

# Congenital, Benign and Inflammatory Lung diseases

Joonho Jung, M.D.

Assistant professor

Department of Thoracic & Cardiovascular Surgery

Ajou University School of Medicine

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- Congenital lesions of the lung
- Congenital vascular lesions of the lung
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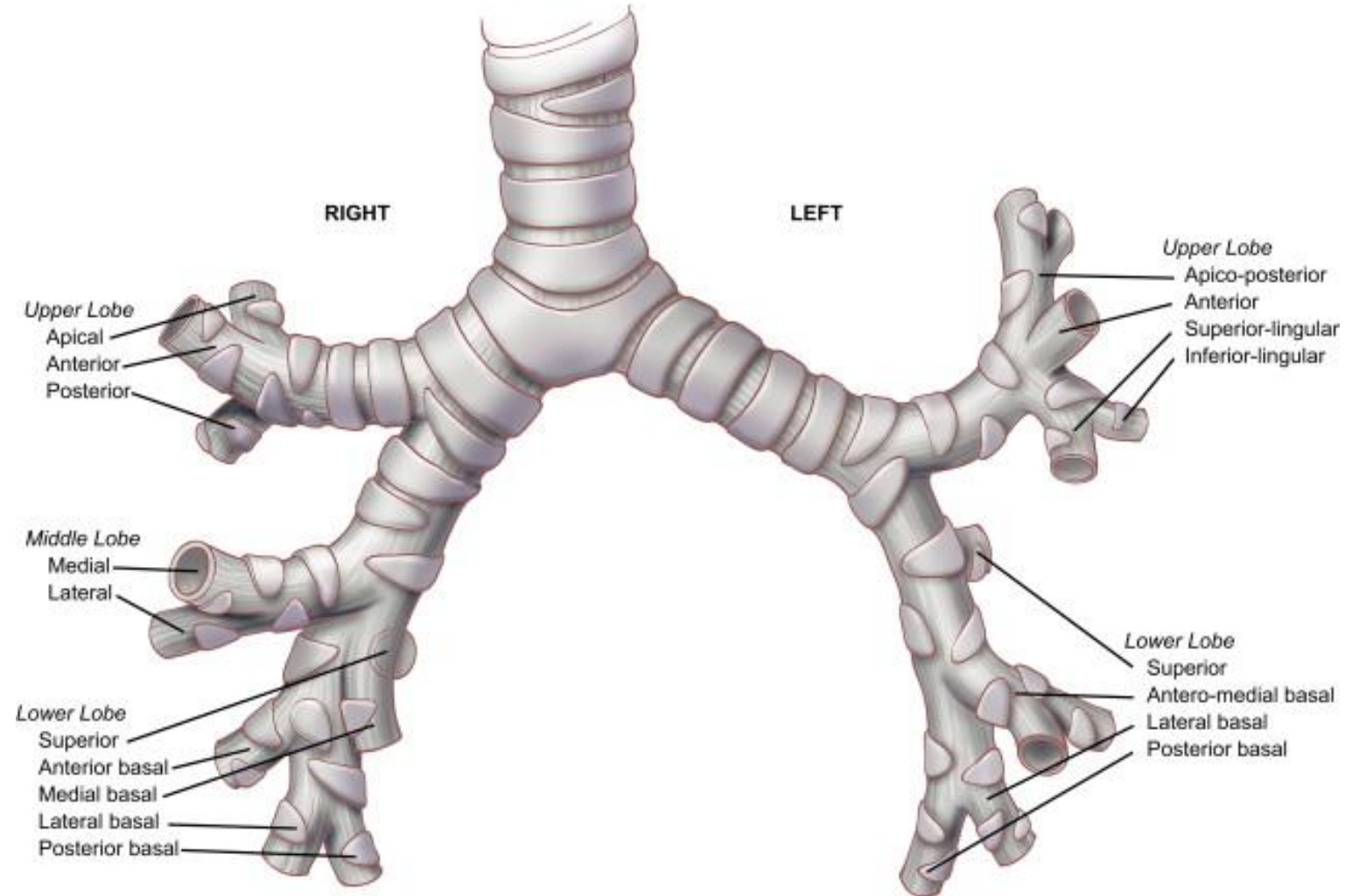
# Congenital Lesions of the Lung

- Tracheal agenesis and atresia
- Bronchial anomalies
  - Tracheal diverticulum
  - Tracheal bronchus
  - Bronchial atresia
- Congenital lobar emphysema
- Pulmonary dysplasia
- Sequestration
- Bronchogenic cyst
- Congenital cystic adenomatoid malformation

# Congenital Lesions of the Lung

- Tracheal agenesis and atresia
- Bronchial anomalies
  - Tracheal diverticulum
  - **Tracheal bronchus**
  - Bronchial atresia
- **Congenital lobar emphysema**
- Pulmonary dysplasia
- **Sequestration**
- **Bronchogenic cyst**
- **Congenital cystic adenomatoid malformation**

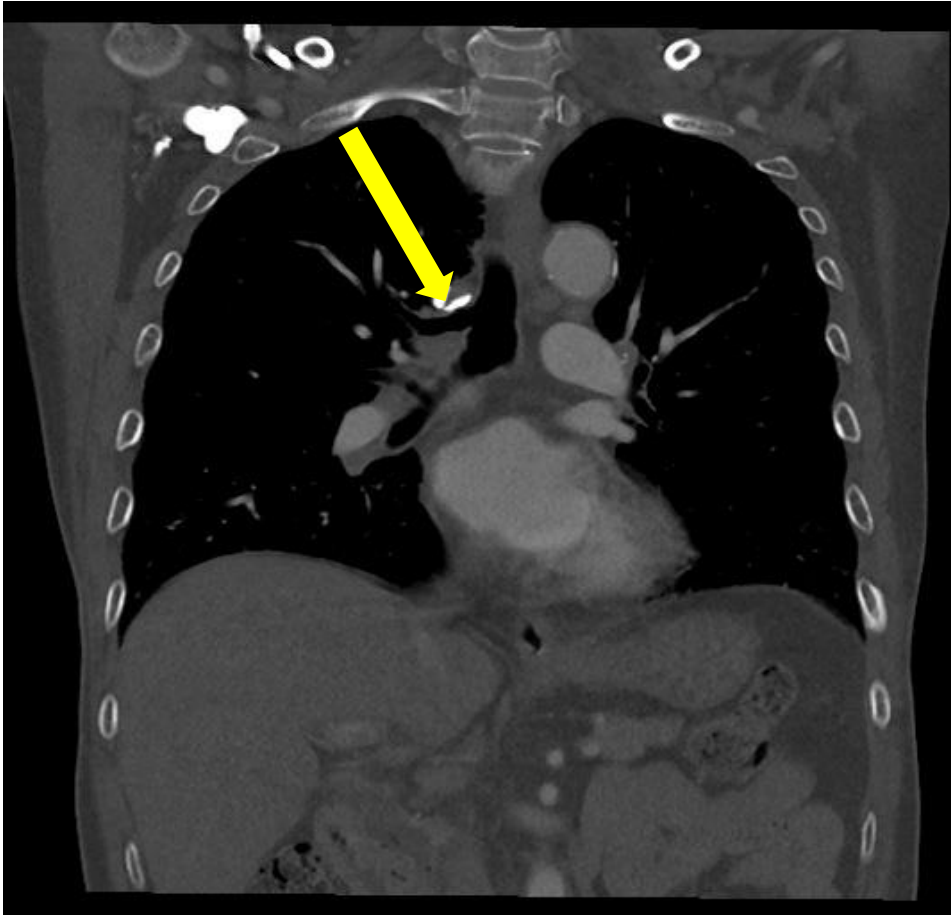
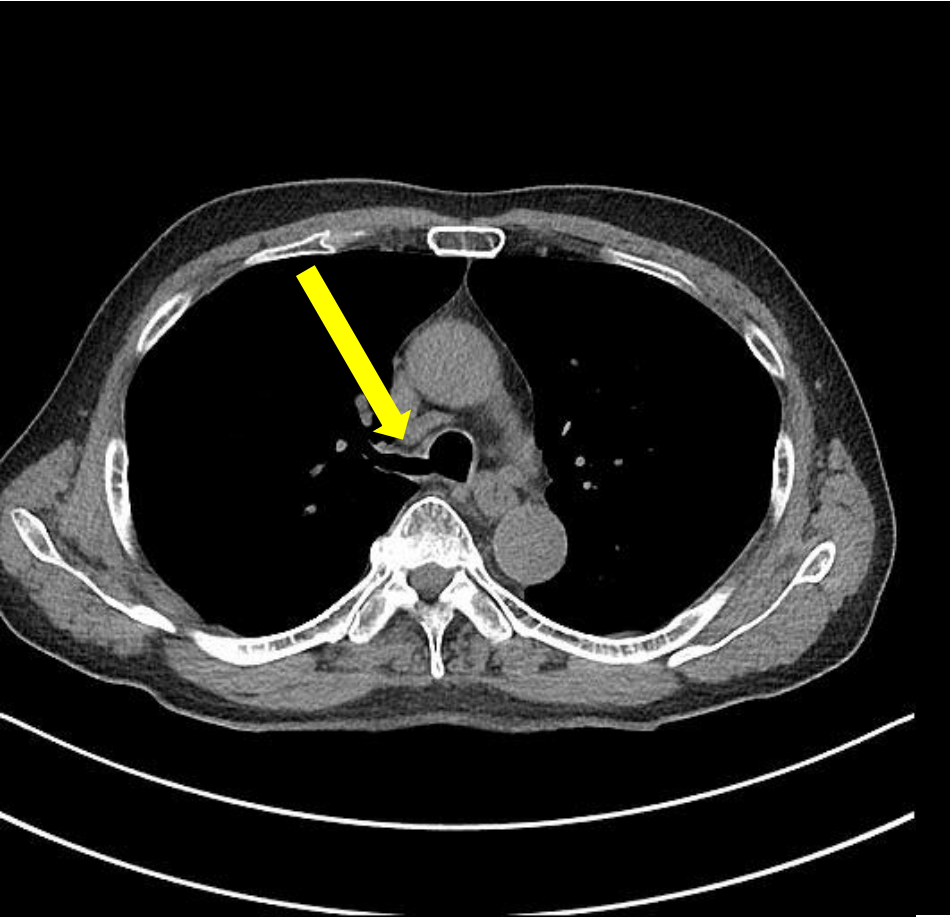
# Tracheal bronchus



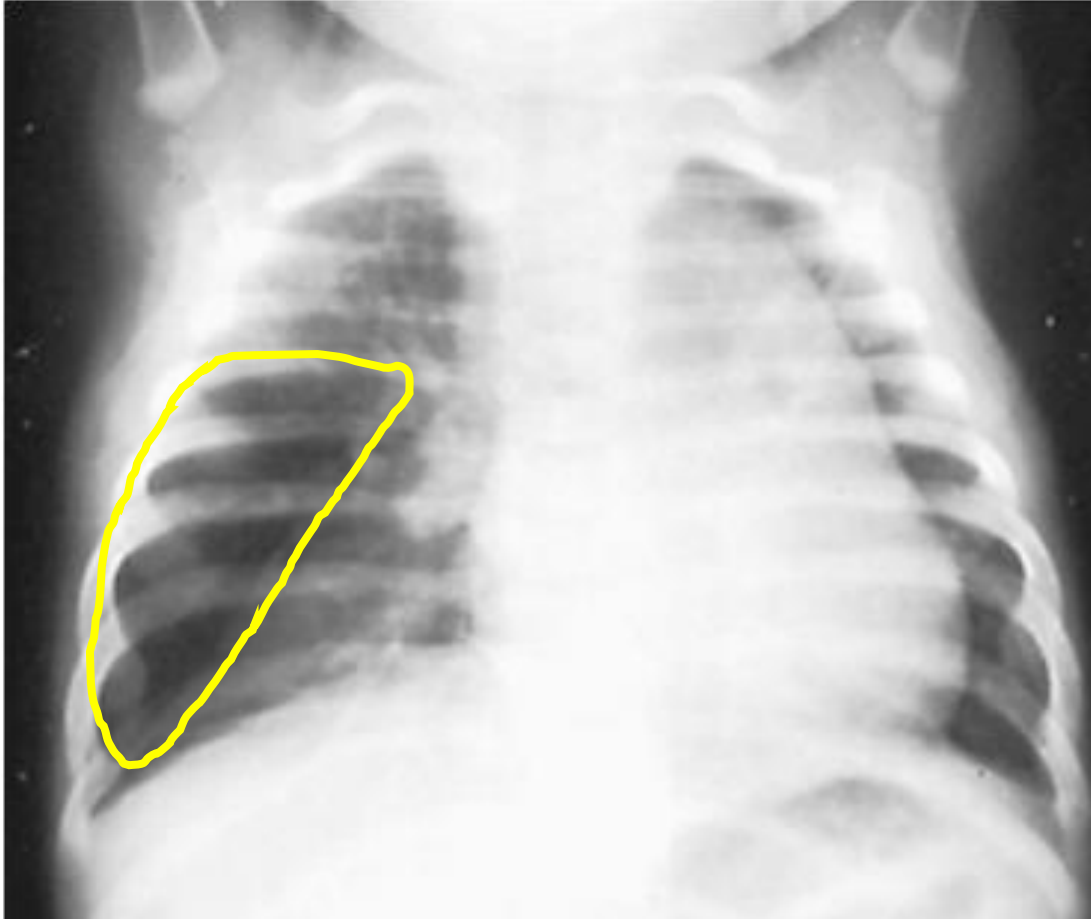
# Tracheal bronchus



# Tracheal bronchus



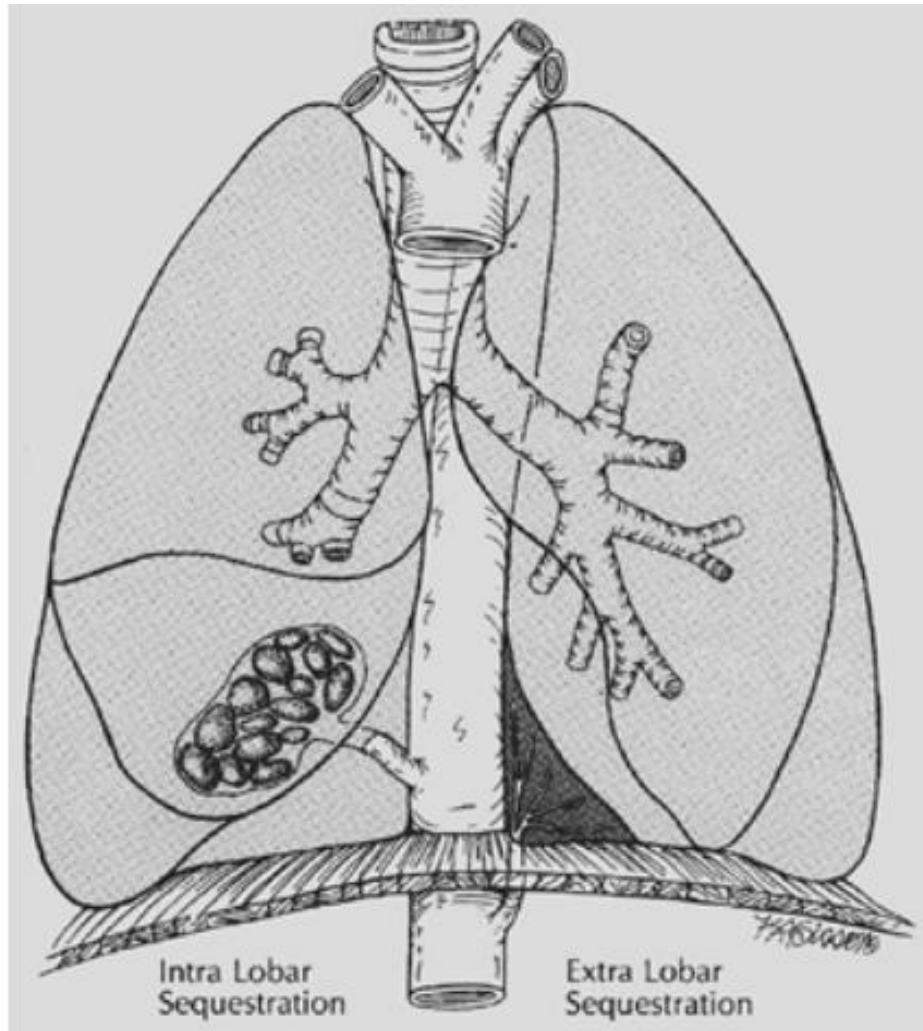
# Congenital lobar emphysema



- Isolated hyperinflation of a lobe without extrinsic bronchial obstruction
- Frequent lobe: LUL, RML
- Severe pulmonary distress since infant
- Treatment: resection



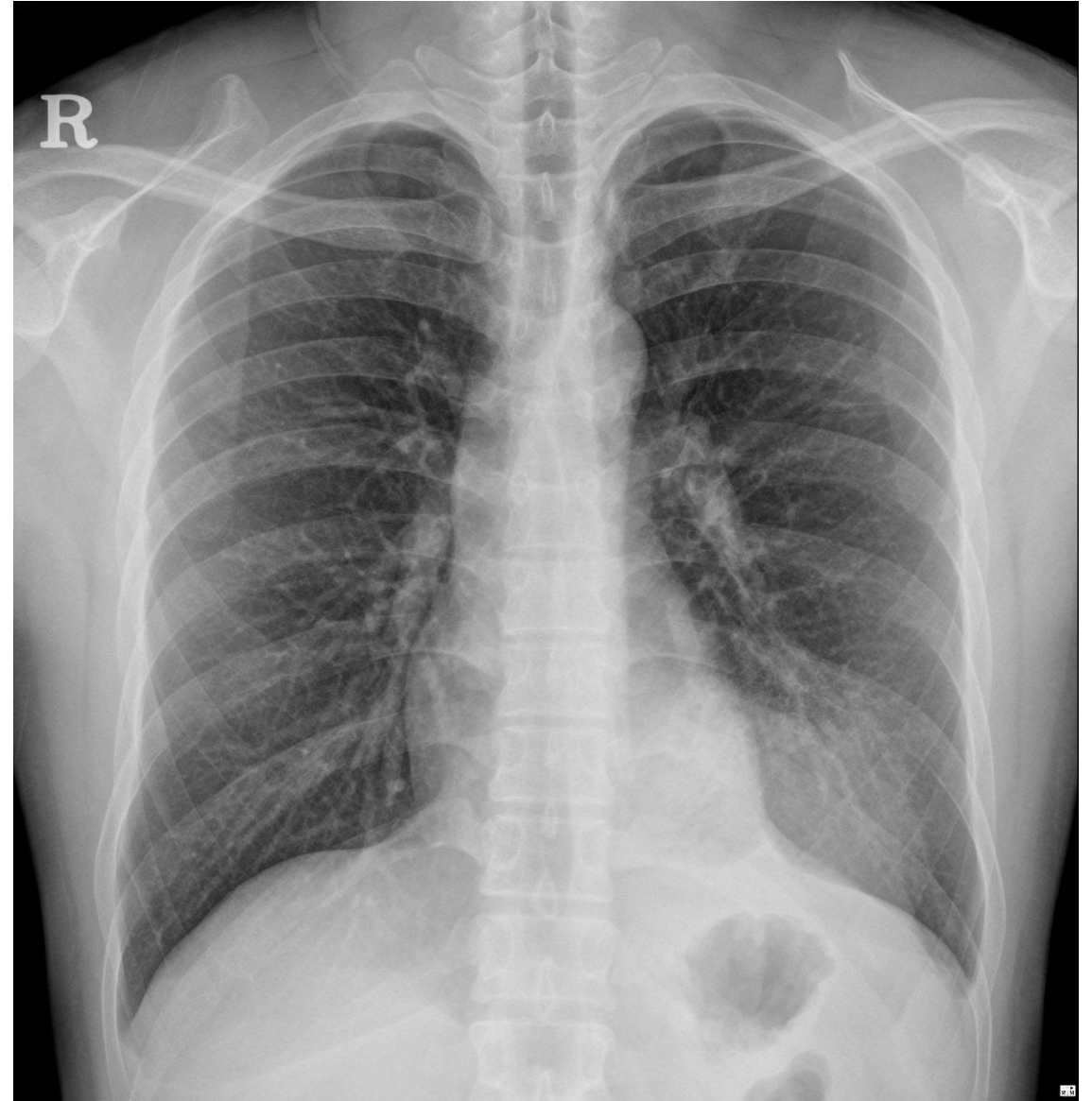
# Sequestration

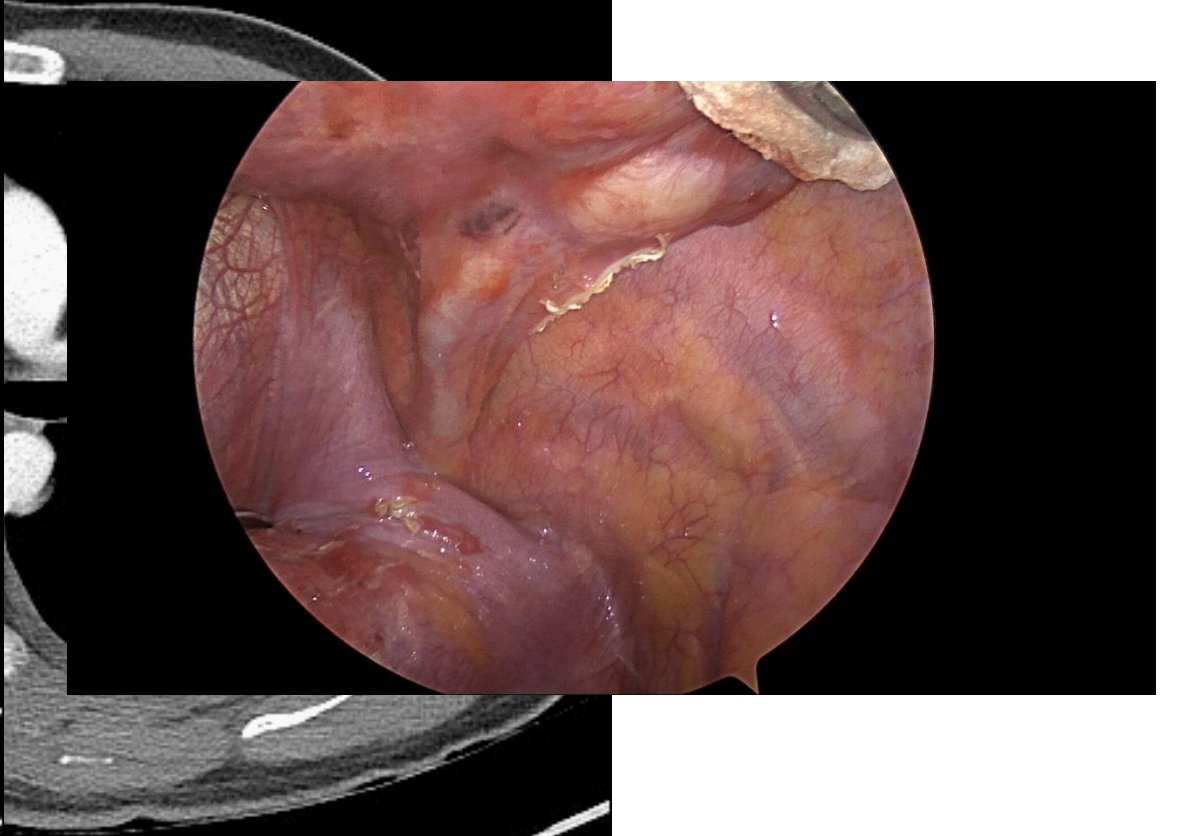


- Definition
  - A segment or lobe of lung tissue that has no bronchial communication with the normal tracheobronchial tree
  - Arterial supply: systemic vessel
  - Venous drainage: pulmonary vein > systemic vein
- Extralobar sequestration
  - Separated from normal lung by own visceral pleura
  - 25% of sequestration
  - Left (90%), posterior CPA angle (frequent), diaphragmatic hernia(30%)
- Intralobar sequestration
  - Within the normal lung parenchyma
  - Communication through the pores of Kohn may lead to chronic infection
  - Left > Right
  - Children and young adults: recurrent pneumonia in LLL

# Sequestration

- 26 year-old male
- Recurrent pneumonia in LLL and frequent hemoptysis





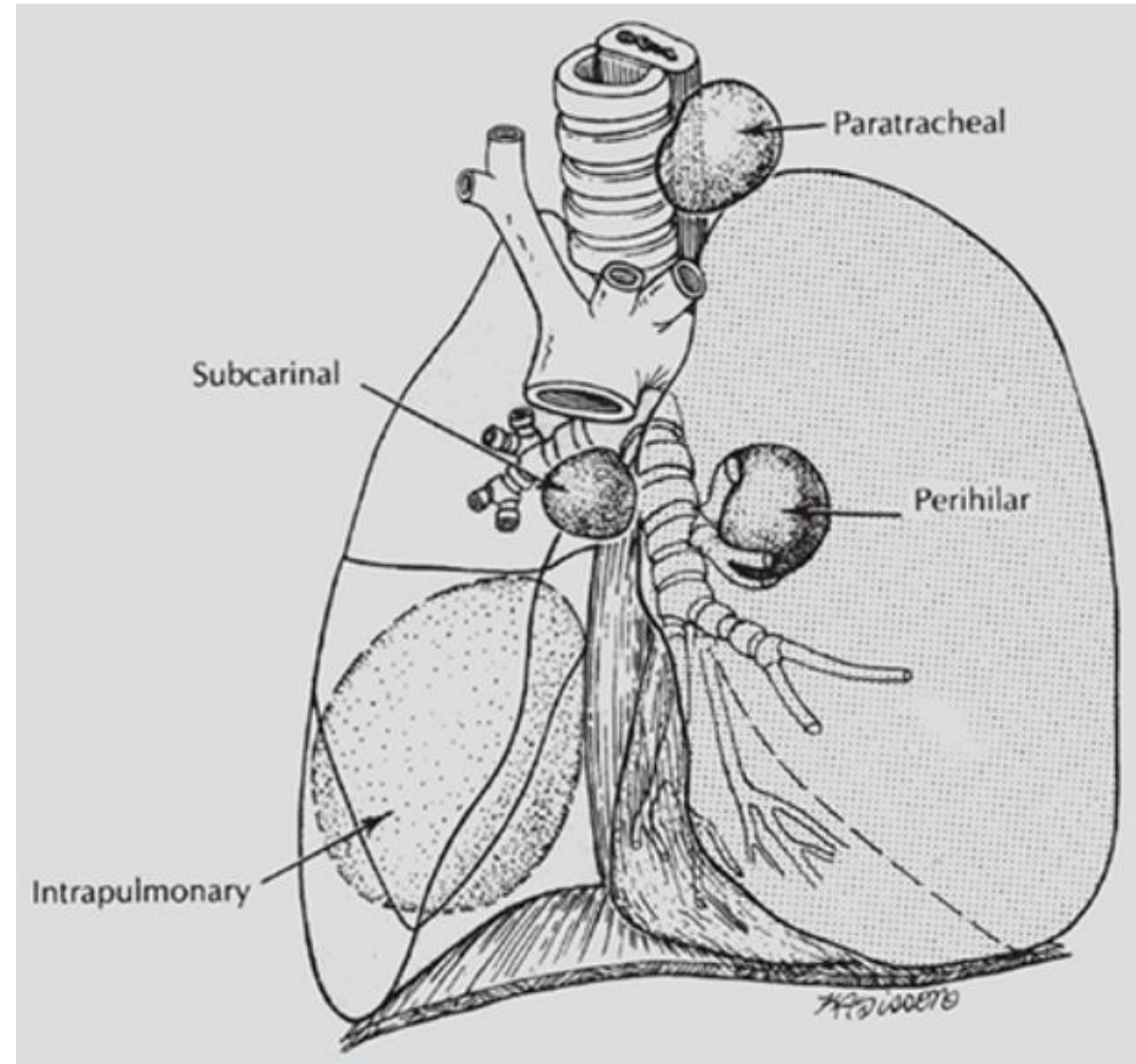




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cm

# Bronchogenic cyst

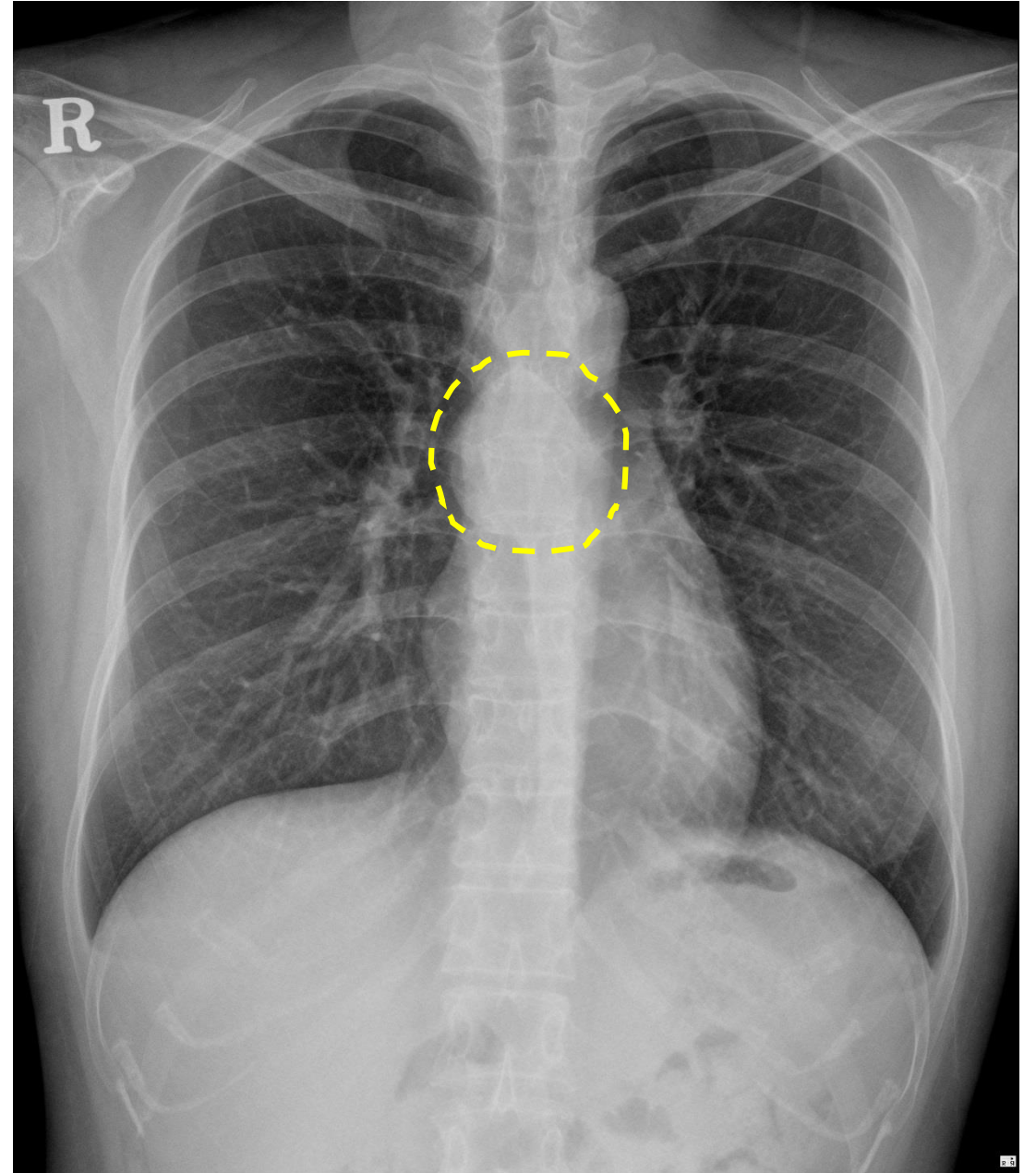
- Bronchogenic cyst
  - Uncommon in extrathoracic locations
  - Lined with ciliated columnar or cuboidal epithelium
  - Thinc wall, may contain cartilage and bronchial glands
  - Treatment: surgical resection
    - Minimally invasive technique
    - Parenchymal cyst: lung resection
    - Morbidity and mortality: zero



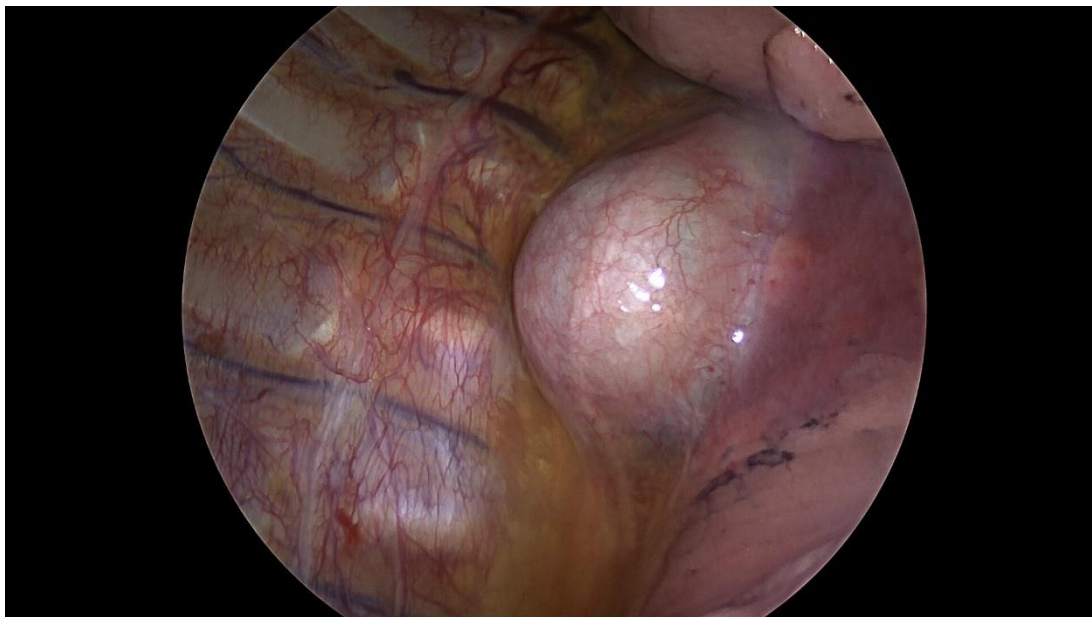


# Bronchogenic cyst

- 31 year-old female
- No symptom
- Incidental finding in medical check-up



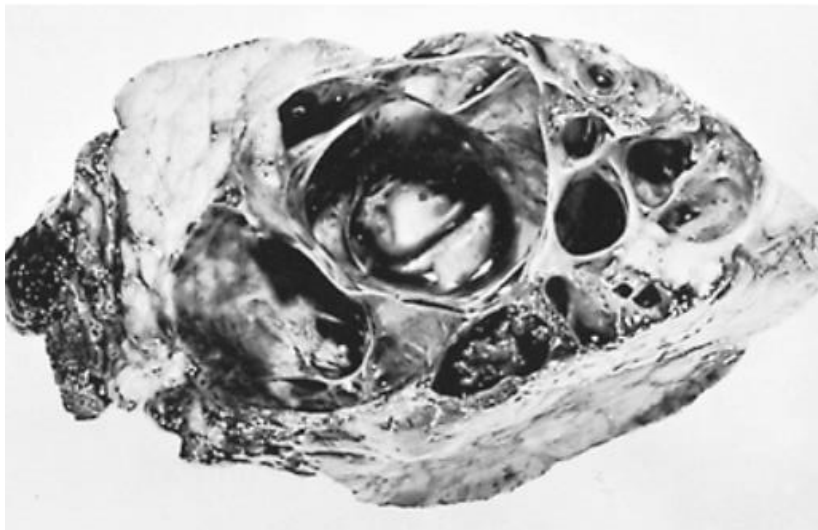
# Bronchogenic cyst



# Congenital cystic adenomatoid malformation

- Definition

- A spectrum of cystic and solid lesions of the lung can be identified histologically as CCAMs.
- An overgrowth of terminal bronchiolar-type tubular structures and a lack of mature alveoli.



- Histology

- An adenomatoid increase of terminal respiratory bronchiole-like structures
  - Lined by ciliated columnar epithelium occurs.
  - Interspersed cysts → immature alveoli
  - Connective stroma → disorganized elastic tissue and smooth muscle
- Mucosa cyst: lined with bronchial-type epithelium, polypoid overgrowth
- Bronchial mucoserous glands and cartilaginous plates: absent



# Classification of CCAM

by Stocker et al. 1977

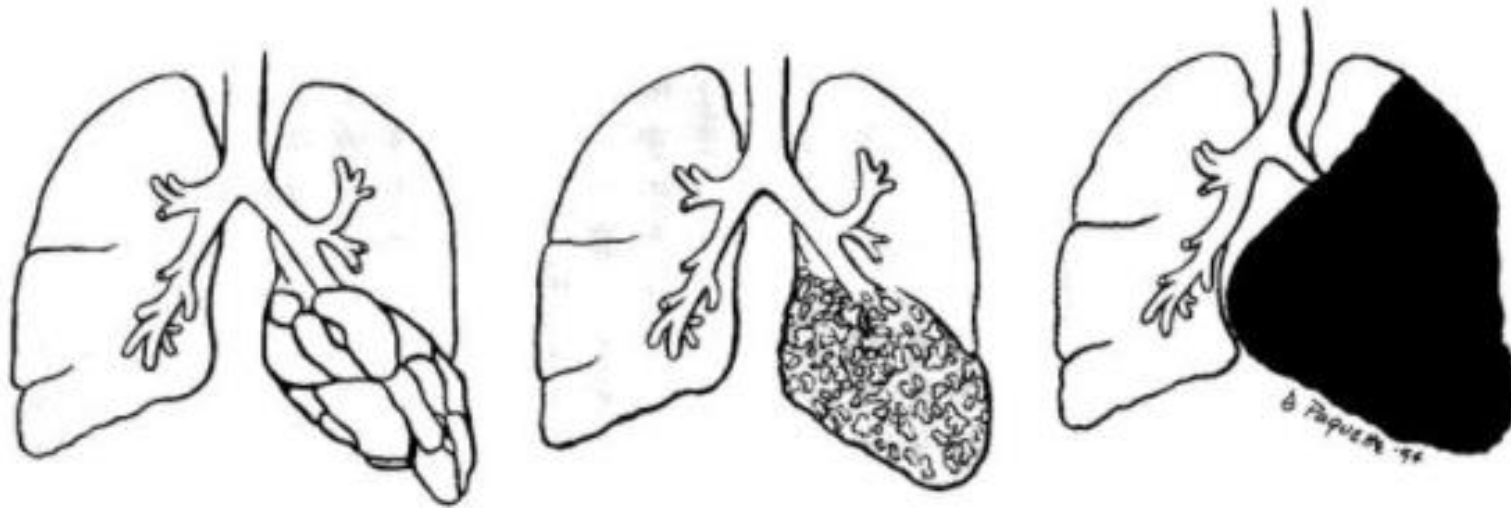


Figure 37-1. Depiction of Stocker's classification of type I, II, and III CCAM.

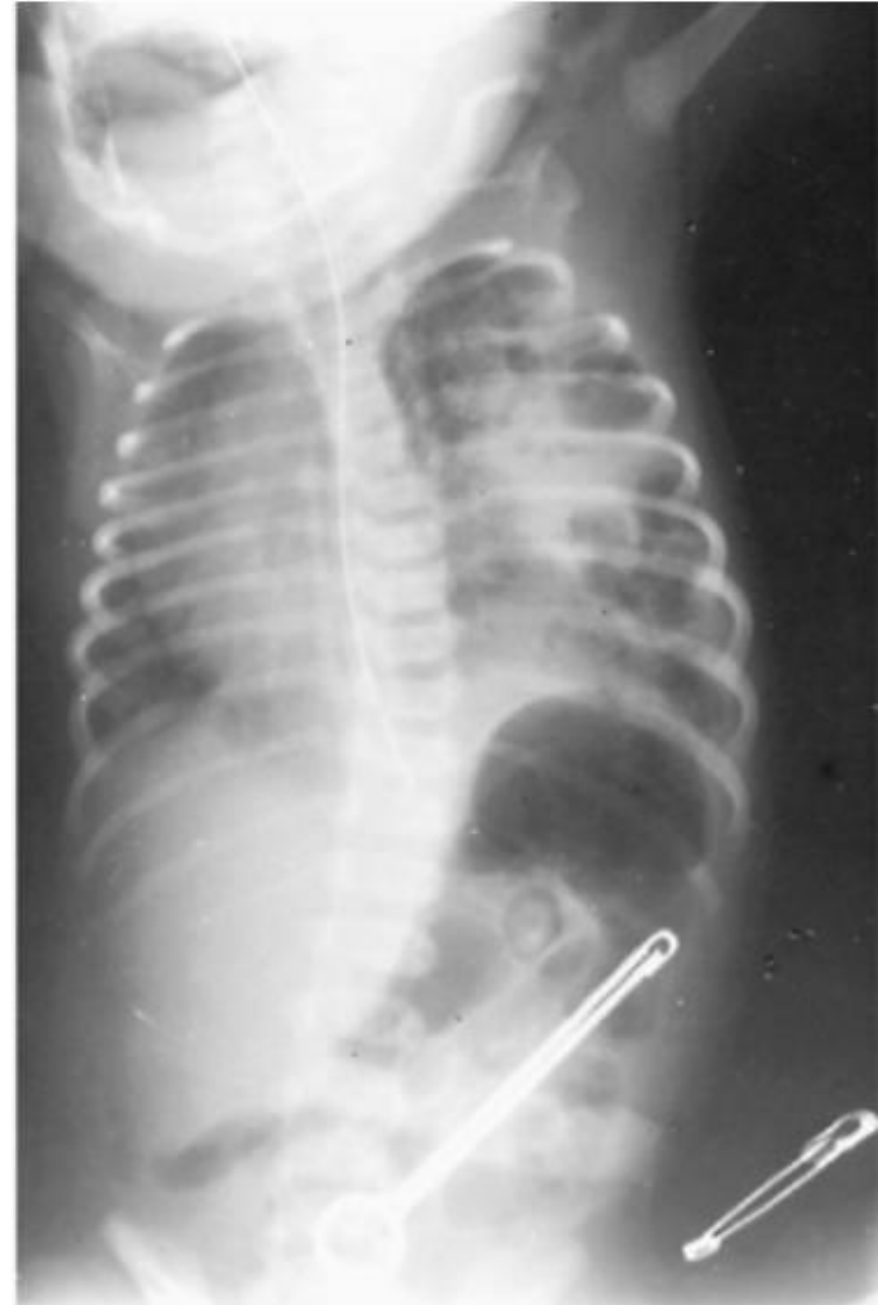
**Type I:** 50% of postnatal cases, favorite outcome

**Type II:** 40% of postnatal cases, high frequency of associated congenital anomalies

**Type III:** 10 % of postnatal cases, large homogenous microcystic mass, mediastinal shift  
poor prognosis → non-immune hydrops fetalis, cardiorespiratory compromise

# CCAM

- Treatment
  - Newborn
    - With large CCAM
      - Respiratory distress
      - Space occupying mass
      - Compression of contralateral lung
    - Surgical resection
  - Older child or adult
    - Surgical resection: source of recurrent pneumonia
- Timing of surgery
- Malignancy potential: some reports



# Congenital Vascular Lesions of the Lungs

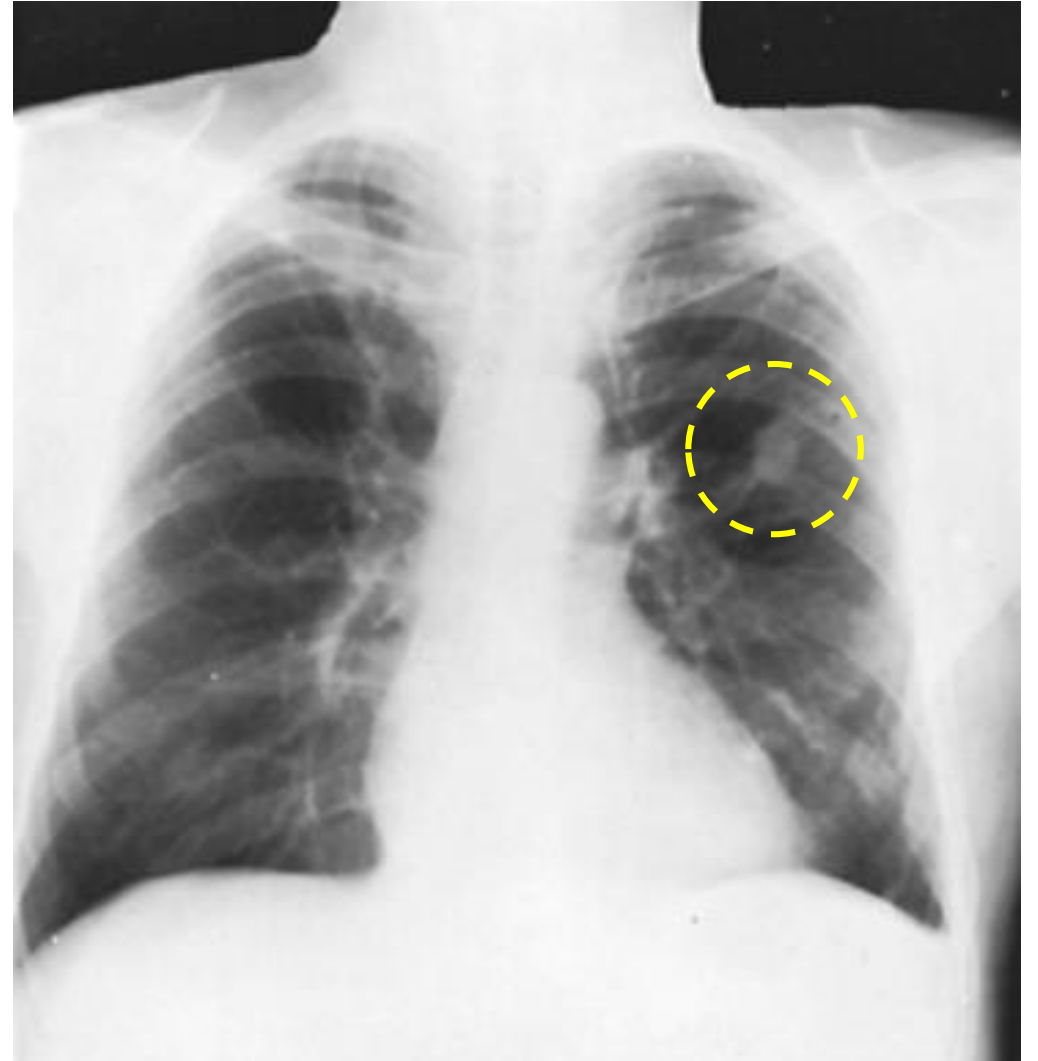
- Agenesis of a pulmonary artery
- Stenosis of a branch or branches of the pulmonary arteries
- Pulmonary arteriovenous fistula
- Abnormal pulmonary venous connection
- Varicosities of the pulmonary veins
- Lymphangiectasia

# Congenital Vascular Lesions of the Lungs

- Agenesia of a pulmonary artery
- Stenosis of a branch or branches of the pulmonary arteries
- **Pulmonary arteriovenous fistula**
- Abnormal pulmonary venous connection
- Varicosities of the pulmonary veins
- Lymphangiectasia

# Pulmonary arteriovenous fistula

- Definition
  - Congenital malformations that result from errant capillary development
  - With incomplete formation or disintegration of vascular septa
- Incidence: 2-3 / 100,000
- Interrupt capillary filter of lung → embolism may occurs



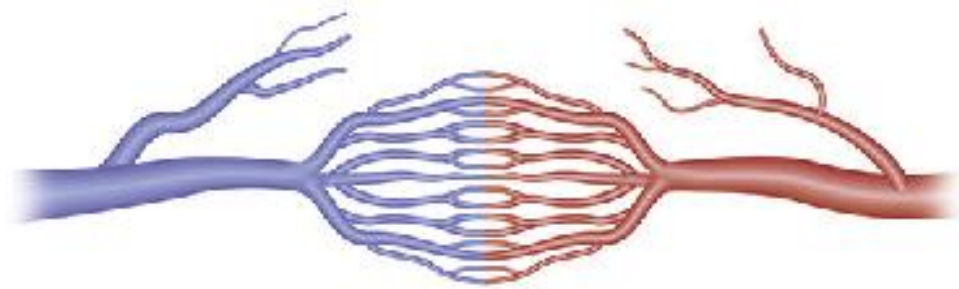
# Pulmonary arteriovenous fistula

- Symptoms

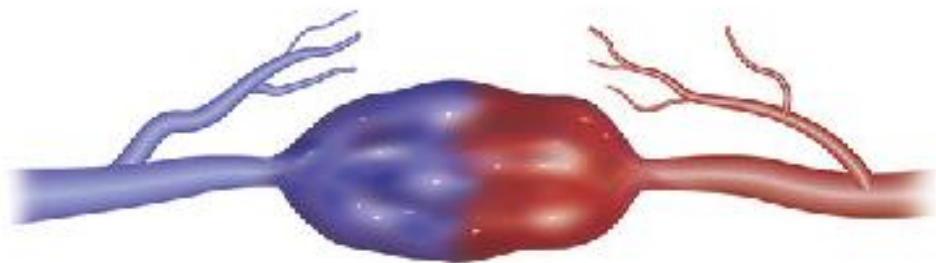
- Massive hemoptysis: uncommon
- Paradoxical embolization and stroke
  - Occurred in 10% of all untreated patients
- Brain abscess

- Treatment

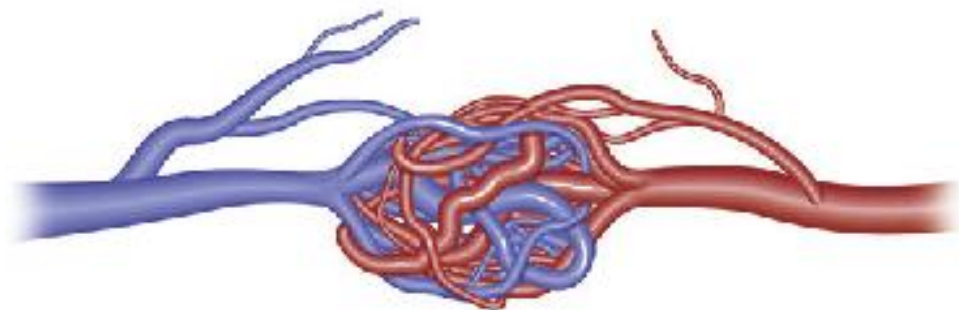
- Surgical resection
  - Best indicated in single lesion
  - Feeding artery > 3mm in diameter
    - High risk of paradoxical embolization and neurologic complications
- Embolic obliteration
  - Selective radiographically guided embolization
    - First line therapy
      - Unsuitable for surgery
      - Multiple PAVFs



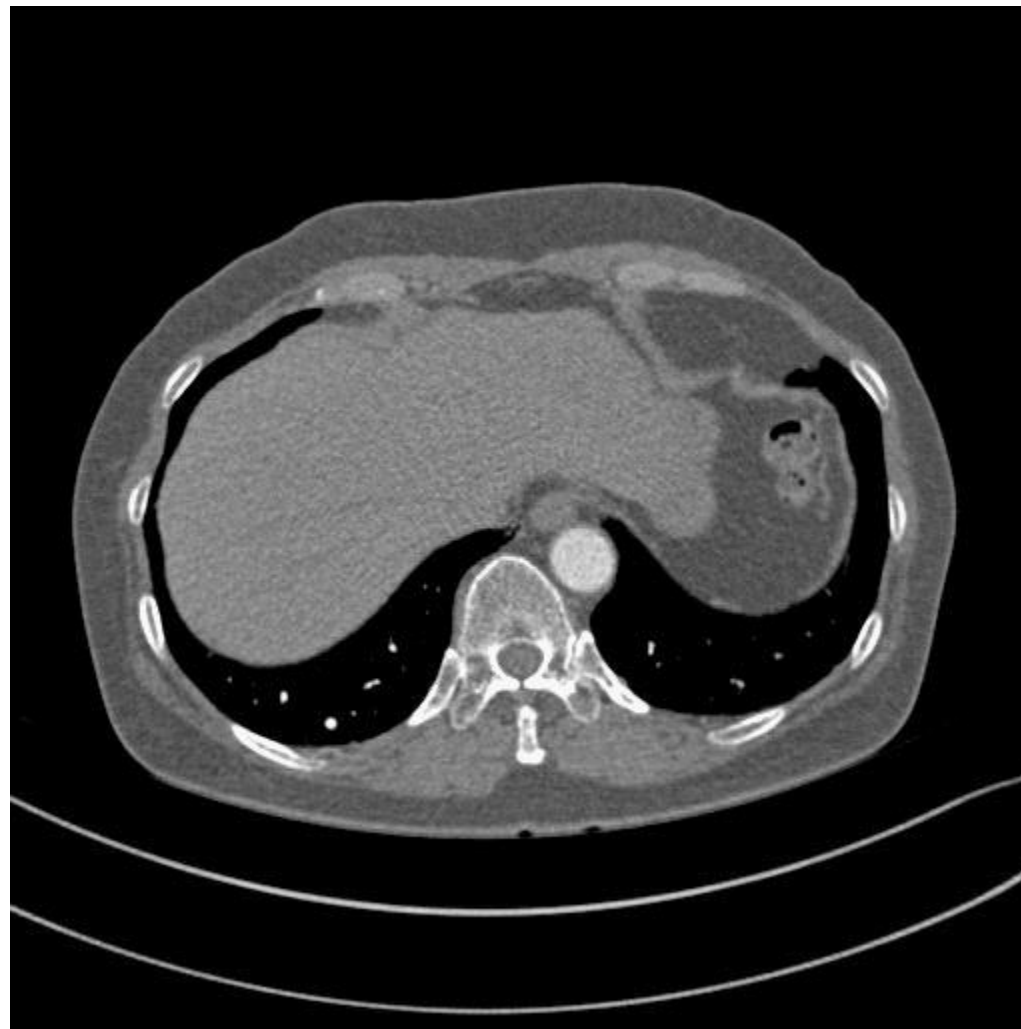
Normal



Simple AVM



Complex AVM





FD 37 cm



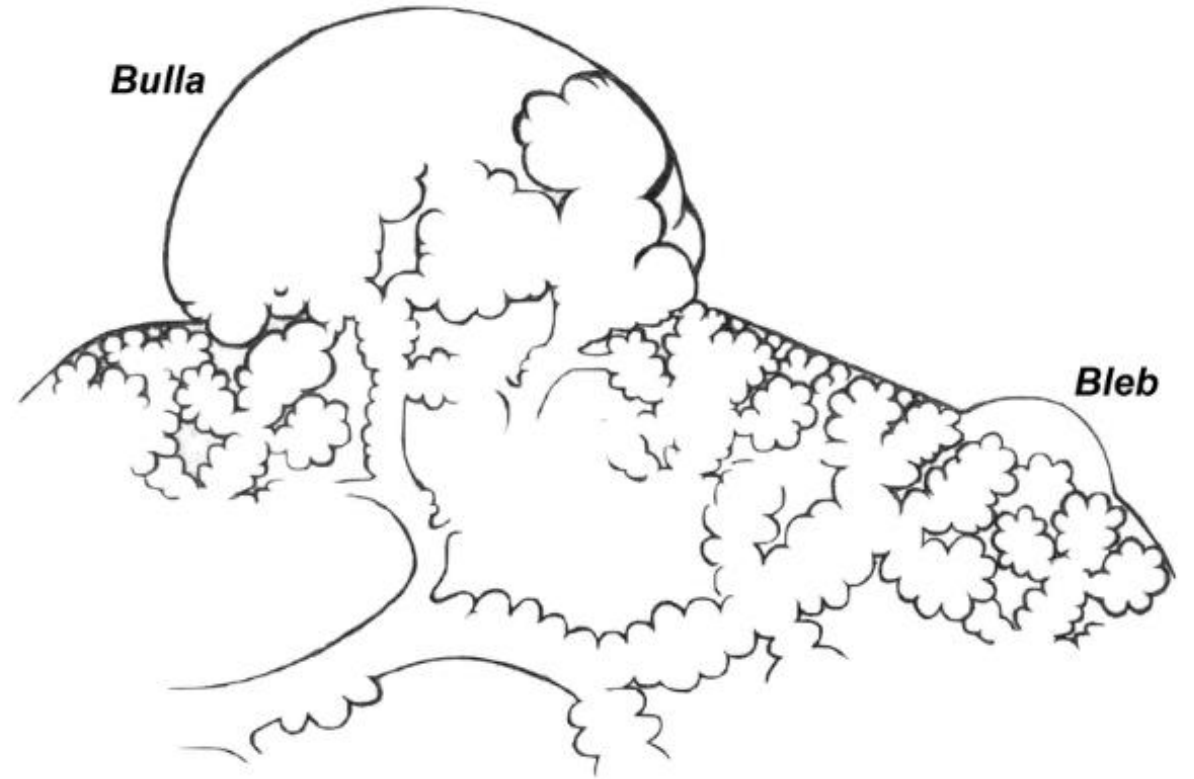
FD 42 cm





# Bullous and Bleb Diseases of the Lungs

- Blebs
  - Well-circumscribed interpleural airspaces separated from underlying parenchyma by thin pleural covering.
  - Result of subpleural alveolar rupture
  - Outer wall: visceral pleura
  - Underlying lung: normal
  - Location: apex
- Bullae
  - Associated with any variety of emphysema
  - Wall: destroyed lung
  - Bronchial opening: usually located at the base of bullae

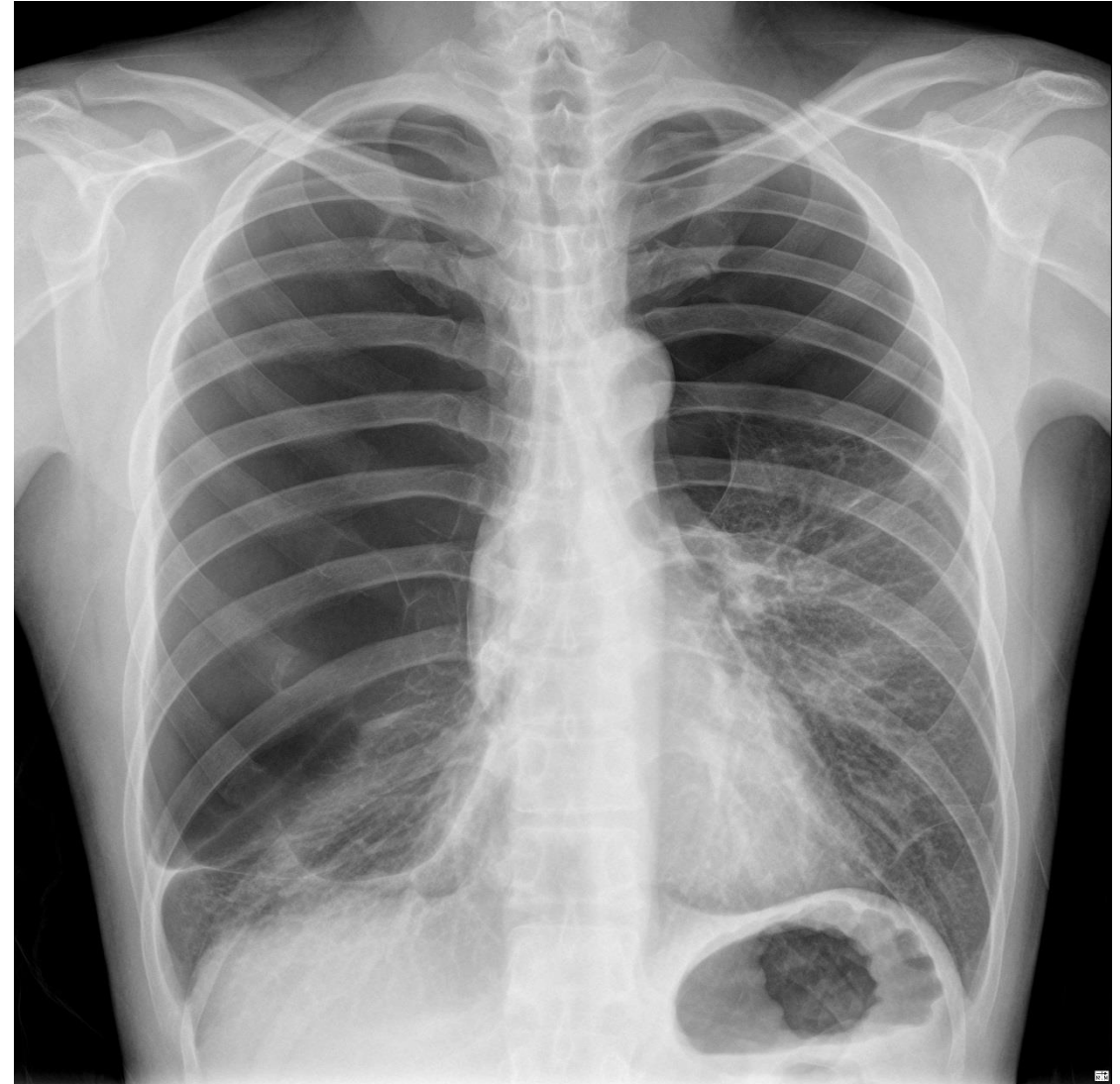


**Table 87-2 Rationale and Indications for Surgery in Patients With Complications of Their Bullae**

<i>Indication</i>	<i>Rationale for surgical approach</i>
Pneumothorax (first episode or recurrence)	Further reduction of function in patients already compromised Prolonged air leak High incidence of recurrences (>50%)
Infection of the bulla/empyema	Failure to respond to medical treatment
Hemoptysis	Management of significant hemoptysis
Chest pain	Pain clearly related to air trapping during hyperventilation
Treatment of lung cancer	Documented or highly cancer suspicious lesion

# Bullous and Bleb Diseases of the Lungs

- 45 year-old male
- Smoker
- Chief complaint: progressive dyspnea
- Misinterpretation in ER
  - Pneumothorax
  - Chest tube insertion → BPF



AJOU UNIVERSITY HOSPITAL-PFT:Elite/Dx

YoungTong-Ku Woncheon-Dong

Suwon-city, Kyungki-Do

Name: YU CHUL JAE	ID: 0532895	BSA: 1.72	Date: 2017-08-07
Tech: Cho In Ae	Height: 172.30	Age: 44	Room: 112W.34.06
Doctor:	Weight: 61.10	Sex: Male	Race: Asian

Diagnosis:

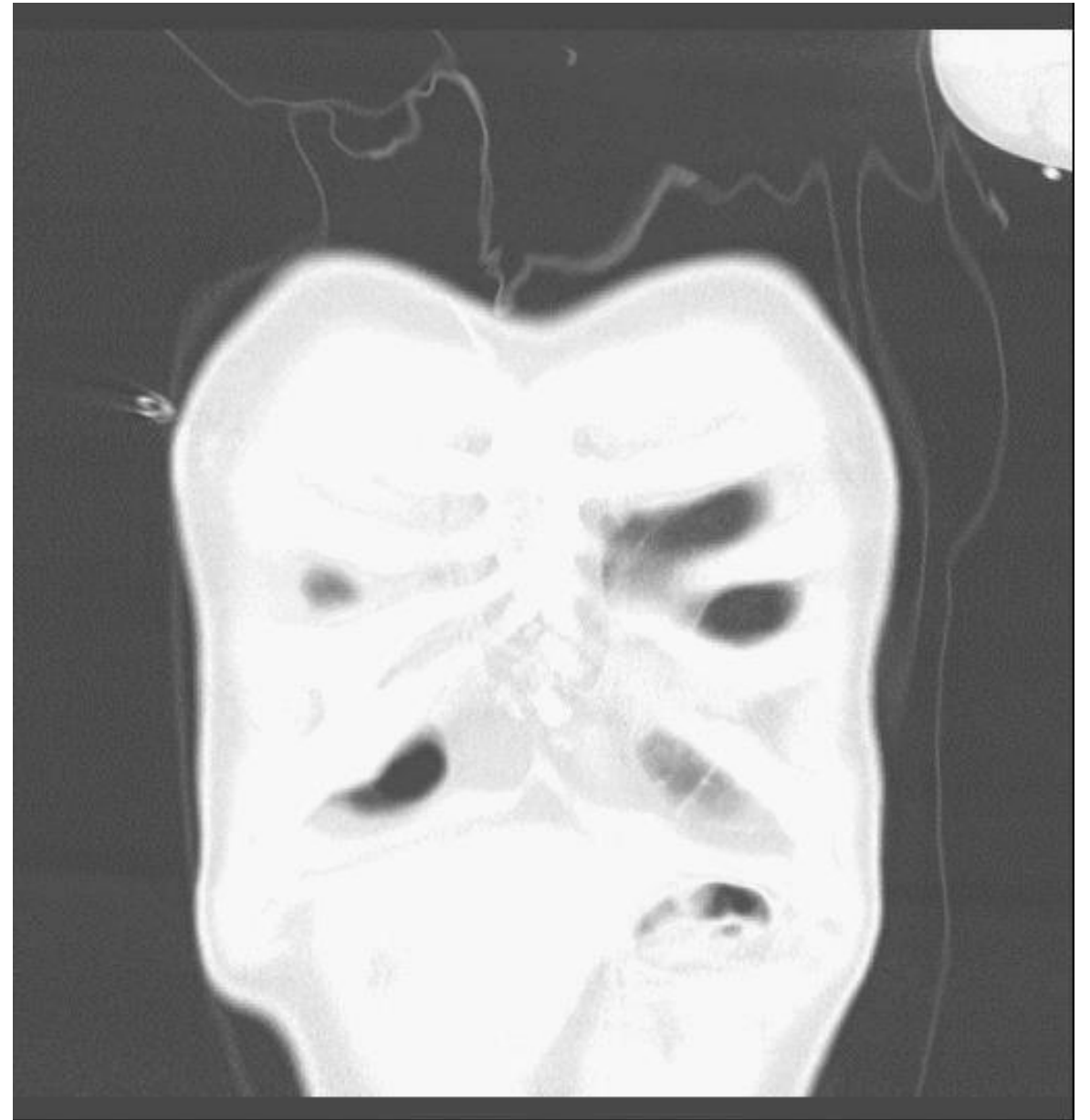
Dyspnea: Cough: Wheeze:  
Tbc Prod: Yrs Smk: Pks/Day: Yrs Quit:

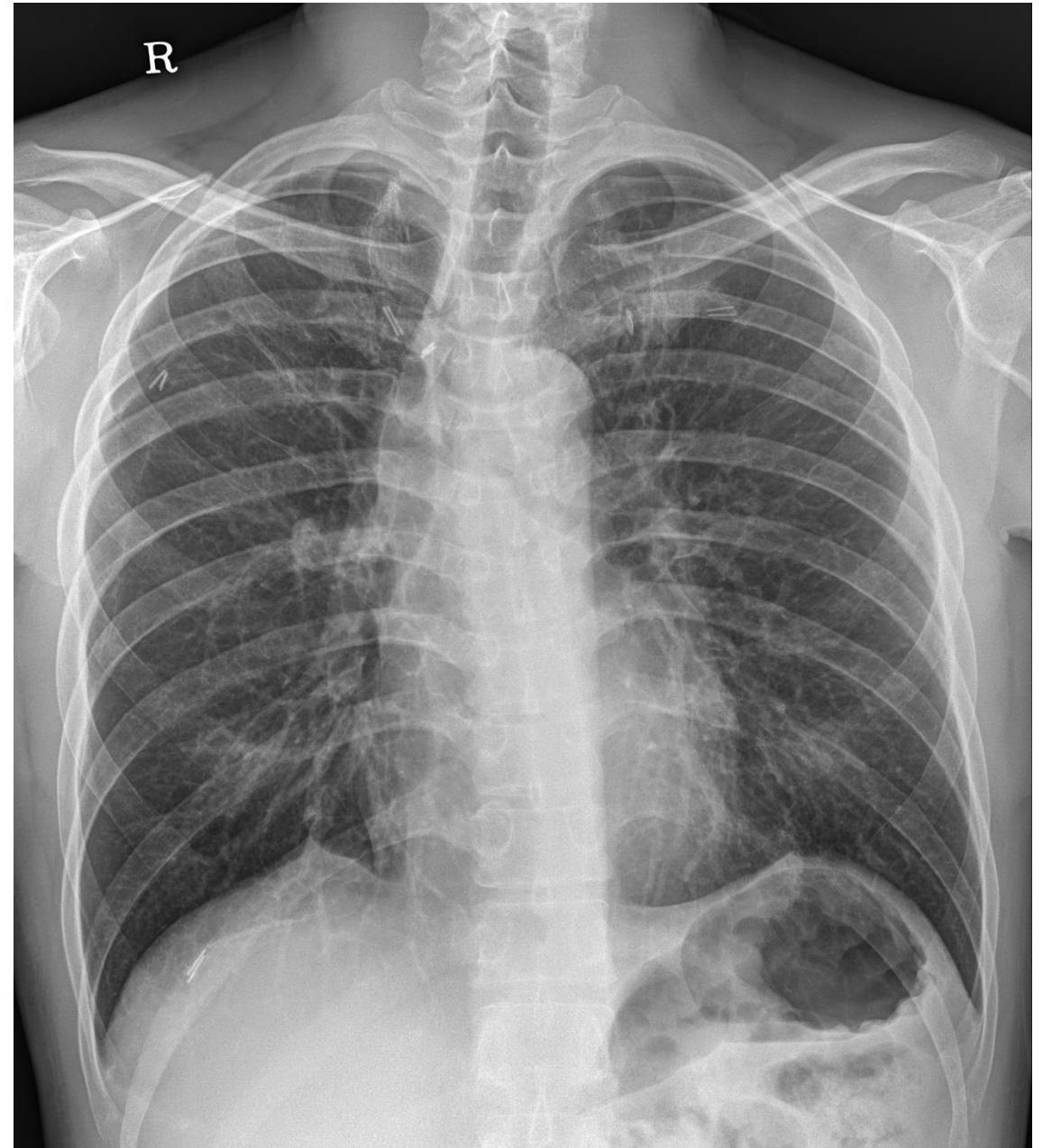
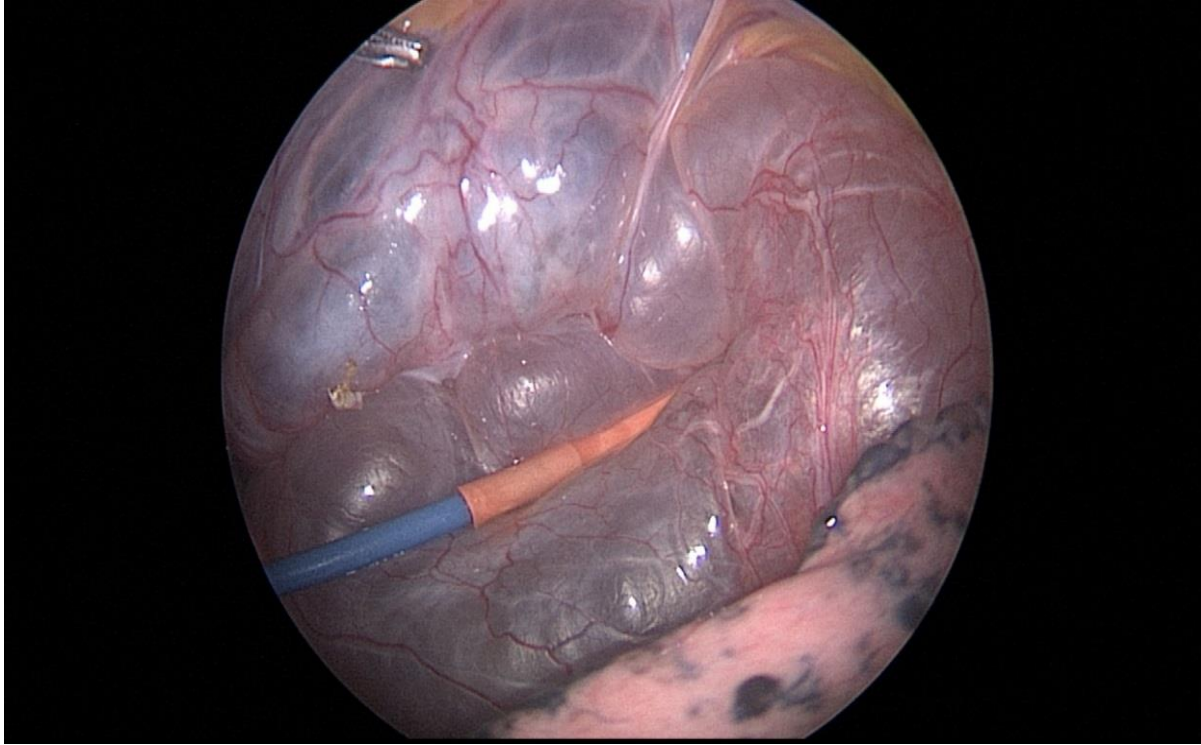
Medications:

Pre Test Comments:

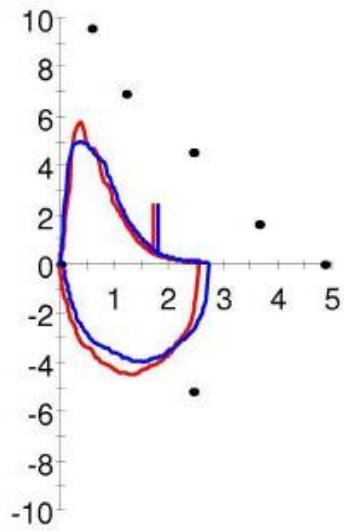
Post Test Comments:

	Pre-Bronch			Post-Bronch		
	Actual	Pred	% Pred	Actual	% Pred	% Chng
--- SPIROMETRY ---						
FVC (L)	2.55	4.85	52	2.73	56	+6
FEV1 (L)	1.74	3.83	45	1.82	47	+4
FEV1/FVC (%)	68	79	86	67	84	-2
FEF 25% (L/sec)	4.65	6.97	66	4.52	64	-2
FEF 75% (L/sec)	0.26	1.61	16	0.34	21	+31
FEF 25-75% (L/sec)	0.88	3.57	24	1.13	31	+28
FEF Max (L/sec)	5.70	9.58	59	4.93	51	-13
FIVC (L)	2.55			2.66		+4
FIF Max (L/sec)	4.47			3.97		-11
--- LUNG VOLUMES ---						
SVC (L)	2.54	4.85	52			
IC (L)	1.66	3.27	50			
ERV (L)	0.88	1.58	55			
--- DIFFUSION ---						
DLCOunc (ml/min/mmHg)	17.73	29.42	60			
DLCOcor (ml/min/mmHg)	17.59	29.42	59			
DL/VA (ml/min/mmHg/L)	5.22	4.52	115			
VA (L)	3.37	6.51	51			
Hgb (gm/dL)	14.9	12-18				









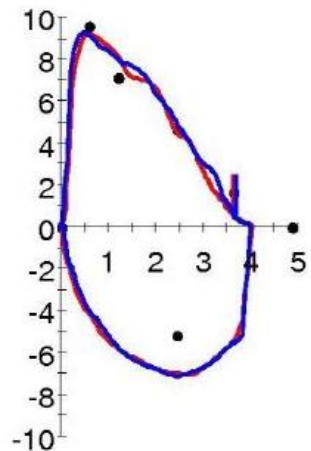
• Pred — Pre — Post

--- SPIROMETRY ---

	Pre-Bronch			Post-Bronch		
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--- LUNG VOLUMES ---

SVC (L)	2.54	4.85	52			
IC (L)	1.66	3.27	50			
ERV (L)	0.88	1.58	55			



• Pred — Pre — Post

--- SPIROMETRY ---

	Pre-Bronch			Post-Bronch		
	Actual	Pred	%Pred	Actual	%Pred	% Chng
FVC (L)	4.04	4.85	83	4.02	82	+0
FEV1 (L)	3.64	3.83	95	3.71	96	+1
FEV1/FVC (%)	90	79	114	92	116	+2
FEF 25% (L/sec)	8.59	7.14	120	8.35	116	-2
FEF 75% (L/sec)	2.54	1.65	153	2.99	181	+17
FEF 25-75% (L/sec)	5.30	3.57	148	5.49	153	+3
FEF Max (L/sec)	9.11	9.58	95	9.15	95	+0
FIVC (L)	4.01			3.97		+0
FIF Max (L/sec)	7.05			7.11		+0

--- LUNG VOLUMES ---

SVC (L)	3.95	4.85	81			
IC (L)	2.46	3.27	75			
ERV (L)	1.48	1.58	93			

# Bacterial Infections of the Lungs

- Bronchiectasis
- Lung abscess

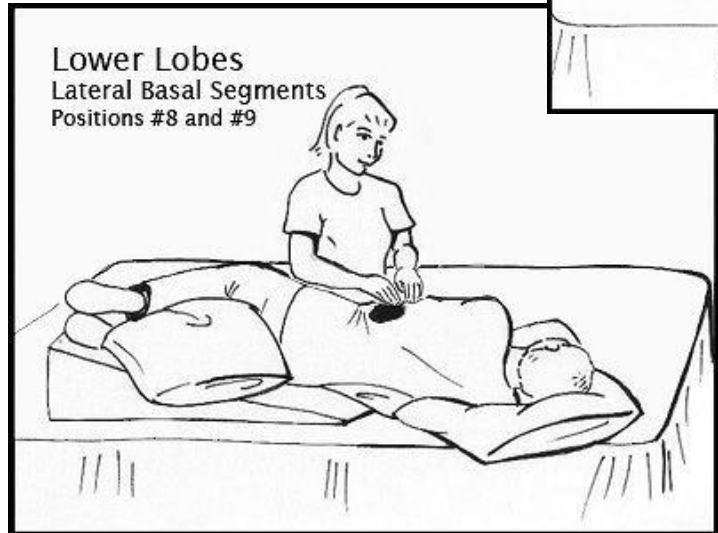
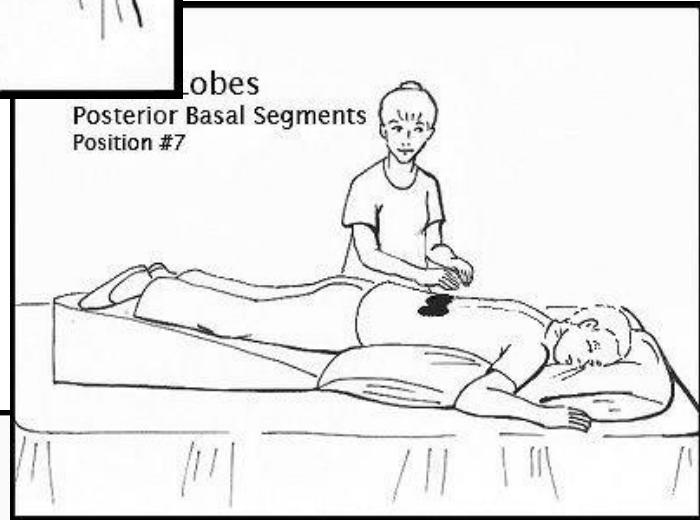
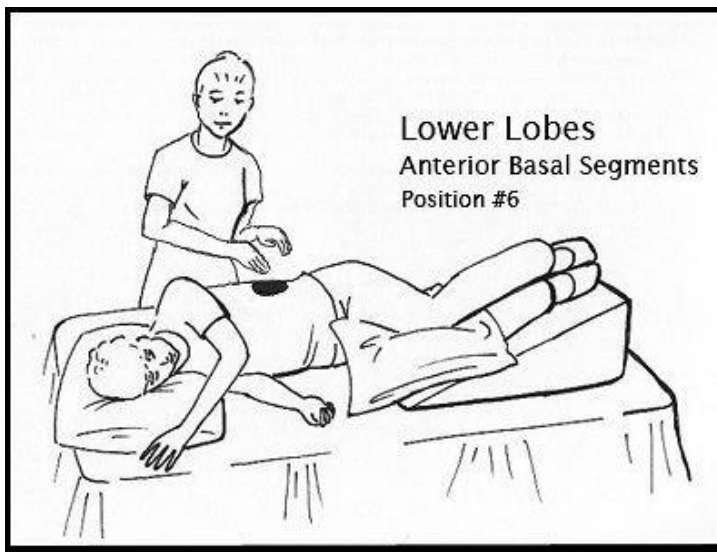
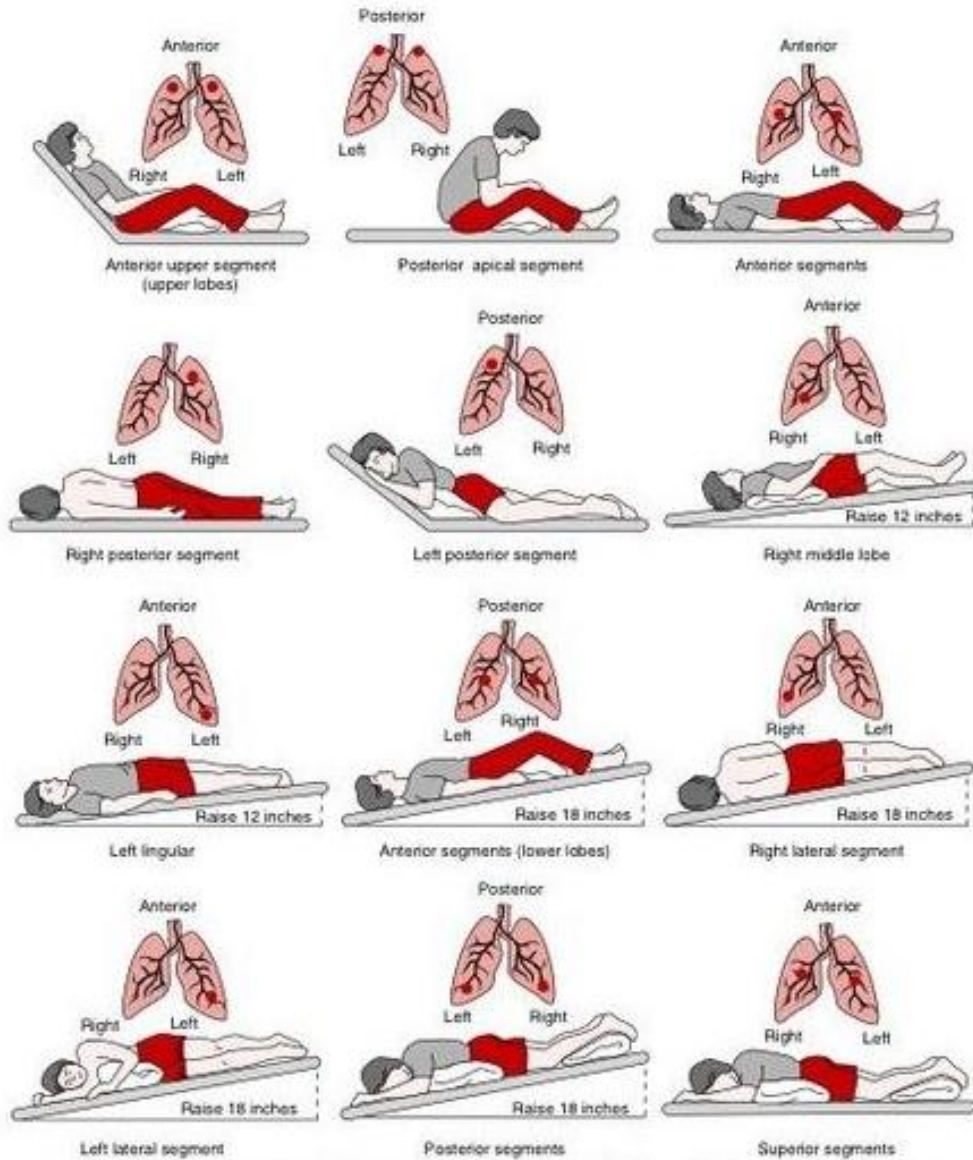
# Bronchiectasis

- Definition
  - Bronchiectasis
    - In *Greek*
      - Bronchus + *Ektasis* (dilatation)
    - Abnormal permanent dilatation of subsegmental airways
- Frequency (in order)
  - Left lower lobe
  - Right middle lobe, left ligular
  - Total left lung
  - Total right lung
- Treatment of bronchiectasis
  - Medical
    - Prevention and Control
    - Antibiotics
    - Postural drainage
  - Surgical
    - Unilateral, segmental, or lobar distribution
    - Persistent, recurrent symptoms when medication is discontinued
    - Recurrent infection and hemoptysis
  - Transplantation



# Patient Positions for Postural Drainage

Cardiopulmonary Assessment and Intervention





# Lung abscess

- Definition
  - Necrosis of the pulmonary tissue and formation of cavities containing necrotic debris or fluid caused by microbial infection
- DDX: Empyema
  - Collection of pus in pleural space

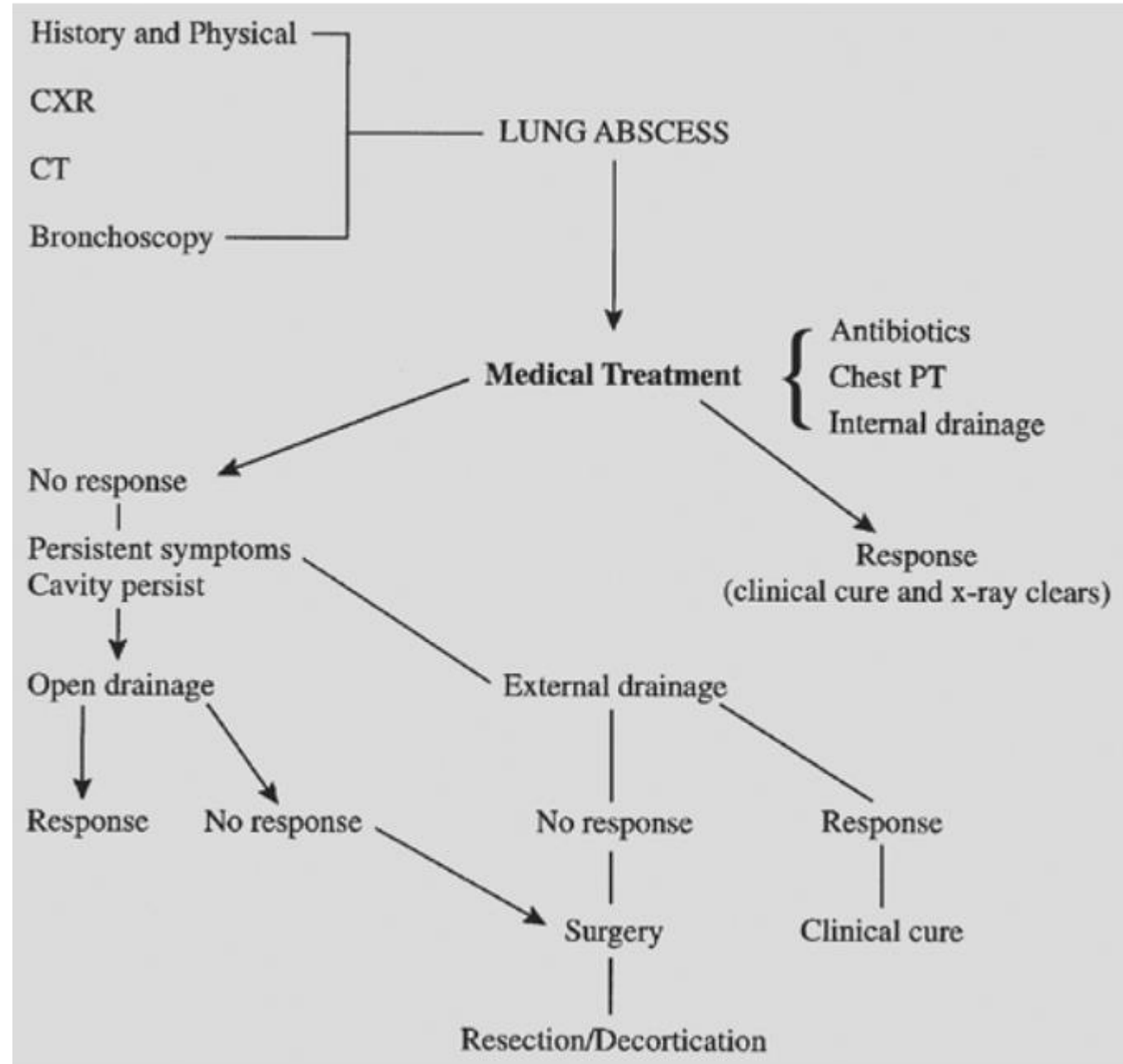
Abscess	Empyema
Intermediately thickened (4–15 mm) walls	Thin walls
Spherical	Lenticular
Surrounded by consolidation	“Split pleura” sign (CT)
Equal-length air-fluid levels on frontal and lateral radiographs	Different-length air fluid levels on frontal and lateral radiographs
Narrow interface with chest wall (CT)	Broad contact with chest wall
Bronchovascular markings extend to abscess	Adjacent compressed lung

### Table 89-9 Contributing Factors to Lung Abscess

- Dental and periodontal disease
- Anesthesia
- Alcohol abuse
- Seizure disorders
- Immunosuppression
- Neuromuscular disorders with bulbar dysfunction
- Esophageal motor disorders
- Bronchial obstruction

### Table 89-10 Bacteriology of Lung Abscess

- Anaerobic
- Bacteroides fragilis*
  - Fusobacterium bacilli*
  - Streptococcus*,  $\beta$ -hemolytic streptococcus
- Aerobic
- Klebsiella pneumoniae*
  - Pseudomonas aeruginosa*
  - Staphylococcus aureus*
  - Streptococcus pneumoniae*
  - Haemophilus influenzae*



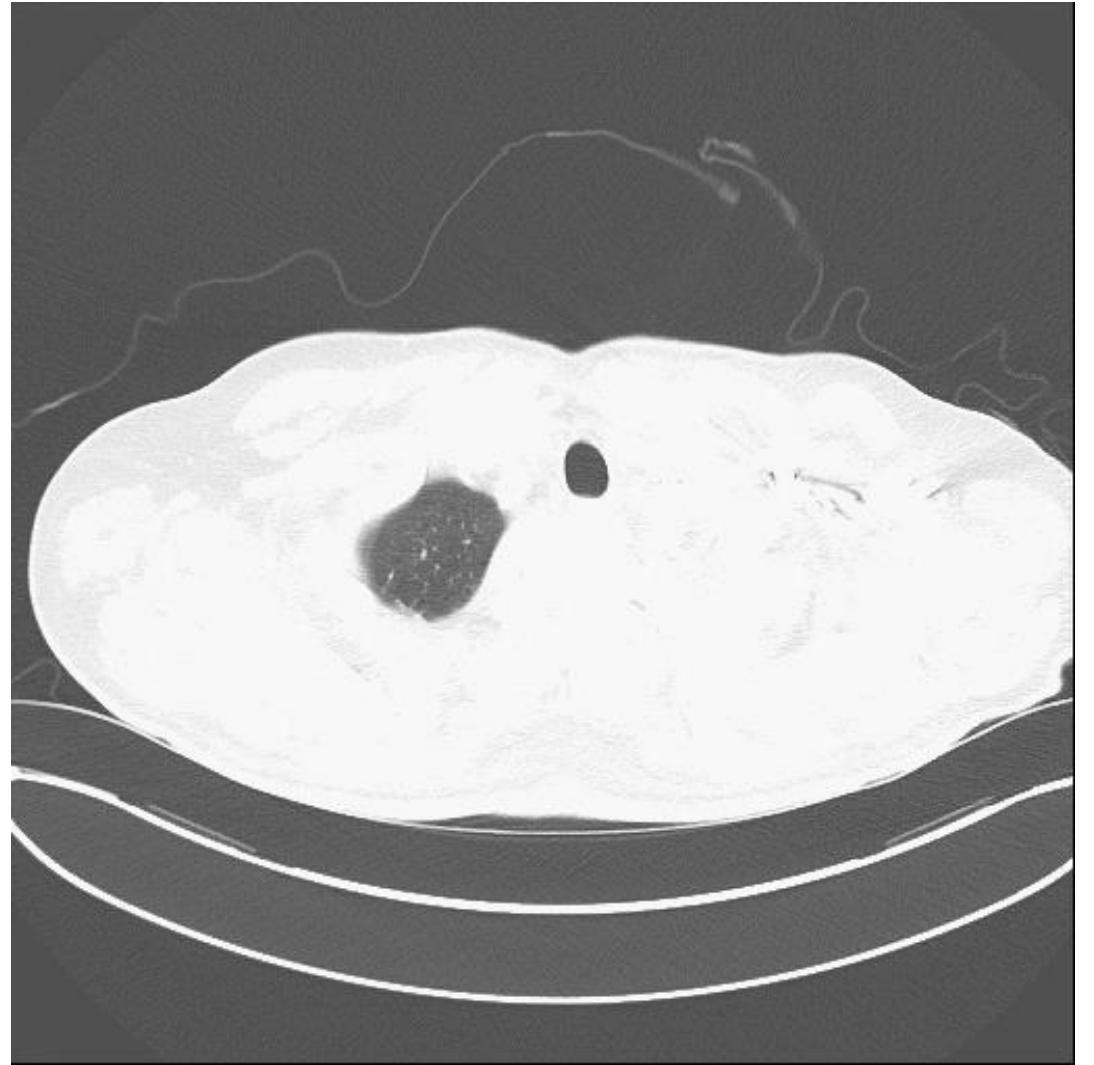
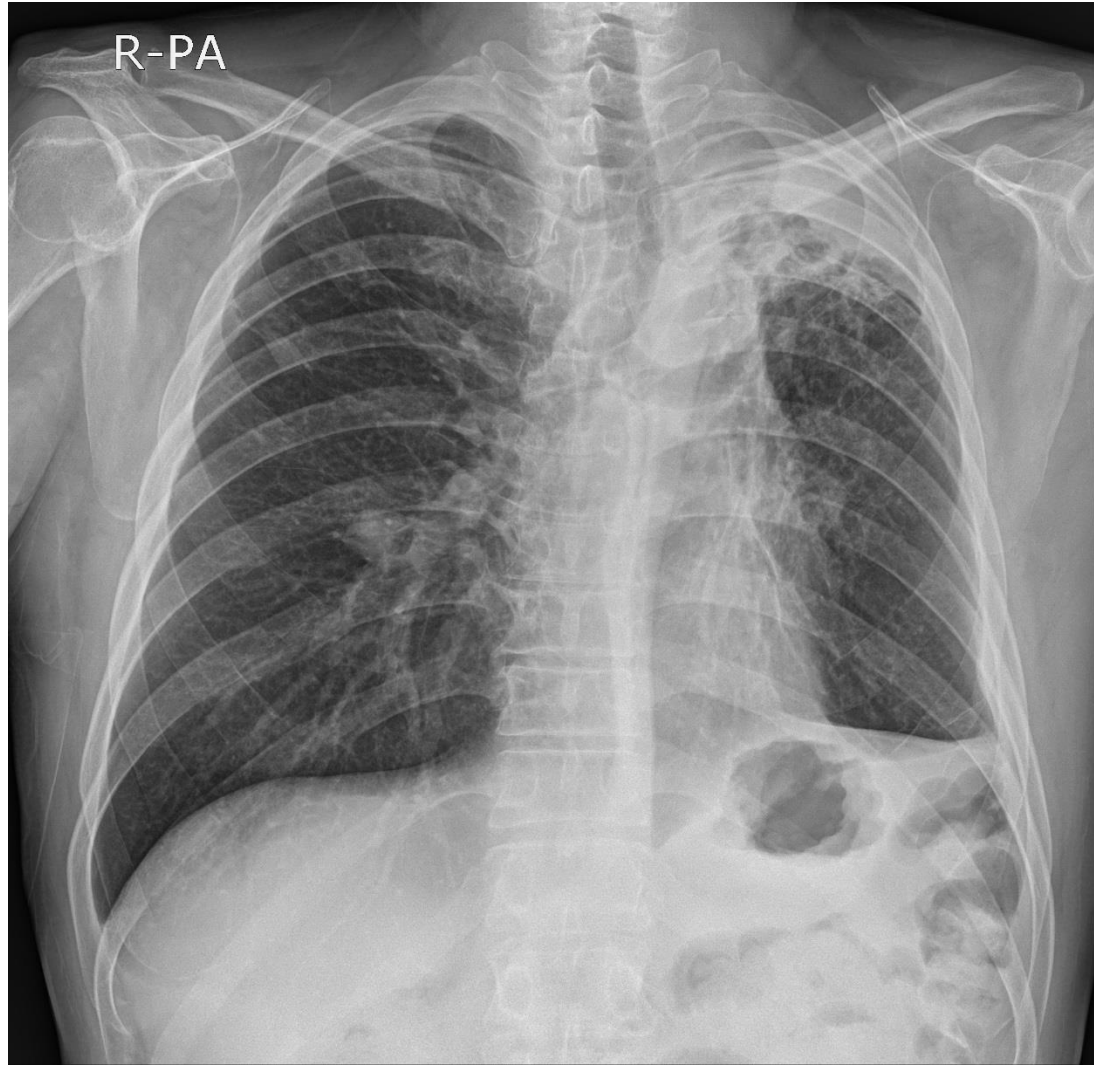
# Pulmonary Tuberculosis

- Classical treatment
  - Collapse therapy (by collapsing cavitory disease, the organisms would be deprived of oxygen and thus die)
- Surgical indications
  - Destroyed lung
  - Cavitory disease, no response in medical therapy
  - Life threatening hemoptysis
  - Bronchial stricture
  - Suspected malignancies
- Frequent complication after operation
  - Empyema
  - Broncho-pleural fistula

# Other Infected Lung Diseases

- Aspergillosis
- Actinomycosis
- Amebiasis
- Candidiasis
- Cryptococcosis
- Paragonimiasis
- Etc.





Rot +8°  
Ang +1°  
FD 48 cm

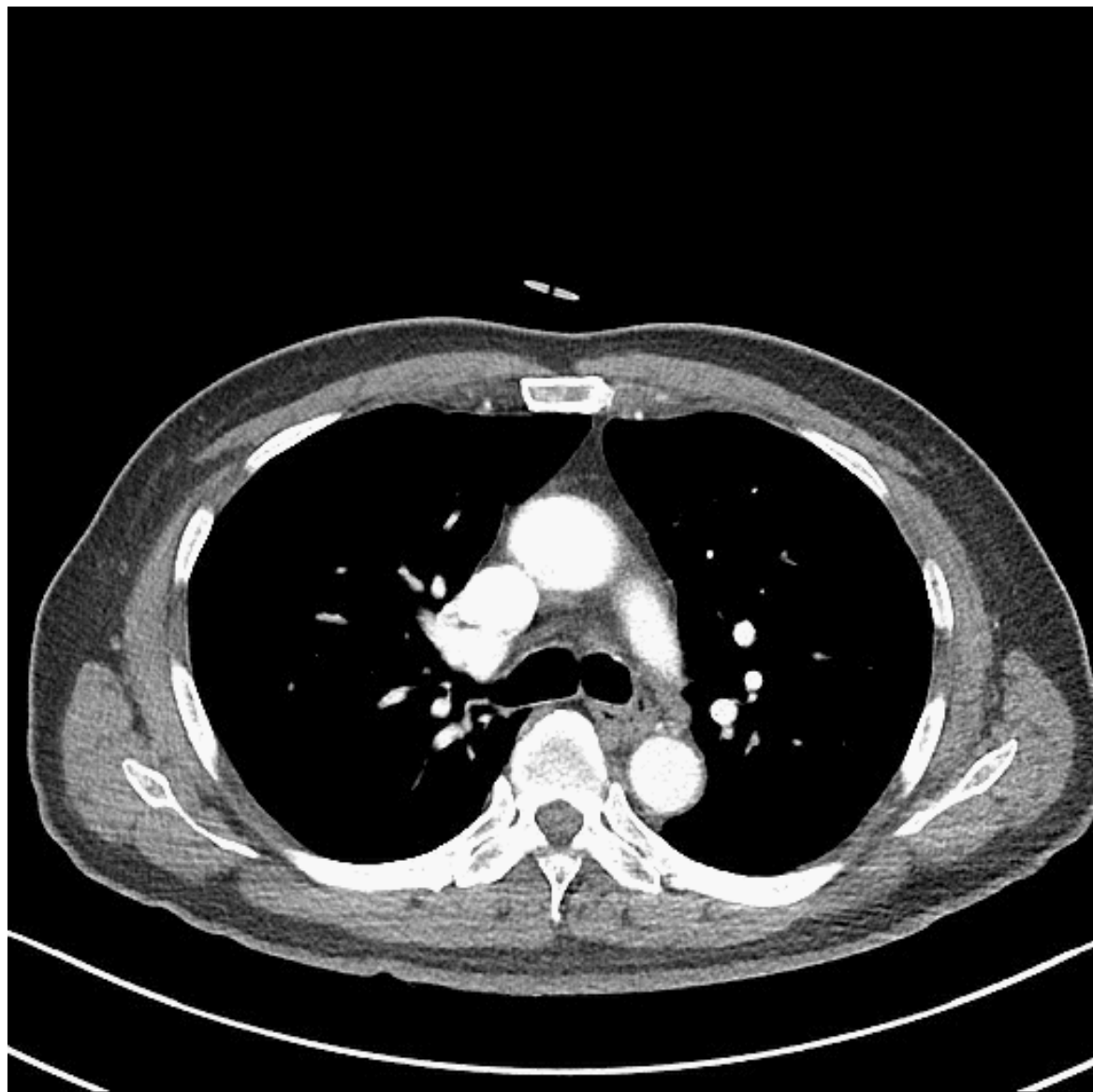
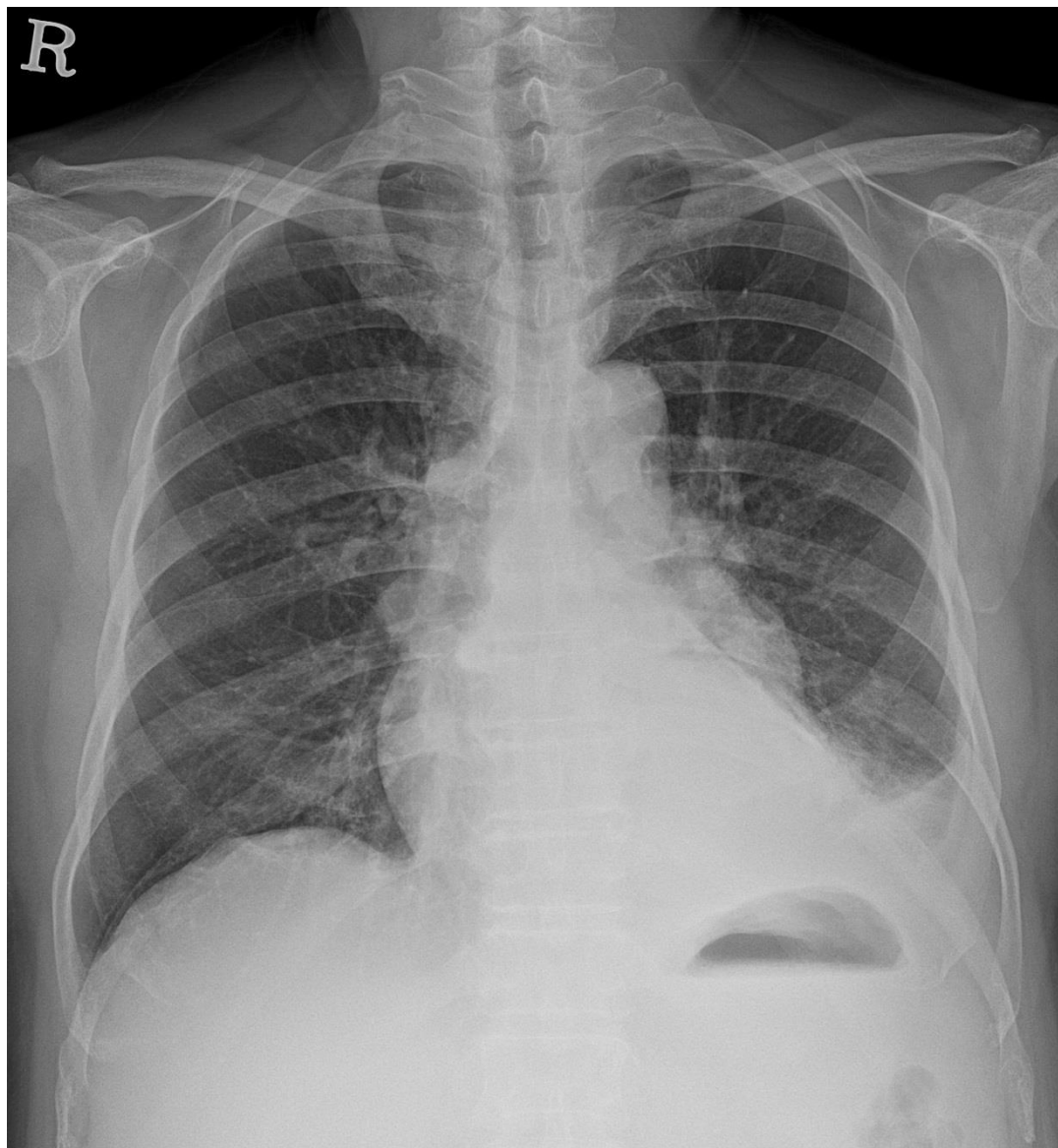


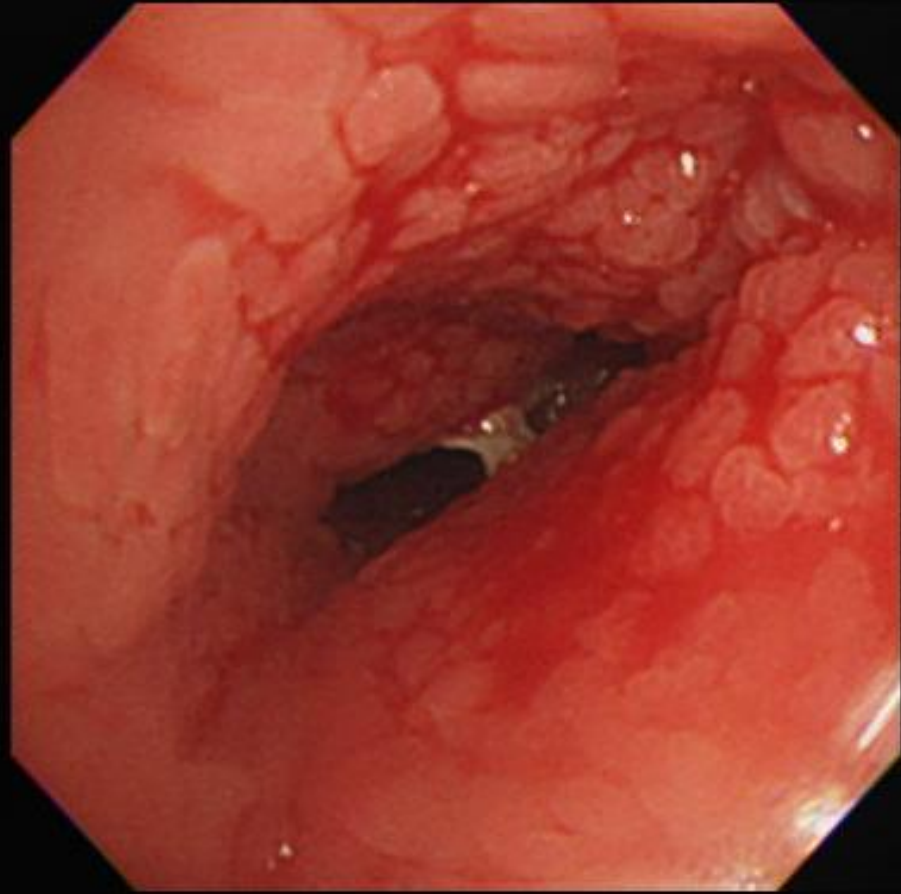
0:00  
3:67  
11:12:58



7  
1-12







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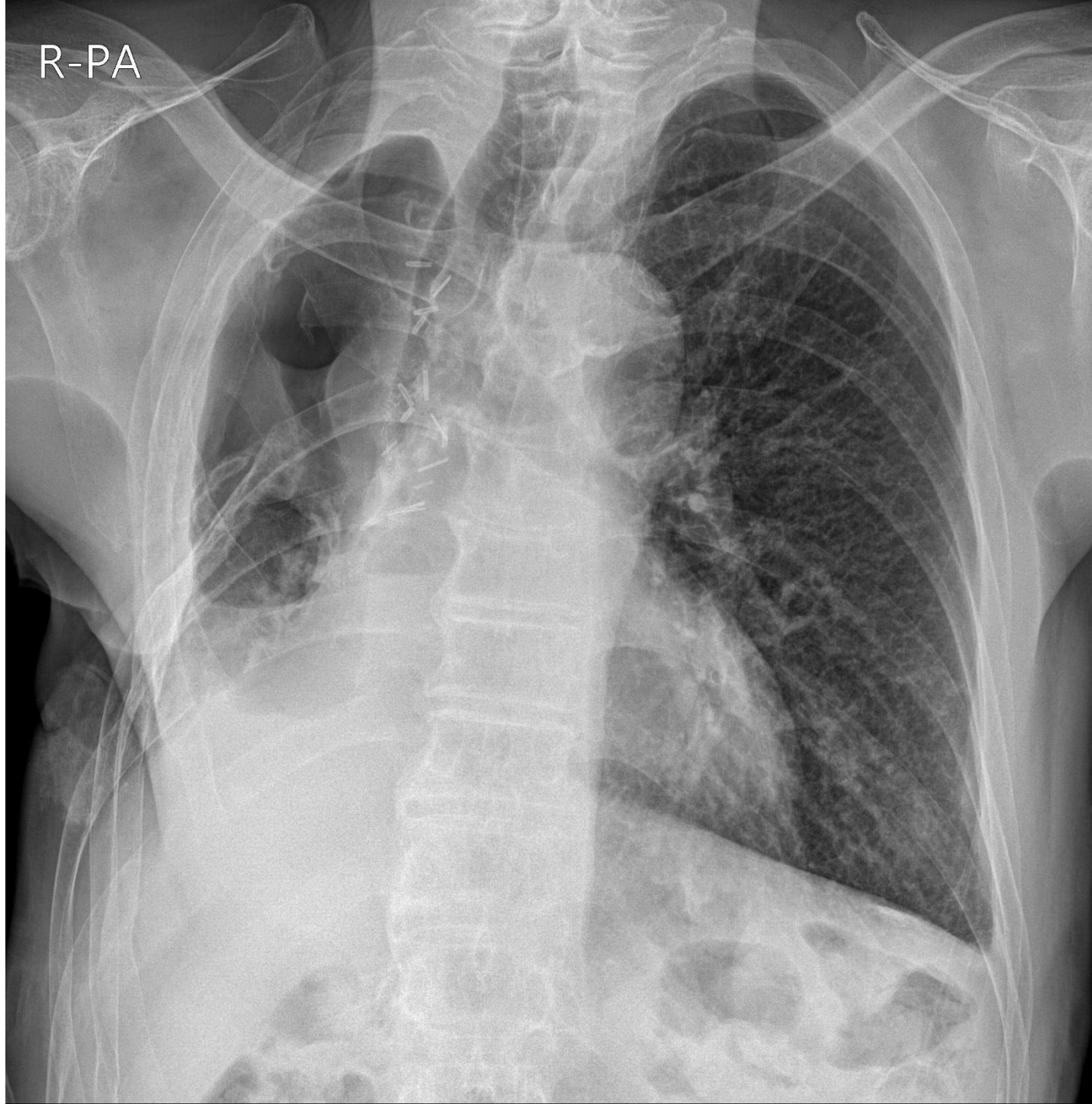




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R-PA





# Solitary Pulmonary Nodule

- Definition
  - Single, spherical, well-circumscribed, radiographic opacity
  - $\leq 3\text{cm}$
  - Surrounded completely by aerated lung
  - Not associated atelectasis, hilar enlargement or pleural effusion

# Solitary Pulmonary Nodule

## Neoplasms

### *Malignant*

Lung cancer

Carcinoid tumor

Metastasis—carcinoma, sarcoma, melanoma, germ cell

Uncommon malignant primary lung tumors—blastoma, carcinosarcoma, lymphoma, melanoma, plasmacytoma, salivary gland—type tumors (adenoid cystic, mucoepidermoid, acinic cell, mixed, oncocytoma), sarcoma, teratoma, thymoma

### *Benign*

Hamartoma

Uncommon benign primary lung tumors—alveolar adenoma, clear cell tumor (sugar tumor), chondroma, Clara cell adenoma, fibroma, fibromyxoma, glomus tumor, granular cell myoblastoma, hibernoma, leiomyoma, lipoma, mucous gland adenoma, neurogenic tumor, sclerosing hemangioma, squamous papilloma, teratoma, thymoma, xanthoma

## Nonneoplastic lesions

Infectious granulomas—histoplasmosis, tuberculosis, coccidioidomycosis, cryptococcosis, blastomycosis, aspergillosis, nocardiosis

Other—abscess, arteriovenous malformation, bronchogenic cyst, pulmonary infarction, intrapulmonary lymph node, organizing pneumonia, parasitic lesions (echinococcus, ascaris, dirofilaria), plasma cell granuloma (inflammatory pseudotumor), postinflammatory fibrosis, rounded atelectasis, sequestration, venous varix

### **Lesions that are usually multiple but may be solitary**

Amyloid nodule, bronchiolitis obliterans—organizing pneumonia (BOOP), endometriosis, eosinophilic granuloma, mucoid impaction, pulmonary hyalinizing granuloma, rheumatoid nodule, sarcoidosis, septic embolus, silicosis, Wegener's granulomatosis

### **Extrapulmonary densities mistaken for solitary pulmonary nodules by plain film**

Blood vessel (dilated or on-end view), bone island, chest wall soft tissue mass, extracorporeal density (e.g., electrocardiographic electrode), overlapping normal structures, nipple shadow, osteophyte, pleural plaque or mass, pseudotumor (fluid in interlobar fissure)

# Harmatoma

- Definition
  - Benign, slow growing or self limiting
  - Abnormal mixture of normal tissues and cells from the area in which it grows
  - Composed of cartilage, connective tissue, muscle, fat, and bone

