

Congenital and Benign Lung diseases

박성용

연세대학교 의과대학 흉부외과학교실

Congenital and Benign lung disease

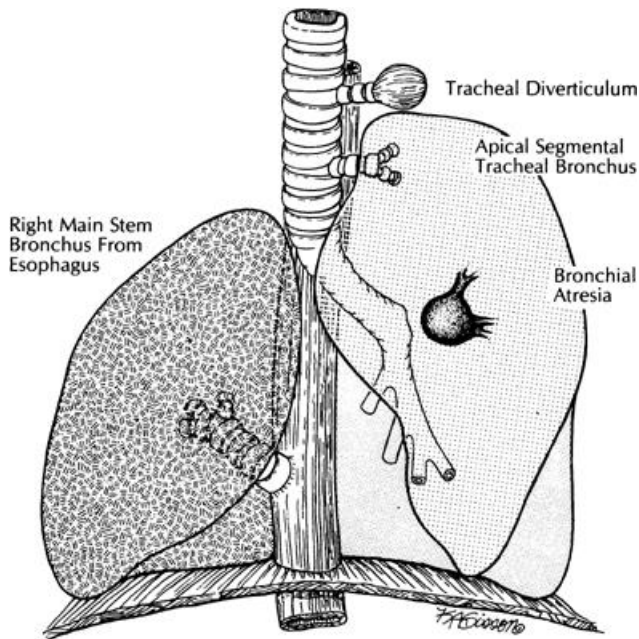
- **Malignant lung diseases**
- **Benign and congenital lung diseases**
 - Congenital lesions of lung and pulmonary vasculature
 - Bullous lung disease
 - Infectious lung disease

Congenital lesions of lung

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

Bronchial anomalies

- Tracheal bronchus and diverticulum



- Bronchial atresia
 - Progressive respiratory symptom due to mucocele
- Anomalous bronchi
 - Common from esophagus

Congenital lobar emphysema



- The isolated hyperinflation of a lobe in the absence of extrinsic bronchial obstruction
- Frequent lobe; LUL, RML
- Severe pulmonary distress since infant

- Tx; resection (lobectomy)

Pulmonary sequestration

■ Definition

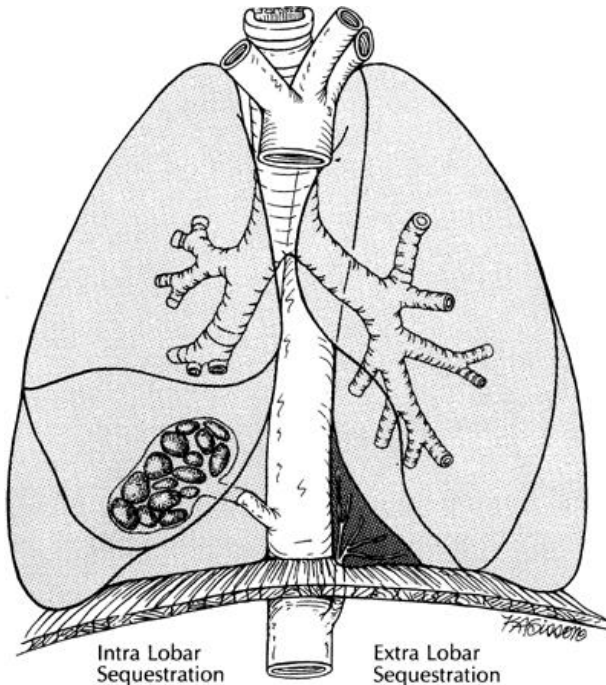
- a segment or lobe of lung tissue that has no bronchial communication with the normal tracheobronchial tree
- The arterial blood supply; from a systemic vessel
- The venous return; usually through the pulmonary veins, sometimes the systemic venous system

■ Extralobar Sequestration

- Separated from normal lung by visceral pleura
- 25% of sequestration
- Left chest; 90%, posterior CPA angle

■ Intralobar Sequestration

- Situated within the normal lung parenchyme
- Communication through the pores of Kohn may lead to chronic infection in the sequestered lobe
- Symptoms; hemoptysis, false aneurysm



Congenital cystic adenomatoid malformation



- **Definition**
 - A spectrum of cystic and solid lesions of the lung can be identified histologically as CCAMs
 - Type I ~ III
- **Histology**
 - An adenomatoid increase of terminal respiratory bronchiole-like structures lined by ciliated columnar epithelium occurs. Interspersed cysts may resemble immature alveoli
 - The mucosa of cysts lined with bronchial-type epithelium may show polypoid overgrowth projecting into the lumen of the cysts
 - Bronchial mucoserous glands and cartilaginous plates are absent throughout the cystic parenchyma

Congenital cystic adenomatoid malformation

■ **Symptoms**

- Severe respiratory distress due to space occupying lesions
 - Recurrent pneumonia
 - Several reports of malignancy arising in CCAM; bronchoalveolar carcinoma, pleuropulmonary blastoma
-
- **Timing of operation?**

Congenital vascular lesions of lung

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

Pulmonary arteriovenous fistula



- Congenital malformations that result from errant capillary development, with incomplete formation or disintegration of the vascular septa that normally divide the primitive connections between the venous and arterial plexuses
- Classification
 - I; Multiple small arteriovenous fistulae without aneurysm
 - II; Large single arteriovenous aneurysm, peripheral
 - IIIa; Large single arteriovenous aneurysm, central
 - IIIb; Large arteriovenous aneurysm with anomalous venous drainage
 - IIIc; Multiple small arteriovenous fistulae with anomalous venous drainage
 - Iva; Large single venous aneurysm with systemic artery communication
 - Ivb; Large single venous aneurysm without fistula, varix of pulmonary vein
 - V; Anomalous venous drainage without fistula

Pulmonary arteriovenous fistula

■ Symptoms

- Decreased pO₂ (shunt effect)
- Stroke
- Cerebral abscess
- Seizure

■ Treatment

- Coil obliteration (10-15mm diameter)
- Surgical resection

Bullous and Bleb Diseases of the Lung

- **Bullae; emphysematous spaces larger than 1 cm in diameter in the inflated lung, usually but not necessarily demarcated from surrounding lung by curved hairline shadows**
- **Blebs; well-circumscribed intrapleural air spaces separated from the underlying parenchyma by a thin pleural covering**
- **Group I; Bullae and Almost Normal Underlying Lung**
- **Group II; Bullae Associated with Diffuse Emphysema**

Rationale and Indications for Surgery

- **Pneumothorax**
 - Prolonged air leak
 - High incidence of recurrences (>50%)
- **Infection of the bulla (uncommon); Failure to respond to medical treatment**
- **Hemoptysis (uncommon)**
- **Chest pain; Pain clearly related to air trapping during hyperventilation**
- **Treatment of lung cancer**
 - Documented cancer or highly suspicious lesion

- **Expansion of previously collapsed lung**
 - Increase in vital capacity and forced expiratory volume in 1 second
 - Improvement in gas exchange (higher ventilation-perfusion ratio and arterial Po₂)
- **Hemodynamic improvement**
 - Increase in cardiac output; better exercise tolerance
- **Restoration of normal curve of diaphragm**
 - Improvement in diaphragmatic contractility and function
- **Restoration of elastic recoil and reduction in airway resistance**
 - Bullae increase the loss of elasticity in the emphysematous lung
 - Loss of elastic recoil causes an extrinsic airway obstruction
- **Removal of an area of dead space ventilation**
 - Reduction in volume of wasted ventilation
 - Decrease in work of breathing

Selection of patients for surgery

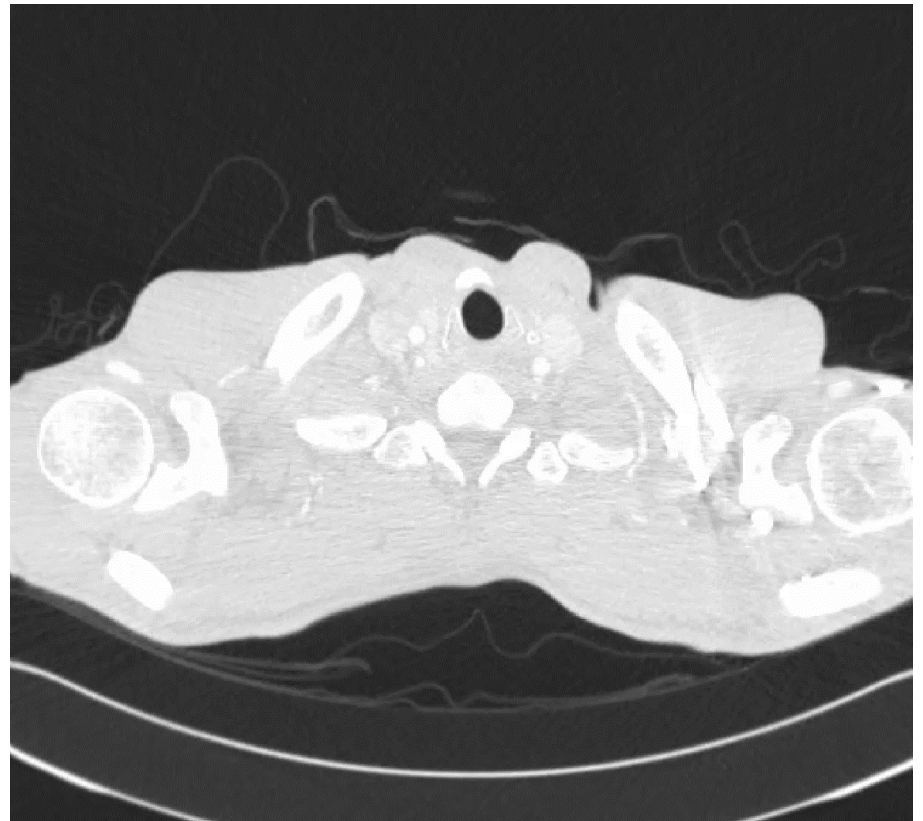
- Is there a localized or enlarging bulla, or both?
- Is the bulla nonfunctional and does it compress adjacent lung, mediastinum, diaphragm?
- Can the compressed lung reexpand and what is its potential to function once reexpanded?
- What is the extent and severity of emphysema in the remaining lung?
- What is the cardiac performance?
- Does the patient have significant associated comorbidities, weight loss, or both?
- Can the patient withstand an operation?

Selection of patients for surgery

Area of Investigation	Technique	Most Suitable for Surgery	Least Suitable for Surgery
Anatomy of bullae	Standard radiographs, CT scan	Large (more than half hemithorax) localized and unilateral bullae Enlargement over time	Multiple, small bilateral bullae No enlargement over time
Function of bullae	V/Q scans, plethysmography	Nonventilated, nonperfused bullae	Ventilated and perfused bullae
Compression index	Standard radiographs, CT scan angiography	High index ($\geq 3/6$)	Low index ($< 3/6$)
State of compressed lung	Angiography, V/Q scans, CT scan	Good capillary filling Good washout of xenon	Poor capillary filling Retention of xenon
Severity of emphysema	CT scan, pulmonary function tests, exercise tests	Minimal or no COPD	Severe COPD Respiratory failure
Medical status	Clinical examination, EKG, echocardiography, nutritional evaluation	Young age Normal heart No comorbidities No weight loss	Older age Cor pulmonale Significant comorbidities Significant weight loss

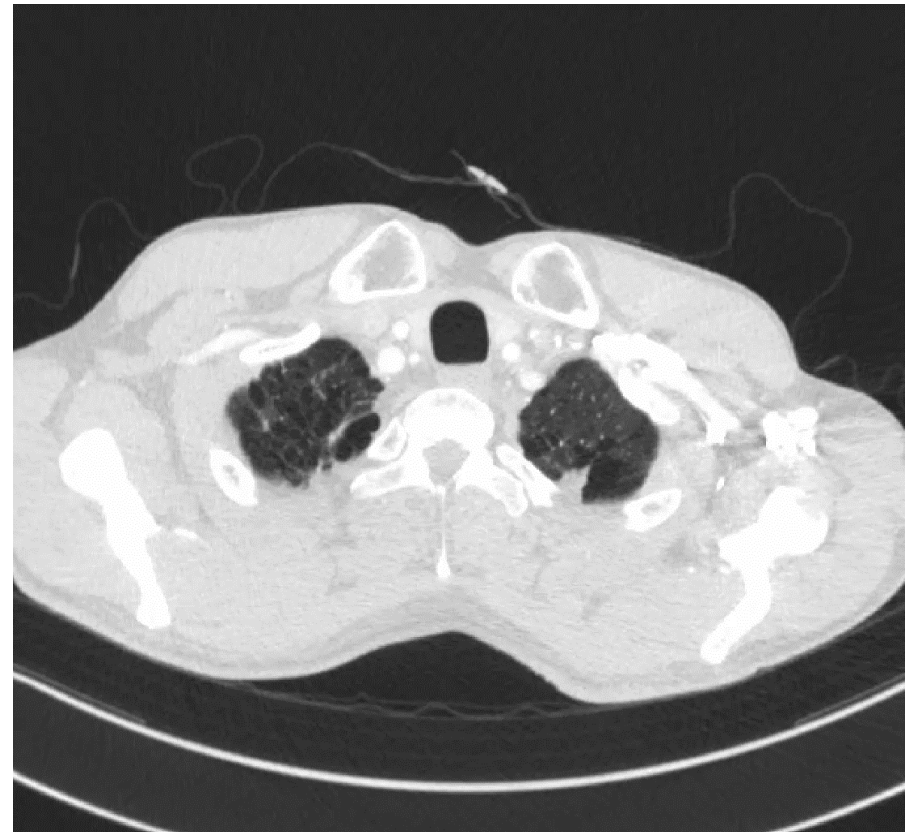
Case; M/59

- **Diagnosis; Emphysema**
- **Operation; Wedge resection of RUL under VATS (2013)**
- **FEV1 1.05 (35%) → 2.32 (66%)**



Case; M/59

- 5 years later,
- FEV1 1.05 (35%) → 2.32 (66%) → 0.84 (29%)



Bacterial Infections of the Lung and Bronchial Compression

- **Spectrum of surgical infectious disease**
 - Bronchiectasis
 - Lung abscess
 - Organizing pneumonia
 - Pulmonary infection in granulomatous disease of childhood
 - Tuberculosis and fungal disease
 - Thoracic empyema

- **Bronchial compressive pulmonary disorders**
 - Right middle lobe syndrome
 - Broncholithiasis
 - Inflammatory lymphadenopathy
 - Congenital processes
 - Sclerosing mediastinitis
 - Cardiovascular disease
 - Congenital
 - Vascular ring
 - Aberrant left pulmonary artery
 - Acquired aortic disease
 - Aortic arch aneurysm
 - Traumatic false aneurysm

Bronchiectasis



- **Definition; abnormal permanent dilatation of subsegmental airways**

- **Treatment**
 - **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**

Lung abscess

- **Principles of Therapy for Lung Abscess**
 - Identification of etiologic organism
 - Prolonged antimicrobial therapy
 - Adequate drainage in acute stage
 - Chest physiotherapy
 - Bronchoscopy
 - Percutaneous catheter drainage
 - Emergency surgical treatment
 - External drainage (only in emergent situation)

- **Indications for Surgery in Lung Abscess**
 - **Acute stage (emergency); Complications**
 - Bronchopleural fistula
 - Empyema
 - Bleeding
 - **Chronic stage (definitive)**
 - Persistent symptoms and signs
 - Recurrent complications (empyema, bronchopleural fistula)
 - Suspicion of carcinoma
 - Persistence of lung abscess larger than 6 cm after 8 weeks of treatment

Surgery for Tuberculosis and NTM

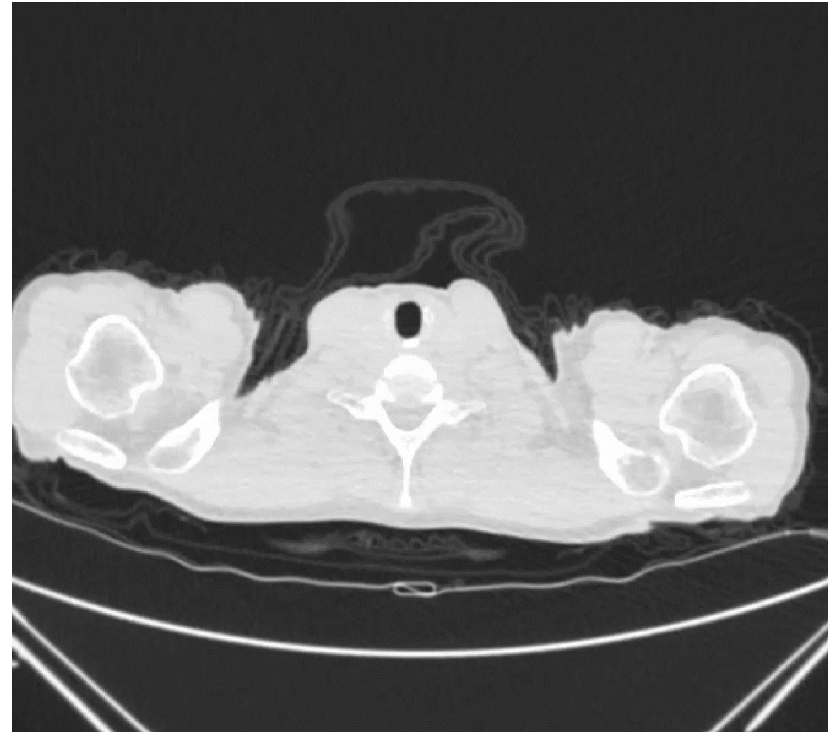
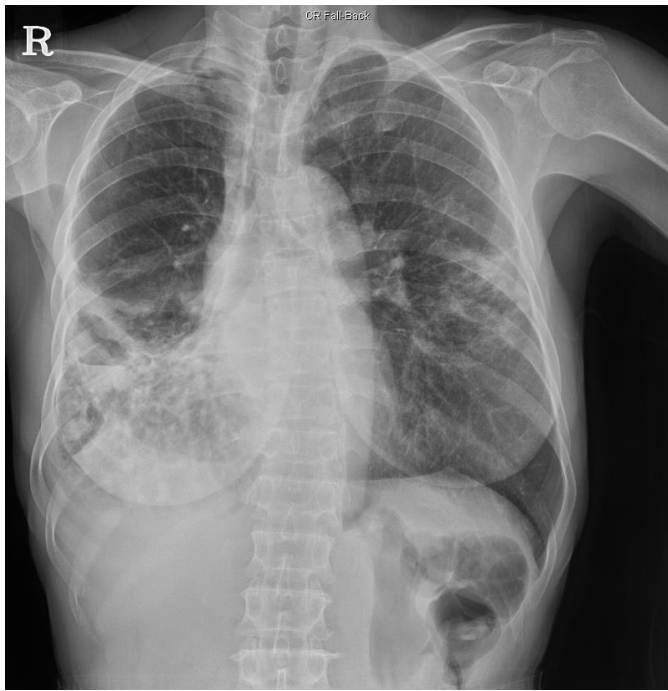
- **Classical treatment**
 - Collapse therapy (by collapsing cavitory disease, the organisms would be deprived of oxygen and thus die)
- **Current surgical indications**
 - Destroyed lung by MDR TB
 - Cavitory disease
 - Life-Threatening Hemoptysis in Patients with Tuberculosis
 - Tuberculous Bronchial Stricture
 - Suspected malignancies
- **Frequent complications after operation**
 - Empyema
 - Broncho-pleural fistula

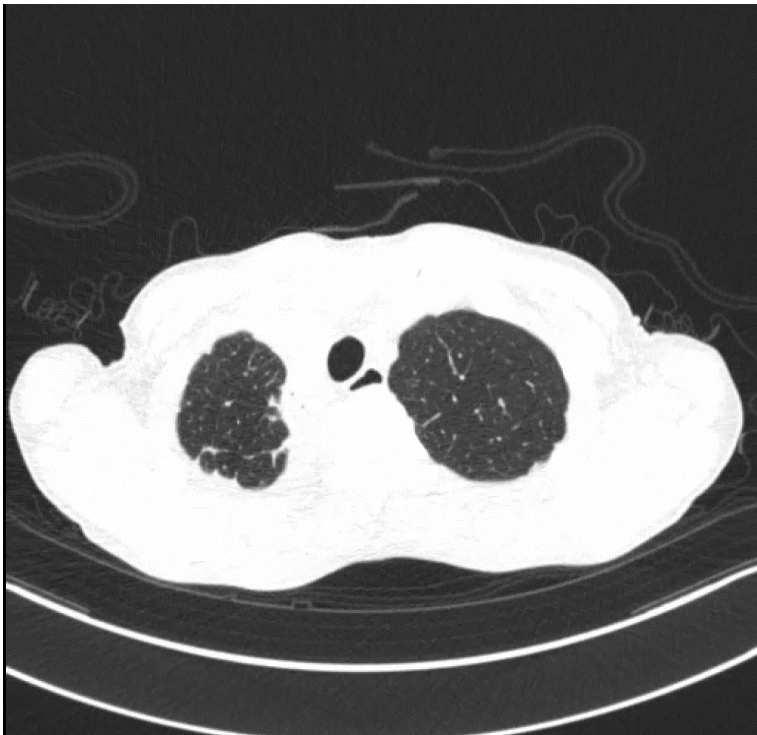
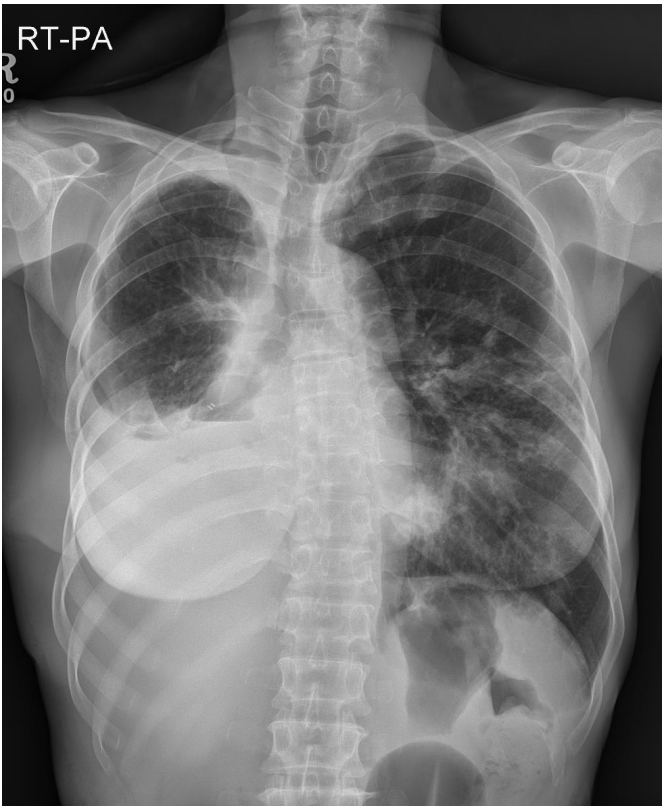
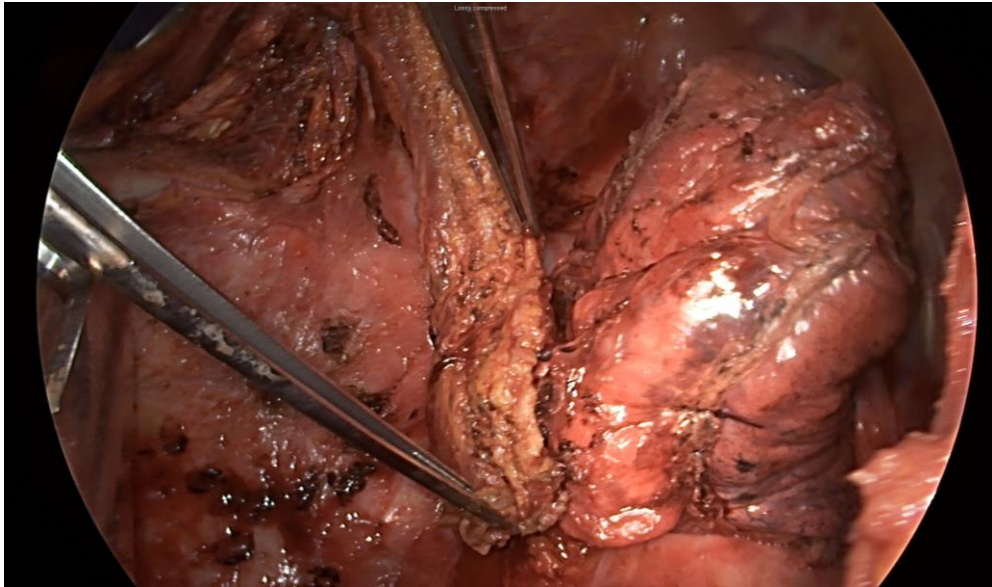
Technical tips for infectious lung disease

- Proper antibiotics during peri-operative periods
- Cover the bronchial stump! (to prevent BPF)

Case; F/59

- **Diagnosis; Drug resistant - NTM infection**
- **Operation; RLL lobectomy, wedge resection of RML, intercostal muscle flap**





Mycotic and Actinomycotic Infections

- **Aspergillosis**
- **Blastomycosis**
- **Candidiasis**
- **Coccidioidomycosis**
- **Cryptococcosis**
- **Histoplasmosis**
- **Mucormycosis**
- **Paracoccidioidomycosis**
- **Pseudallescheriasis**
- **Sporotrichosis**

- **Actinomycosis**
- **Nocardiosis**