightness, or noisy inspiration during the episodes.

On physical examination, vital signs and pulmonary examination are normal.

Baseline spirometry is normal. Exercise testing demonstrates a significant decrease in FEV₁ from baseline.

Which of the following inhaled medications is the most appropriate next step in treatment?

- A Salmeterol daily
- B Budesonide twice daily
- C Budesonide-formoterol before exercise
- D | Ipratropium before exercise

GINA Guidelines for Starting Treatment in Adults and Adolescents with a Diagnosis of Asthma

	Step 1	Step 2	Step 3	Step 4	Step 5
Track 1	Symptoms <4-5 d/wk	Symptoms <4-5 d/wk	Symptoms most days or waking with asthma ≥ once/wk	Daily symptoms, or waking with asthma ≥ once/wk, and low lung function	
Controller regimen	As-needed low-dose ICS-formoterol		Low-dose maintenance ICS- formoterol	Medium-dose maintenance ICS- formoterol	Add on LAMA
					Assess phenotype
					Consider high-dose maintenance ICS-formotero +/- anti-IgE, anti-IL5/5R, anti-IL4R, anti-TSLP
Reliever (preferred)	As-needed low-dose ICS-formoterol				
Track 2	Symptoms < twice/mo	Symptoms ≥ twice/mo but <4-5 d/wk	Symptoms most days or waking with asthma ≥ once/wk	Daily symptoms, or waking with asthma ≥ once/wk, and low lung function	
Controller	Take ICS whenever	Low-dose maintenance	Low-dose maintenance ICS-	Medium/high-dose maintenance ICS-	Add on LAMA
regimen	SABA taken	ICS	LABA	LABA	Assess phenotype
					Consider high-dose maintenance ICS-LABA +/-
					anti-IgE, anti-IL5/5R, anti-IL4R, anti-TSLP
Reliever (alternative)	As-needed SABA				

Anti-TSLP = thymic stromal lymphopoietin; ICS = inhaled corticosteroid (glucocorticoid); LABA = long-acting β2-agonist; LAMA = long-acting muscarinic antagonist; SABA = short-acting β2-agonist.

Data from Global Initiative for Asthma. Global Strategy for Asthma Management and Prevention. Updated 2022. https://ginasthma.org/wp-content/uploads/2022/07/GINA-Main-Report-2022-FINAL-22-07-01-WMS.pdf.



Antileukotrienes

Some patients respond really well; trial these drugs early.

Particularly effective for AERD.



Biologics

Omalizumab (Anti-IgE)	Subq	Anaphylaxis, increased risk of malignancy
Mepolizumab Benralizumab (Anti-IL5)	Subq	EOSINOPHILS
Reslizumab (Anti-IL5)	IV Infusion	EOSINOPHILS
Dupilumab (Anti-IL4/IL13)	Subq	EOSINOPHILS



Omalizumab takes months to work.



Acute Exacerbation

Check the Peak Flow (< 50% of baseline is concerning)

A 25-year-old man is seen in follow-up examination for asthma diagnosed 2 months ago. He reports that his symptoms are now well controlled without use of his rescue inhaler, and results of the Asthma Control Test confirm well-controlled asthma. During the visit, he describes feeling down on many days, with difficulty falling asleep and early morning awakening. Depression screening with the Patient Health Questionnaire-2 is positive. Medications are beclomethasone, montelukast, and albuterol.

On physical examination, vital signs and pulmonary examination are normal.

Which of the following is the most appropriate treatment?

- A Begin escitalopram
- B Begin salmeterol
- C Stop beclomethasone; begin budesonide-formoterol
- D Stop montelukast



COPD

Cough.

Sputum.

Dyspnea.

Causes to look for:

- Smoking (duh)
- Biomass fuel exposure.
- Genetics (CF, AAT)
- Pollution
- Childhood asthma or pulmonary infections.

Don't screen for it - only test symptomatic patients.



COPD Therapy

(similar to asthma)

*Don't forget pulmonary rehab.

SABA
LAMA or LABA
LAMA and LABA
LAMA / LABA / ICS [Most ICS benefit in patients with eosinophilia]
LAMA / LABA / ICS / Roflumilast (frequent / chronic bronchitis only)

Consider NAC or Azithro here (controversial).

* LABA monotherapy is fine, unlike in asthma. Don't use it as monotherapy in overlap syndromes.



Theophylline for either COPD or Asthma

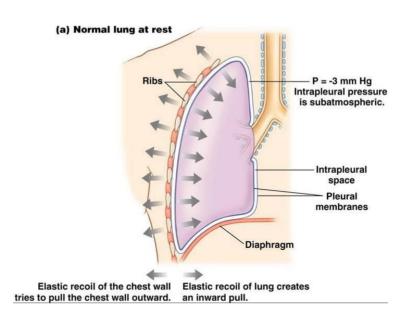
Toxic, but effective if you don't have any other good options.



COPD Therapy

End-Stage Adjuncts

- Home BiPAP
- Lung Volume Reduction Surgery (heterogeneous emphysema [upper lobe], FEV 20-45%)
 (Remove nonfunctioning bullae to increase elastic recoil leading to widened bronchioles and less expiratory air trapping)
- Bronchoscopic therapy is an option.
- Lung Transplant (homogenous emphysema, FEV < 20%)



¡COPD Exacerbación!

Oxygenation to a pulse ox goal of 90% (hyperoxygenation can precipitate VQ mismatch and lead to CO2 narcosis).

Albuterol/Ipratropium

Prednisone 40-80mg daily x 5 days

Use antibiotics for moderate-severe exacerbations OR infectious symptoms (change / increase in sputum, fever). **No comparative evidence between antibiotics.**

- Targets: Haemophilus influenzae, Moraxella catarrhalis, Streptococcus pneumoniae
- Lower risk: Azithromycin vs Cefpodoxime
- Higher risk: Amoxicillin-clavulanate vs Fluoroquinolone



Bronchiectasis

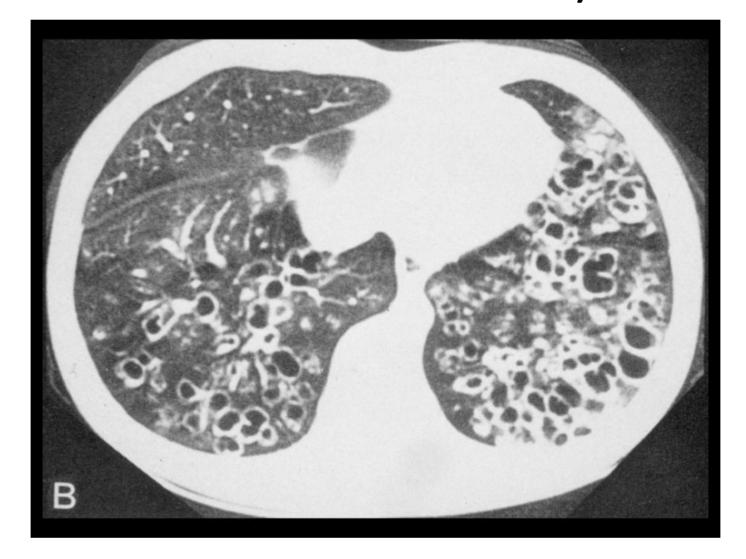
Characterized by chronic, productive cough.

Lots of causes. Look them up. Then forget them, because half the cases are idiopathic.

Diagnosis: Clinical + high-resolution CT scan.



Cystic Fibrosis - Mean survival of 41 years.



Cystic Fibrosis

CFTR mutation makes mucus secretions thicker.

Mucus is found in the ...

Sinuses: Sinusitis, Nasal polyps

Lungs: Bronchiectasis with pulmonary infections (ABPA, Burkholderia, Pseudomonas).

Pancreas: Pancreatitis, Diabetes

Vas deferens: Male infertility

Also, osteoporosis and liver disease.



Cystic Fibrosis

Sweat chloride testing is not sensitive.



Bronchiectasis/CF Treatment

Chronic:

Pulmonary Rehab
Inhaled Hypertonic Saline
Chest PT, Flutter Valves
Inhaled Tobramycin
Oral Azithromycin (rule out mycobacterial infection first)
Decolonization of staph or pseudomonas from the lung.

Inhaled DNAse (Dornase alfa) - Works in CF. Inhaled NAC - Unclear benefit.

SABA, ICS, and LABAs help with symptoms; no impact on outcomes unless patients also have COPD.

Bronchiectasis and CF flares: Empiric antibiotics (ie. fluroquinolones).

CF Flares with bronchospasm: Systemic steroids (controversial).



Lung Cancer Chapter 7

Pulmonary Nodules

The bigger, the badder.

> 3cm = Cancer

Spiculated / lobulated borders are scary.



SOLID NODULES GROW FAST

Risk Factors for Lung Cancer?	Size	Recommended Follow-Up
No (Low-risk patient)	<6 mm	No follow-up
	6-8 mm	CT at 6-12 months then consider CT at 18-24 months
	>8 mm	Consider CT at 3 months, PET/CT, or tissue sampling
Yes (High-risk patient)	<6 mm	Optional CT at 12 months
	6-8 mm	CT at 6-12 months then CT at 18-24 months
	>8 mm	Consider CT at 3 months, PET/CT, or tissue sampling



GROUND-GLASS NODULES GROW SLOW

Imaging Findings	Size	Recommended Follow-Up
Pure ground glass	<6 mm	No follow-up
	≥6 mm	CT at 6-12 months to confirm persistence, then CT every 2 years until 5 years
Part solid nodule	<6 mm	No follow-up
	≥6 mm	CT at 3-6 months to confirm persistence. If unchanged and solid component remains <6 mm, annual CT should be performed for 5 years



Lung Cancer

Adeno, Large Cell - Peripheral (Newer treatments targeting EGFR / ALK / ROS1)

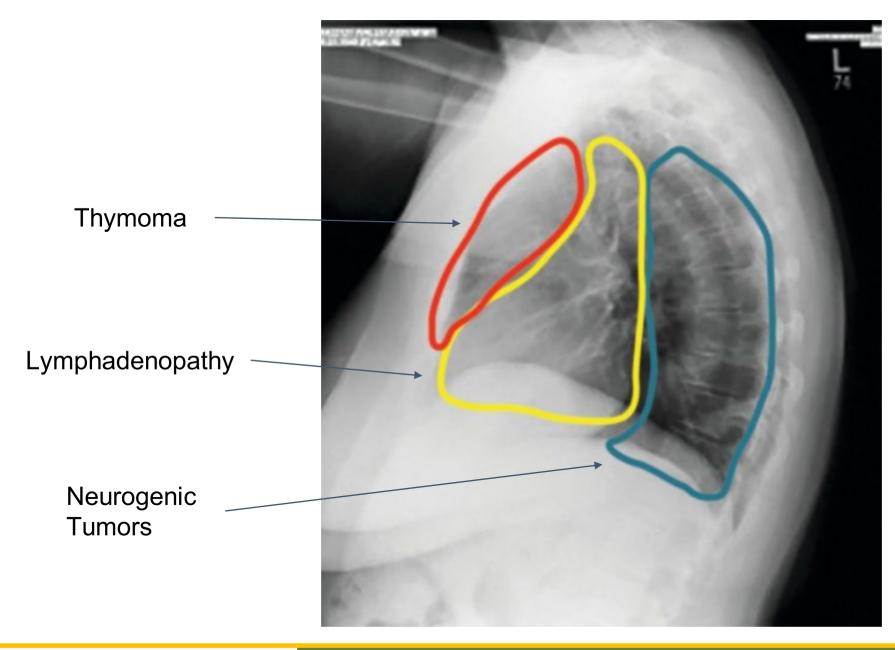
Squamous - Central, Smokers

Small Cell - Poor prognosis, widely metastatic at diagnosis. Smokers.

Predilection for mediastinum and paraneoplastic syndromes.

Screening: Low-dose CT annually. 55 to 80, 30-pack-years, have smoked within the last 15 years.





A 62-year-old man is evaluated in follow-up examination for COPD. Despite smoking cessation, adherence to his medical regimen, good inhaler technique, and participation in pulmonary rehabilitation, he continues to experience breathlessness with mild exertion and has diminished quality of life. He has a minimal dry cough, and he has never required treatment for an acute exacerbation of COPD. Medications are fluticasone-umeclidinium-vilanterol and albuterol inhalers as needed. Immunizations are up to date.

On physical examination, vital signs are normal. Oxygen saturation \triangle is 93% with the patient breathing ambient air. There are diminished breath sounds

A 6-minute walk test shows a minimum oxygen saturation \triangle of 90% with the patient breathing ambient air. Spirometry shows an FEV₁ \triangle of 35% of predicted and a DLCO of 42% of predicted.

Chest imaging shows upper-lobe-predominant emphysema.

Which of the following is the most appropriate treatment? A Long-term azithromycin therapy B Lung volume reduction surgery C Roflumilast D Supplemental oxygen

A 27-year-old woman is evaluated for a cough and chest tightness that occur during and after exercise. She has been training for her first marathon, but she has been unable to increase her training intensity because of these symptoms. She denies cough or chest tightness at any other time. She reports no stridor, throat tightness, or noisy inspiration during the episodes.

On physical examination, vital signs and pulmonary examination are normal.

Baseline spirometry is normal. Exercise testing demonstrates a significant decrease in FEV₁ from baseline.

Which of the following inhaled medications is the most appropriate next step in treatment?

- A Salmeterol daily
- B Budesonide twice daily
- C Budesonide-formoterol before exercise
- D | Ipratropium before exercise

A 57-year-old woman is evaluated at a follow-up visit after discharge from the hospital for a COPD exacerbation. This was her third hospitalization for a COPD exacerbation in the past 6 months. She no longer smokes cigarettes. Medications are fluticasone-umeclidinium-vilanterol and albuterol inhalers. She has been adherent with her medication, has excellent inhaler technique, and continues to use home oxygen. Between exacerbations she has a cough productive of thin, colorless sputum.

On physical examination, vital signs are normal. Oxygen saturation is 93% breathing oxygen, 2 L/min by nasal cannula. Scattered expiratory wheezing is heard. Cardiac examination is normal.

Spirometry shows postbronchodilator reduced FEV₁/FVC ratio and an FEV₁ \triangle of 45% of predicted.

Oral N-acetylcysteine

Influenza, COVID-19, and pneumococcal pneumonia vaccinations are brought up to date. The patient is enrolled in a pulmonary rehabilitation program. α_1 -Antitrypsin level is normal.

Which of the following additional long-term treatments is most appropriate?

A \(\alpha_1\text{-Antitrypsin augmentation therapy} \)

B \(\text{Azithromycin} \)

C \(\text{Metoprolol} \)

A 72-year-old woman is evaluated for exacerbation of bronchiectasis symptoms over the past 3 days. Chronic productive cough has worsened in frequency and severity, and her sputum has increased in amount, become thicker, and changed in color from white to dark yellow. She has been using albuterol and hypertonic saline nebulization with a positive expiratory pressure device three times daily since her symptoms increased.

On physical examination, vital signs are normal. Lung examination reveals bibasilar crackles. There is no accessory muscle use.

Prior sputum cultures have grown Pseudomonas aeruginosa and Haemophilus parainfluenzae.

Chest radiograph reveals chronic interstitial markings but no acute change.

Which of the following is the most appropriate treatment? A Azithromycin B Ciprofloxacin C Inhaled dornase alfa D Prednisone

Interstitial Lung Disease



Common Drug Toxicities:

Amiodarone

Methotrexate

Nitrofurantoin (generally long term)

Chemotherapy (ie. bleomycin)

Chest radiation (4-12 weeks after radiation; generally treated with steroids; subsequent fibrosis in 6 months)

Smoking (especially RBILD [centrilobular micronodules], DIP, Langerhans cell histiocytosis, SRIF [smoking-related interstitial fibrosis [3]])

Chronic Aspiration

Pneumoconiosis

Autoimmune disease (Scleroderma, RA, Lupus, DM/Polymyositis [Anti-synthetase syndrome], etc.)

- Examine the skin & joints.

Hypersensitivity Pneumonitis (poorly formed granulomas)



When to suspect ILD:

- Cough
- Shortness of breath
- Atypical presentation
- Treatment unresponsive
- PNEUMONIA

How to work it up:

Start with spirometry, follow through with pulm consult and high res CT.

Normal CXR and spirometry do not exclude early ILD.

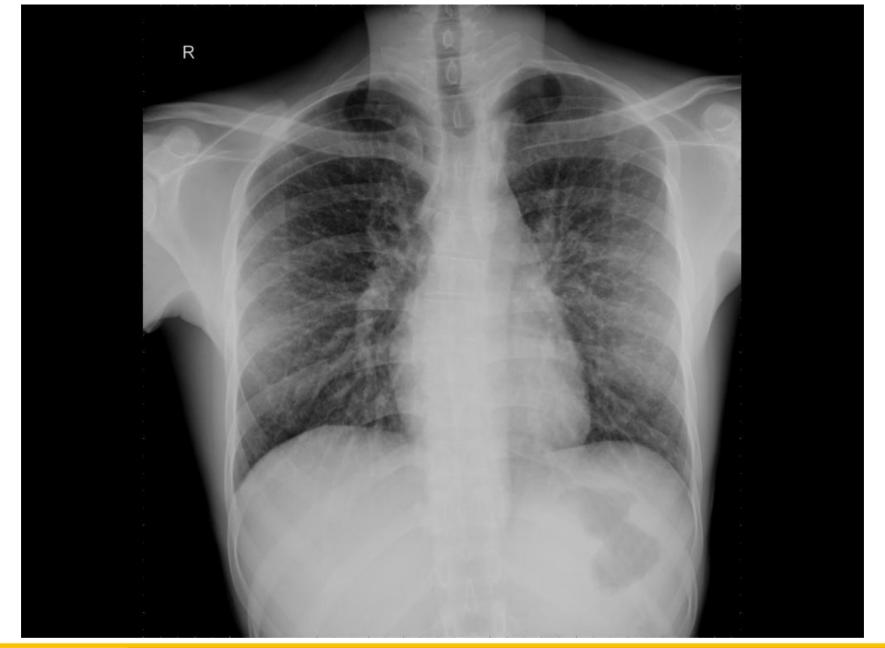
Final Diagnosis:

VATS with lung biopsy. Transbronchial biopsy yield is low.



RETICULAR INFILTRATES (intersecting lines)

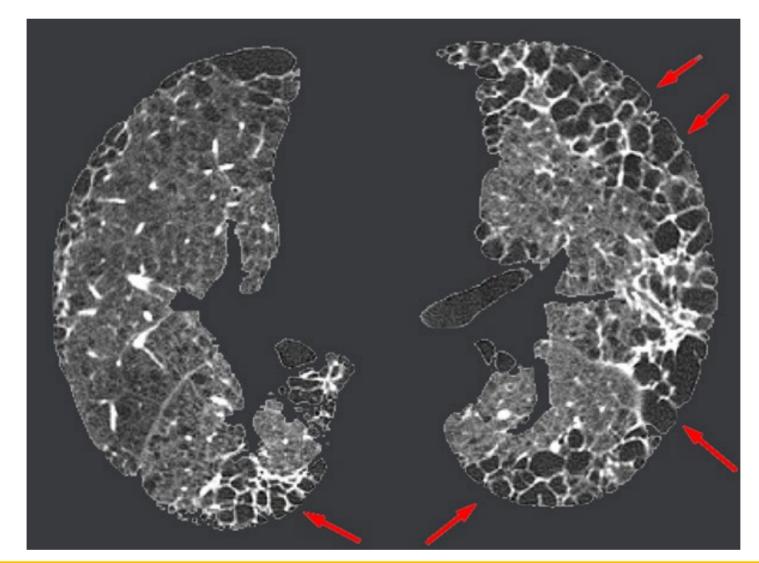




TRACTION BRONCHIECTASIS

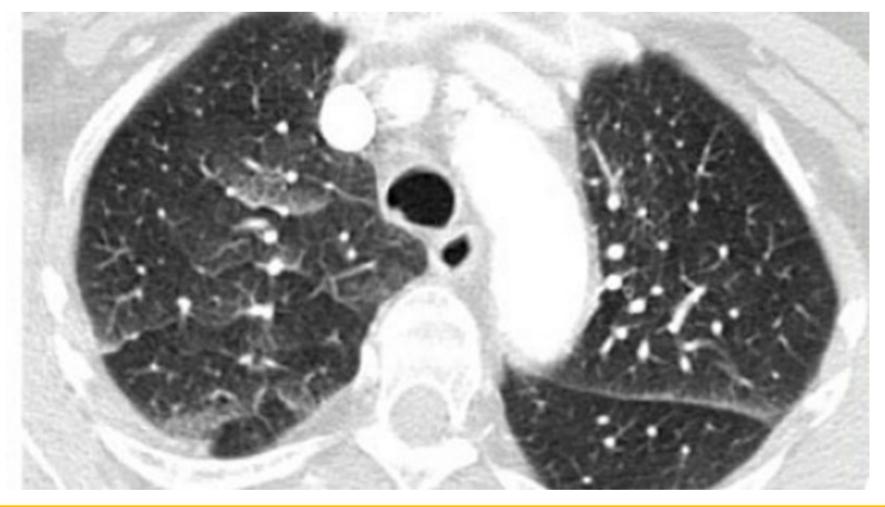


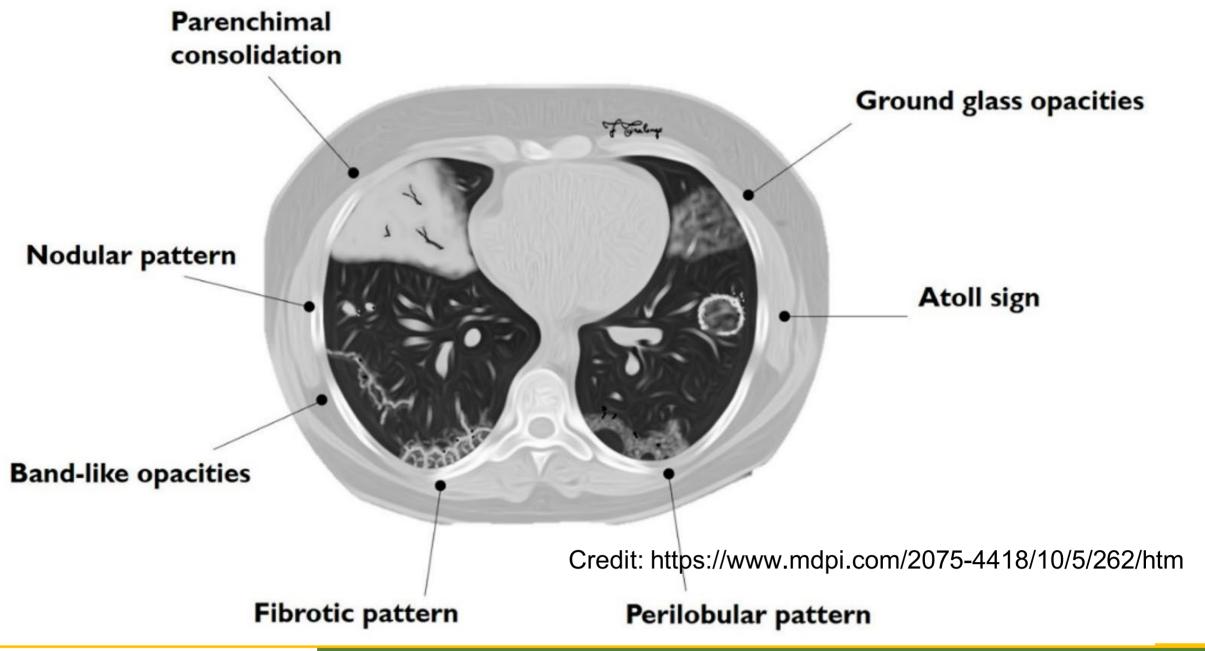
HONEYCOMBING





GROUNDGLASSING



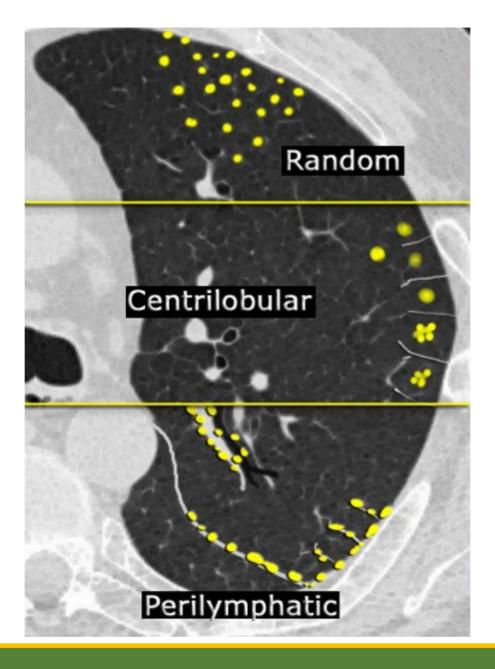




SEPTAL LINE THICKENING

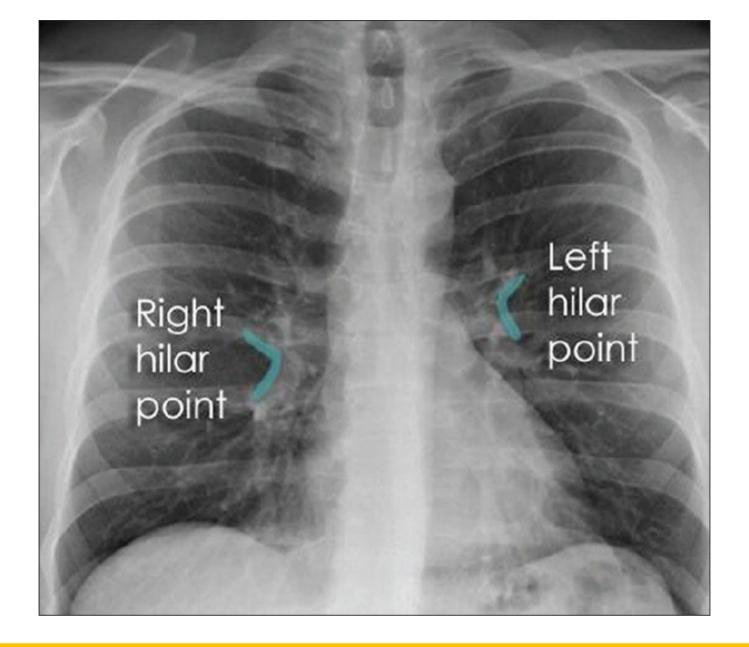


NODULAR DISTRIBUTION

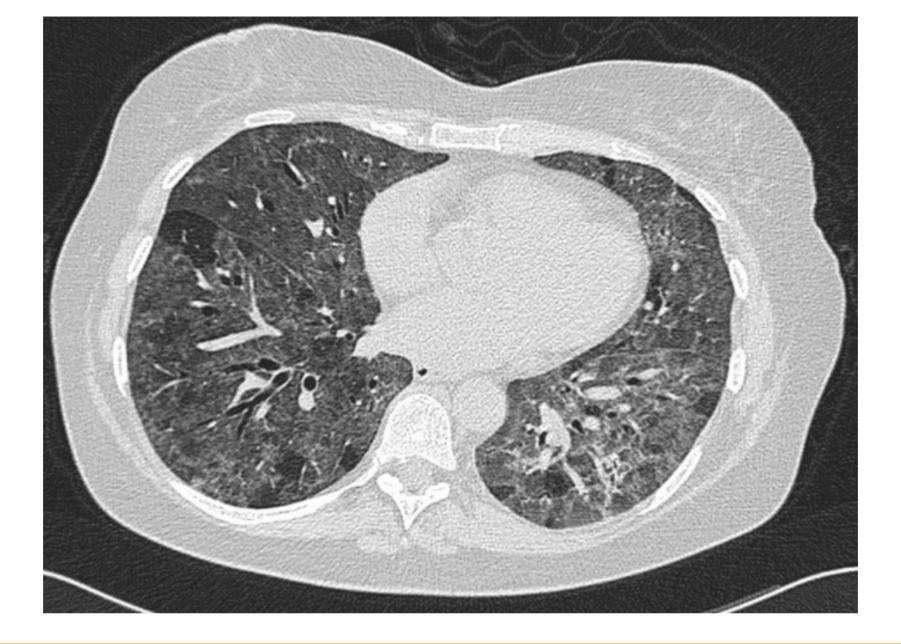


Hilar Adenopathy



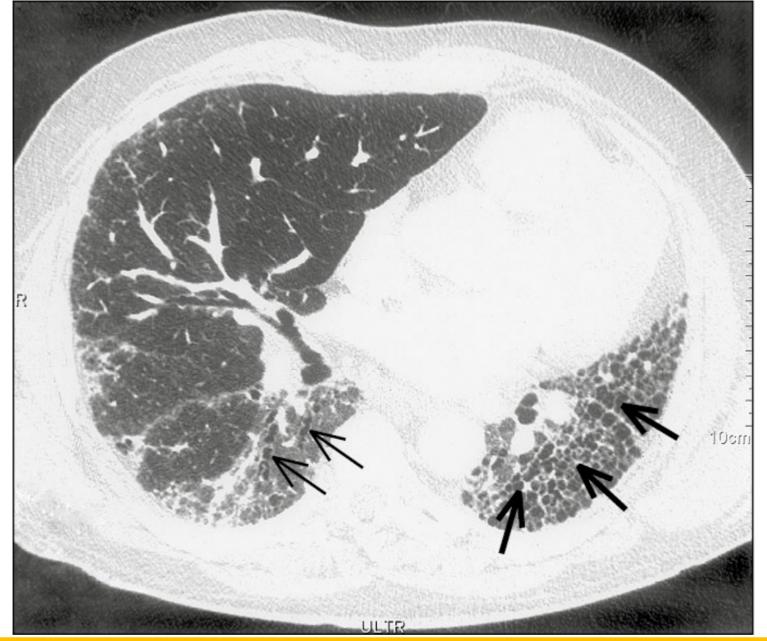






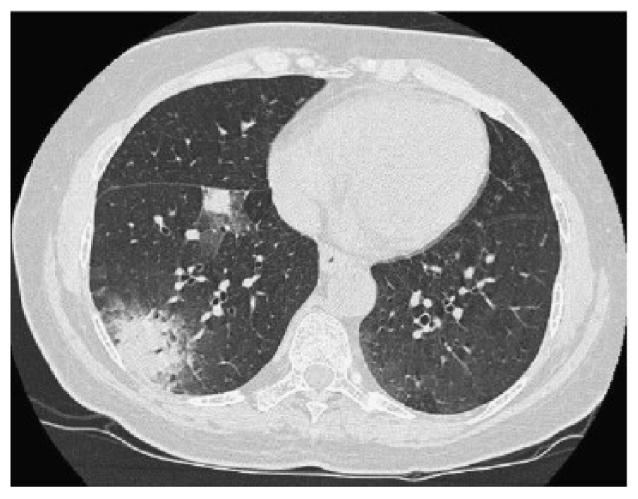
Hypersensitivity Pneumonitis

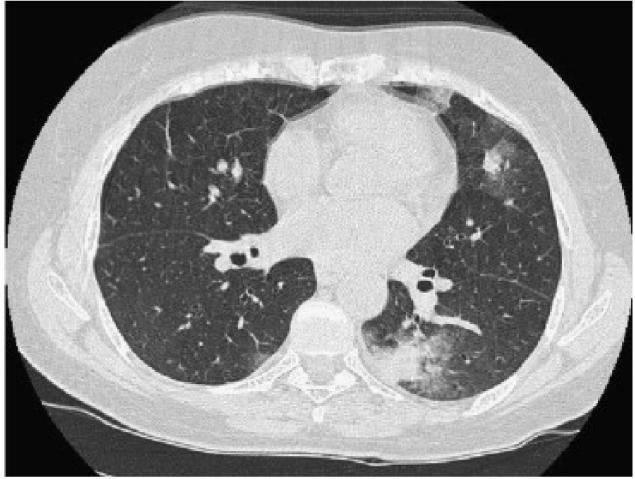




IPF







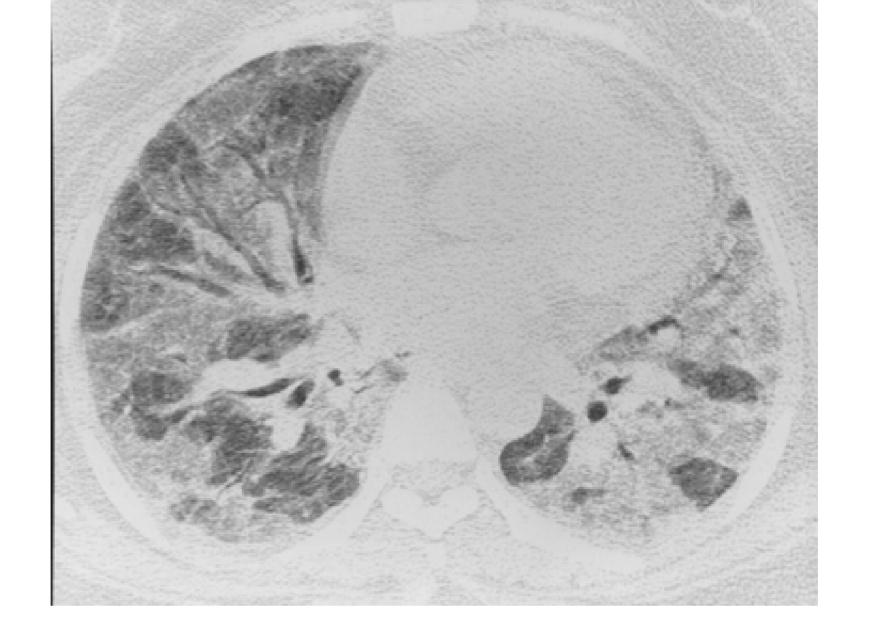
COP / BOOP





Sarcoidosis





AIP



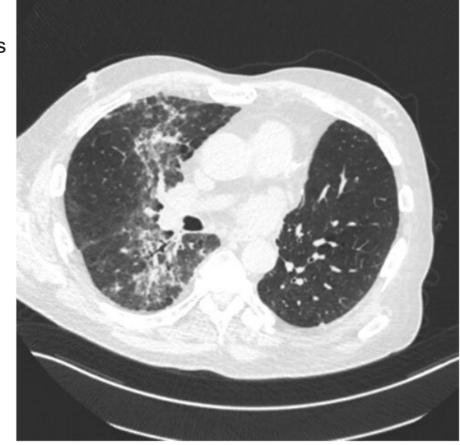
A 55-year-old man is evaluated in the hospital for fever, dry cough, and shortness of breath of 3 days' duration. He is a horrific historian; after a 30 minute interview you now know he has a history of lung cancer and have a deep

understanding of his political views.

On physical examination, temperature is 37.8 °C (100.0 °F), blood pressure is 110/70 mm Hg, pulse rate is 102/min, and respiration rate is 22/min. Oxygen saturation is 92% with the patient breathing ambient air. BMI is 17. Crackles are present in the right infrascapular region. Cardiovascular examination is normal.

The radiology resident is currently in the emergency room having a STEMI.

- A) Bacterial pneumonia
- B) Radiation pneumonitis
- C) Acute interstitial pneumonia
- D) Cryptogenic organizing pneumonia
- E) Bleomycin toxicity



Hypersensitivity Pneumonitis - focus on the history

Actinomyces, fungi, bird droppings are most common (but could be any antigen exposure)

Acute HP looks like ATYPICAL PNEUMONIA

Symptoms improve 1-2 days after removal from exposure.

Chronic HP (ie. Bird fancier's) leads to fibrosis / irreversible ILD.

Steroids for severe disease.

HRCT findings (please don't memorize them):

Upper/mid lung zones (pseudoscience: less antigen reaches lower lung zones)

Centrilobular micronodules.

Groundglassing



A 66-year-old man is evaluated for gradually progressive dyspnea and dry cough over the past 6 months. He has a 30-pack-year history of smoking; he stopped 10 years ago. Medical history is otherwise unremarkable.

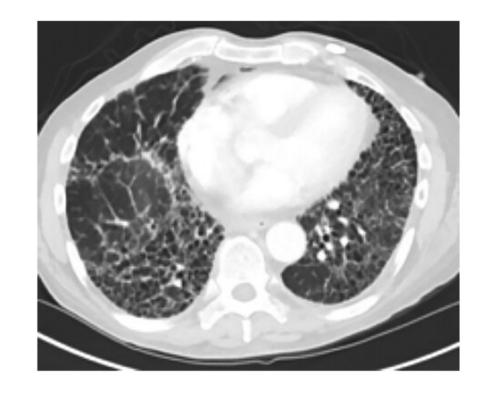
On physical examination, respiration rate is 22/min. Oxygen saturation \triangle is 93% with the patient breathing ambient air. Auscultation of the lungs reveals fine end-inspiratory Velcro-like crackles at both lung bases. Clubbing is present.

Spirometry shows an FVC \triangle of 55% of predicted, an FEV₁/FVC \triangle ratio of 0.91, and a DLCO of 42% of predicted.

A high-resolution CT scan of the chest is shown.

Which of the following is the most likely diagnosis?

- A Cryptogenic organizing pneumonia
- B Idiopathic pulmonary fibrosis
- C Nonspecific interstitial pneumonia
- D Respiratory bronchiolitis-associated interstitial lung disease





Idiopathic Pulmonary Fibrosis (Idiopathic UIP)

Patients are 50+ years old.

Poor prognosis (3-5 years).

Supportive care only for exacerbations (steroids don't work).

Long-term treatments:

- Nintedanib (minimal benefit, if any)
- Pirfenidone (minimal benefit, if any)
- Transplant

HRCT findings (no need to memorize):

- Lower lung zones
- Septal thickening
- Honeycombing
- Traction bronchiectasis



A 67-year-old man is evaluated for progressive shortness of breath and dry cough over the past 3 months. He has a 40-pack-year history of cigarette smoking. His only medication is an albuterol inhaler.

On physical examination, pulse rate is 90/min and respiration rate is 24/min. Oxygen saturation \triangle is 88% with the patient breathing ambient air. Auscultation demonstrates bibasilar crackles. Clubbing is present.

Spirometry shows an FVC \triangle of 110% of predicted, an FEV₁ \triangle of 95% of predicted, an FEV₁/FVC \triangle ratio of 0.68, and a DLCO of 36% of predicted.

An echocardiogram shows normal left ventricle ejection fraction. Right ventricle systolic pressure is 66 mm Hg.

Representative high-resolution CT chest images are shown.

- A Acute interstitial pneumonia
- B Combined pulmonary fibrosis and emphysema
- C Desquamative interstitial pneumonia
- D Idiopathic pulmonary fibrosis



Combined Pulmonary Fibrosis and Emphysema

Is now a thing ©



Non-specific Interstitial Pneumonia

- Common in MCTD
- Contrast to IPF: <u>NSIP is inflammatory</u>.
- 5-year mortality of 15-25%.

Therapy

- Mild: Observation
- Moderate: Steroids (Grade IIC like everything else)
- Severe: Immunosuppressants

HRCT findings (no need to memorize):

- Lower lung zones
- Reticular infiltrates
- Groundglassing



Cryptogenic Organizing **Pneumonia**

- Unresponsive to antibiotics (duh)
- Steroid responsive.
- Associated with autoimmune disease.

HRCT findings (no need to memorize):

- Lower lung zones
- Groundglassing
- Consolidation



Sarcoidosis

- Hilar adenopathy.
- Pulmonary nodularity/fibrosis.
- Noncaseating granulomas on bronchoscopic lymph node FNA.
- Steroid responsive (use low or alternate day dosing).
- Can affect any organ system.

Don't biopsy these (because they are self limiting):

Hilar adenopathy alone.

Hilar adenopathy + polyarthralgias + E. nodosum = *Lofgren syndrome*

Hilar adenopathy + uveoparotid fever + facial nerve palsy = *Heerfordt syndrome*

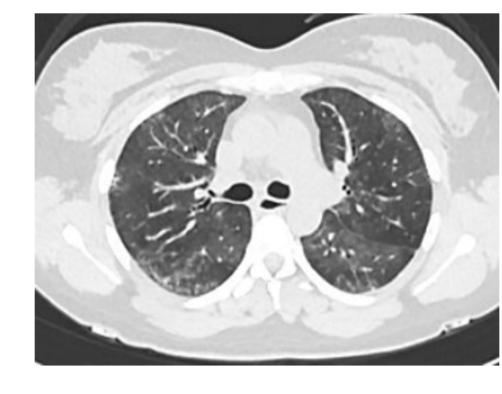
Steroids are an option for severe Lofgren's or Heerfordt's.



A 32-year-old woman is evaluated in the hospital for a 3-week history of progressive shortness of breath, arthralgia, and fatigue. Her only medication is pantoprazole for gastroesophageal reflux disease. Medical history is significant for Raynaud phenomenon treated with cold avoidance. She works in an accounting firm and has no unusual environmental exposures.

On physical examination, blood pressure is 140/90 mm Hg, pulse rate is 98/min, and respiration rate is 24/min. Oxygen saturation is 90% breathing oxygen, 2 L/min by nasal cannula. Auscultation reveals crackles at both lung bases. She has puffy-appearing fingers. The remainder of the examination is normal.

Laboratory studies:	
Hemoglobin A	11 g/dL (110 g/L)
C-reactive protein 🛆	4 mg/dL (40 mg/L)
Antinuclear antibody	Positive
Antitopoisomerase I (anti-ScI-70) antibody	Positive



Chest radiograph shows bilaterally increased markings with a basilar predominance without lymphadenopathy.

- A) RBILD
- B) Hypersensitivity pneumonitis
- C) IPF
- D) Sarcoidosis

- E) NSIP
- F) SRIF
- G) COP/BOOP
- H) ACGME/ECFMG

Acute Interstitial Pneumonia

- Looks like ARDS / pulmonary edema.
- Biopsy shows diffuse alveolar damage.
- Steroid responsive.
- High mortality.



ARDS/DAD Highlights

- Pulmonary HTN
- Decreased Lung Compliance (Higher Pressures for Lower Volumes)
- Hypoxia despite high FiO2 & PEEP
 - * Be conservative with fluids, target CVP ~ 4
 - * Prone positioning seems to have a mortality benefit.



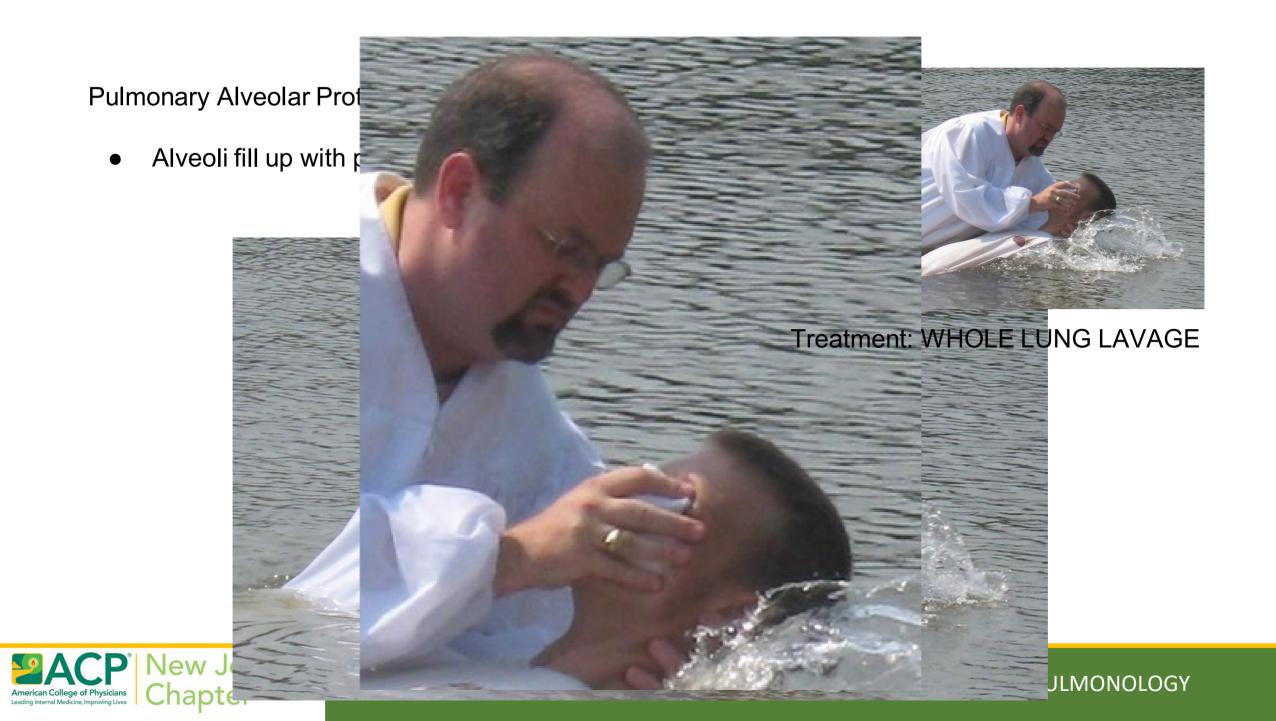
Lymphangioleiomyomatosis (smooth muscle grows into the alveoli)

- Young women.
- Pneumothorax.
- Chylous effusions.
- Cystic pulmonary changes.
- Sirolimus?



Eosinophilic Pneumonia

Eosinophils



Silicosis

- Upper lobe predominant (pseudoscience: dust doesn't go too deep)
- Increased risk for TB and lung cancer.
- No treatment.



A 25-year-old man is evaluated in the emergency department for worsening shortness of breath and right-side pleuritic chest pain, which developed 1 hour ago. He has an 8-pack-year history of smoking cigarettes. His medical history is otherwise unremarkable, including the absence of lung disease. He is a professional scuba diver.

On physical examination, blood pressure is 150/70 mm Hg, pulse rate is 105/min, and respiration rate is 30/min.

Oxygen saturation is 92% with the patient breathing ambient air. There are decreased breath sounds, reduced expansion, and hyperresonance to percussion on the right side.

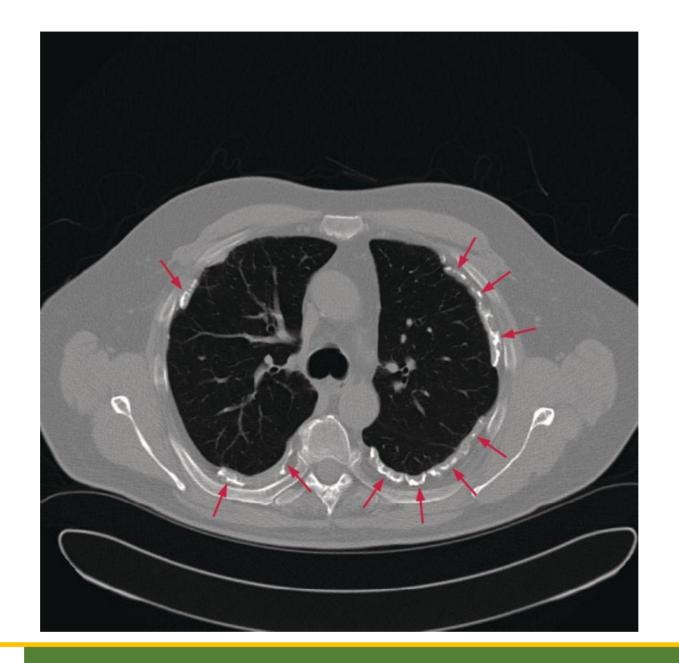
Chest radiograph reveals a large right pneumothorax and no signs of tension.

Smoking cessation counseling and an offer of varenicline are planned at the time of discharge.

Which of the following is the most appropriate additional pneumothorax management?

- A Catheter thoracostomy followed by pleurodesis
- B Needle aspiration
- C Observation
- D Supplemental oxygen and observation





Asbestosis

Exposures:

- Cars
- Ships
- Factories
- Construction
- Mining

Complications:

- ILD
- Exudative Pleural Effusions & Plaques
- Lung cancer (mesothelioma); multiplicative risk when coupled with smoking.



Spontaneous Pneumothorax - High-flow oxygen.

- Primary (healthy lungs)
 - Small (<2cm): observe
 - Large: needle decompression, chest tube if it re-expands
 - Recurrent primary pneumothorax: Pleurodesis
- Secondary (crap lungs)
 - Small (<2cm): observe
 - Large: chest tube
 - Pleurodesis when stable.
- Tension: Needle decompression immediately followed by chest tube.

No air travel until it's gone. No diving ever (unless mechanically addressed).

Stop smoking!



Pulmonary Hypertension

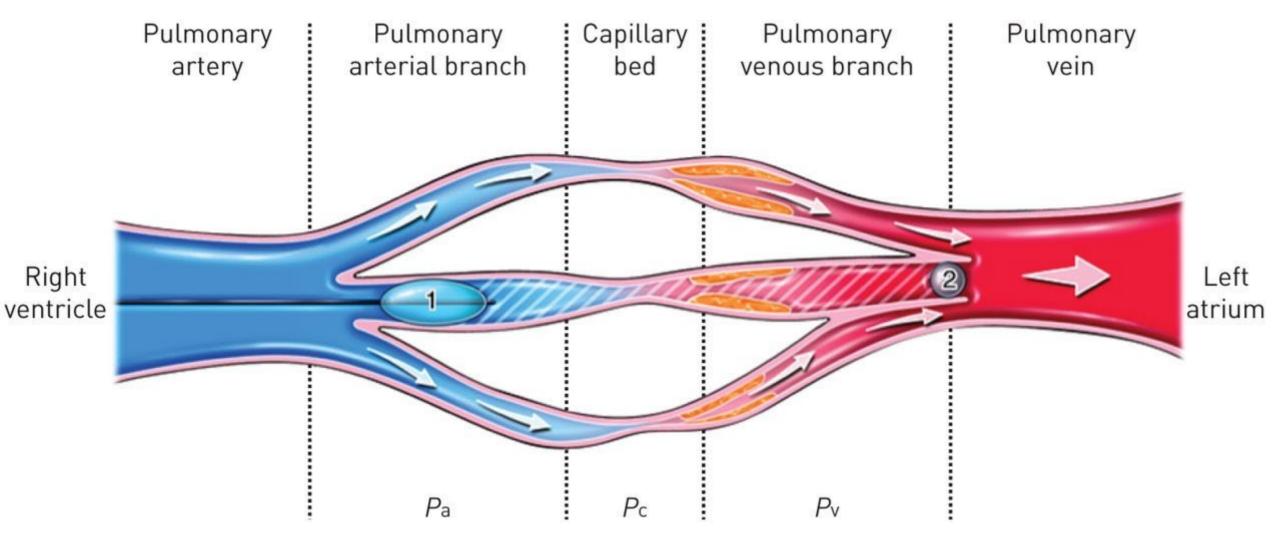
Normal RVSP is 15 - 25 mmHg



Pulmonary Hypertension PA > 25 mm Hg	Group 2 Heart Disease Systolic Left Heart Failure Diastolic Left Heart Failure Aortic Valve Disease Mitral Valve Disease	Group 3 Lung Disease / Hypoxemia COPD ILD OSA Obesity Hypoventilation Syndrome
Group 4 Chronic Thromboembolic Pulmonary Hypertension	Group 1 Pulmonary Arteriole Disease - Responds to vasodilator therapy. Idiopathic / Genetic MCTD Drug-Induced HIV Portopulmonary Hypertension Schistosomiasis (Biochemical/Mechanical) Chronic Hemolytic Anemia (Biochemical) *Pulmonary Veno-Occlusion	Group 5 Unknown Pathophysiology Hematologic Disorders Sarcoidosis Sickle Cell (Recurrent Infarction) Glycogen Storage Disease



PVOD



*Normal PCWP doesn't rule this disease out.

Credit: https://erj.ersjournals.com/content/47/5/1518



Making the Diagnosis

- 1) ECHO first (not 100% sensitive)
- 2) Right Heart Cath if ECHO negative or if you want to do vasoreactivity testing (to evaluate response to CCB treatment).

A 72-year-old man is evaluated in the emergency department for exertional dyspnea of 1 year's duration and recent onset of orthopnea. He has diabetes mellitus, hypertension, and nonischemic cardiomyopathy. Medications are atorvastatin, sacubutril-valsartan, metoprolol, and metformin.

On physical examination, blood pressure is 150/82 mm Hg; the remaining vital signs are normal. Jugular venous pressure is elevated. An S3 is present. Pulmonary examination reveals occasional crackles. There is 2+ pitting edema to the knees.

A chest radiograph shows an enlarged cardiac silhouette and pulmonary vascular congestion.

An echocardiogram reveals a global left ventricular hypokinesis with an ejection fraction of 20% and a right ventricular systolic pressure of 60 mm Hg.

Which of the following is the most appropriate next step in management?

- A) Bosentan
- B) IV Dobutamine
- C) IV Furosemide
- D) Sildenafil
- E) Right heart catheterization with vasodilator challenge



Evaluation Steps

- 1) Lung Disease: PFTs
- 2) Hypoxemia: PSG & Overnight oximetry
- 3) Thromboembolism: V/Q Scan
- 4) MCTD ANA, RF, ANCA
- 5) HIV ELISA
- 6) Portal Hypertension LFTs



A 42-year-old man with 6 months of progressive dyspnea is diagnosed with chronic thromboembolic pulmonary hypertension following right heart catheterization and pulmonary artery angiography. Medical history is otherwise unremarkable, and he takes no medications.

Apixaban is initiated.

Which of the following is the most appropriate management?

- A Add riociguat
- Balloon pulmonary angioplasty evaluation
- C Surgical thromboendarterectomy evaluation
- D No additional treatment

CTEPH after Acute PE

Data unclear regarding whether or not thrombolytics / thrombectomy reduces the ultimate risk of developing CTEPH.

CTEPH Evaluation

Part of the standard testing for pulmonary hypertension.

VQ scan has almost 100% sensitivity.

After diagnosis, plan right heart cath and surgical evaluation (pulmonary thrombendarterectomy).



CTEPH Treatment

Inoperative? Second surgical opinions are standard of care.

Still inoperative? Consider balloon angio, riociguat.

Type I Treatment

- Sildenafil / Tadalafil
 - If no response, consider prostaglandins [epoprostenol, treprostinil, iloprost, selexipag], riociguat, or bosentan [ambrisentan, macitentan].
- Nifedipine (CCBs)

Portopulmonary HTN -> HAS PHTN
Hepatopulmonary syndrome -> DOES NOT



Pleural Effusions!

Most common causes:

- CHF (Bilateral effusions)
- Pneumonia (Parapneumonic effusion)
- Malignancy



DDx

Transudate: Organ Failure States (CHF, Cirrhosis, Nephrotic Syndrome)

* Hepatic Hydrothorax, Urinothorax, Peritoneal Dialysis, Hypothyroidism, SVC Syndrome

Exudate: Everything Else.

Bacterial Infection / Tuberculosis, Malignancy, Pulmonary Embolus, Rheumatoid Arthritis, Pancreatitis, Esophageal Rupture, Chylothorax, Blah Blah (30 other causes)



Diagnostic Evaluation

Thoracentesis - Exudate if any 3 of Light's Criteria Met

Pleural / Serum Protein > 0.5

Pleural / Serum LDH > 0.6

Pleural LDH > $\frac{1}{3}$ upper limit of normal.



Diagnostic Evaluation - FYI

Pleural Effusion secondary to CHF can appear exudative after diuresis.

High pleural NT-proBNP is highly specific for CHF as the etiology.



Diagnostic Evaluation - FYI

If you were expecting a transudative effusion, but got an exudative one...

Check the serum-pleural protein gradient. High gradient (>3.1g/dL) means that it's transudative (pleural fluid shouldn't contain protein). Can also do "SAAG" with cutoff of 1.2g/dL.

Elevated Amylase - Malignancy, Pancreas, Esophagus

Low Glucose - Malignancy, Bacterial Infection, Rheumatoid Arthritis

Elevated ADA, AFB Stain/Cultures, Interferon - Tuberculosis



Does the patient have a PE? Maybe you should CTA or VQ.

Pleural fluid cytology & AFB is insensitive. Increase yield by repeating thoracentesis. If still negative, diagnose malignancy with pleural biopsy.



An 81-year-old man is evaluated in the hospital for pneumonia.

On physical examination, temperature is 38.0 °C (100.4 °F), blood pressure is 119/65 mm Hg, pulse rate is 110/min, and respiration rate is 24/min. Oxygen saturation is 88% with the patient breathing ambient air. There are coarse rhonchi and decreased breath sounds and dullness to percussion over the left lower half of the chest. CT scan of the chest shows left lower lobe consolidation and loculated left pleural effusion. A thoracentesis is performed.

Which of the following tests is necessary in directing subsequent therapy?

A Pleural lactate dehydrogenase

B Pleural pH

C Serum Procalcitonin

D Pleural Total protein

E Chest x-ray



Complicated Parapneumonic Effusion Must Be Completely Drained! Thoracentesis -> Chest Tube -> VATS

Antibiotics should include anaerobic coverage.

How do we diagnose it?

If **Left Lateral Decubitus CXR** shows >10mm of pleural fluid between lung and chest wall (or is non-layering), perform thoracentesis.



Left Lateral Decubitus Film



What Makes it Complicated?

Loculations

Acidity (pH < 7.2)

Hypoglycemic (Glu < 60mg/dL)

Positive Gram Stain/Culture

Gross Pus



Still don't know what it is?

Viral pleural effusion is a diagnosis of exclusion. If completely resolved with thoracentesis, can follow clinically.

Malignant Effusions

Manage with serial thoracentesis, pleurex cathether, or talc pleurodesis.

Uncommon alternatives: Pleurectomy, pleuroperitoneal shunt.



A 63-year-old man is evaluated for dyspnea. The patient has newly diagnosed stage IV lung adenocarcinoma with a right malignant pleural effusion. A thoracentesis removed 1200 mL of bloody fluid 5 days ago with relief of dyspnea. Follow-up chest radiograph documented incomplete expansion of the right lung, with air replacing the removed pleural fluid. He has returned to the emergency department today with increasing dyspnea.

On physical examination, vital signs are normal. Respiration rate is 18/min. Oxygen saturation \triangle is 93% with the patient breathing ambient air. Lung examination is consistent with a large right pleural effusion.

Repeat chest radiograph shows a large right-sided pleural effusion with loculations.

Which of the following is the most appropriate management?

- A Chemical pleurodesis
- B Indwelling pleural catheter
- C Repeat thoracentesis
- D Surgical decortication



* Post-thoracentesis chest CT

Boring things we didn't cover:

- Sleep medicine.
- Obstructive and central sleep apnea.
- Obesity hypoventilation syndrome.
- Mountain sickness.
- High altitude cerebral & pulmonary edema.
- Air travel with pulmonary disease.

