

Cystic Fibrosis Complications

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 INTRODUCTION 

 PNEUMOTHORAX

 HEMOPTYSIS

 RESPIRATORY FAILURE

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Cystic Fibrosis

✚ Autosomal Recessive

✚ Genetically transmitted disorder affecting 30,000 individuals in US and 60,000 worldwide

✚ US incidence: 1 per 1900-3700 Caucasians

✚ Previously a pediatric disease

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 1990: 30% patients in US CFF Registry older than 18yrs

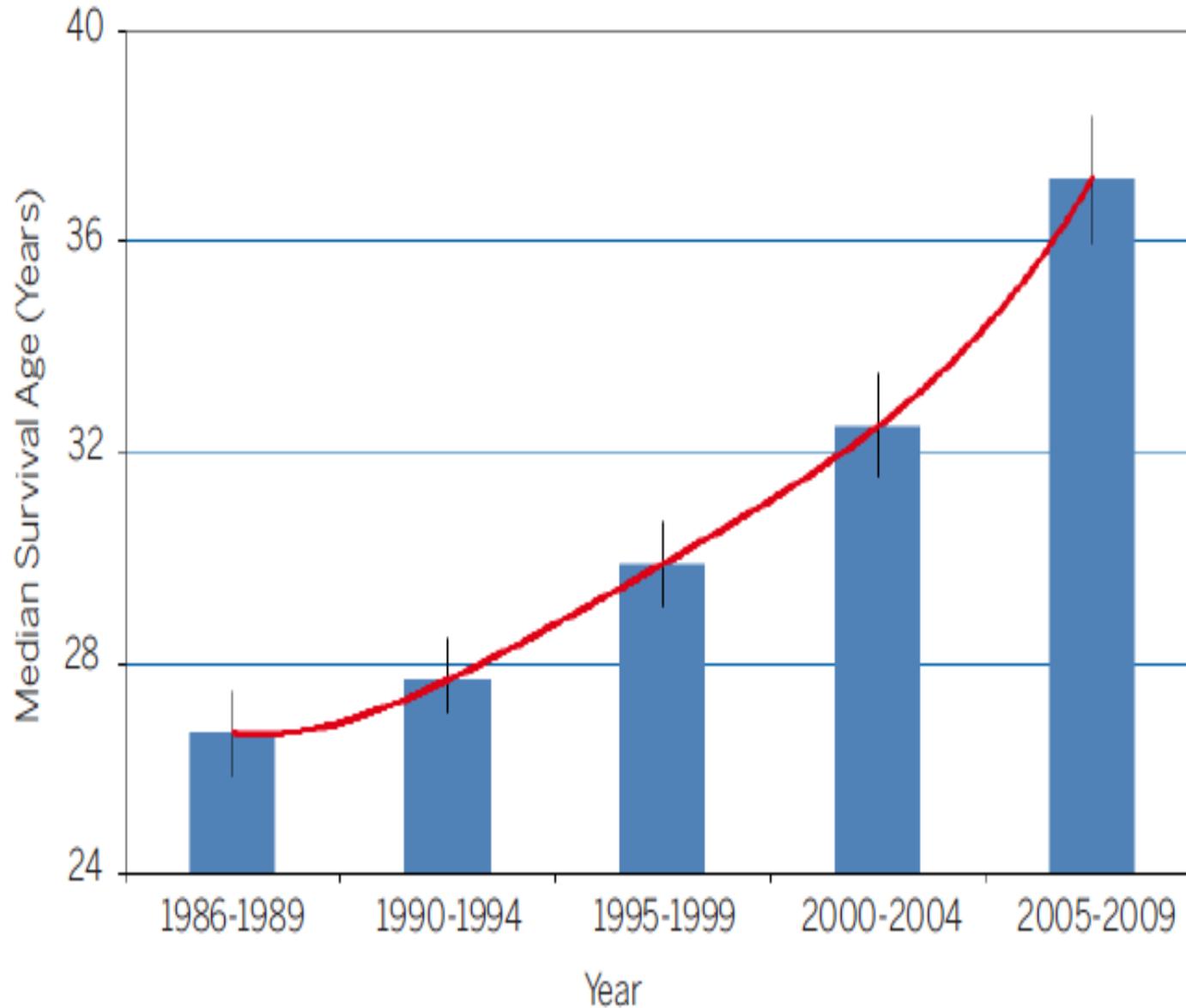
 2012: More than 45% older than 18yrs.

 Assuming improving care and therapies, the projected median survival is approximately 50 years of age for those born after 2000.

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Median Predicted Survival Age, 1986-2009
Over 5 Year Bands



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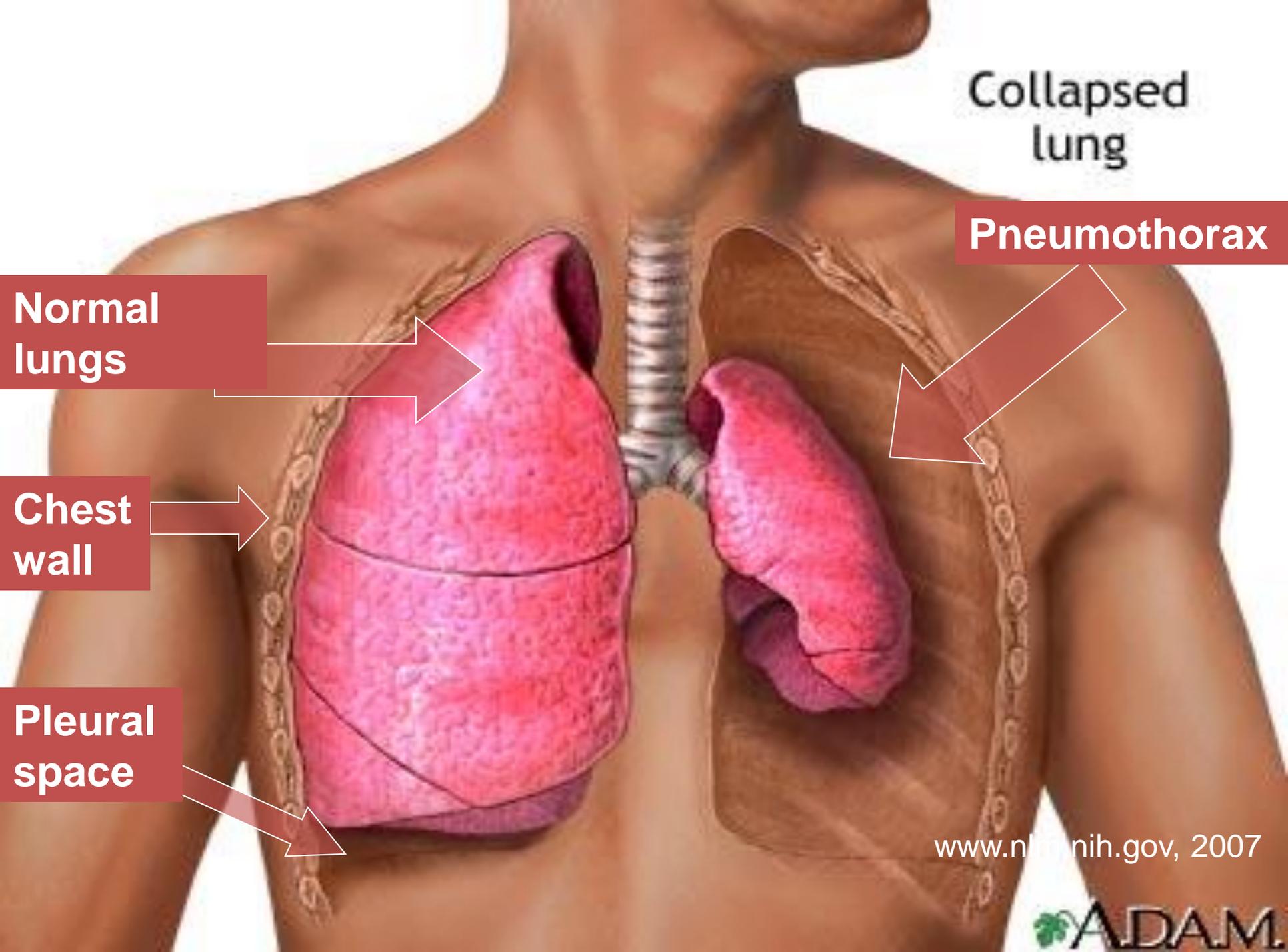
 RESPIRATORY FAILURE

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What is a pneumothorax (PTx)?

- 1803: First description in patients with pulmonary tuberculosis
- Accumulation of air in the pleural space defined as the space between the lungs and chest wall
- Primary Pneumothorax:**
 - Spontaneous occurrence in patients with normal lungs
- Secondary Pneumothorax:**
 - Spontaneous occurrence in patients with *abnormal* lungs i.e Emphysema, Cystic Fibrosis



Collapsed lung

Pneumothorax

Normal lungs

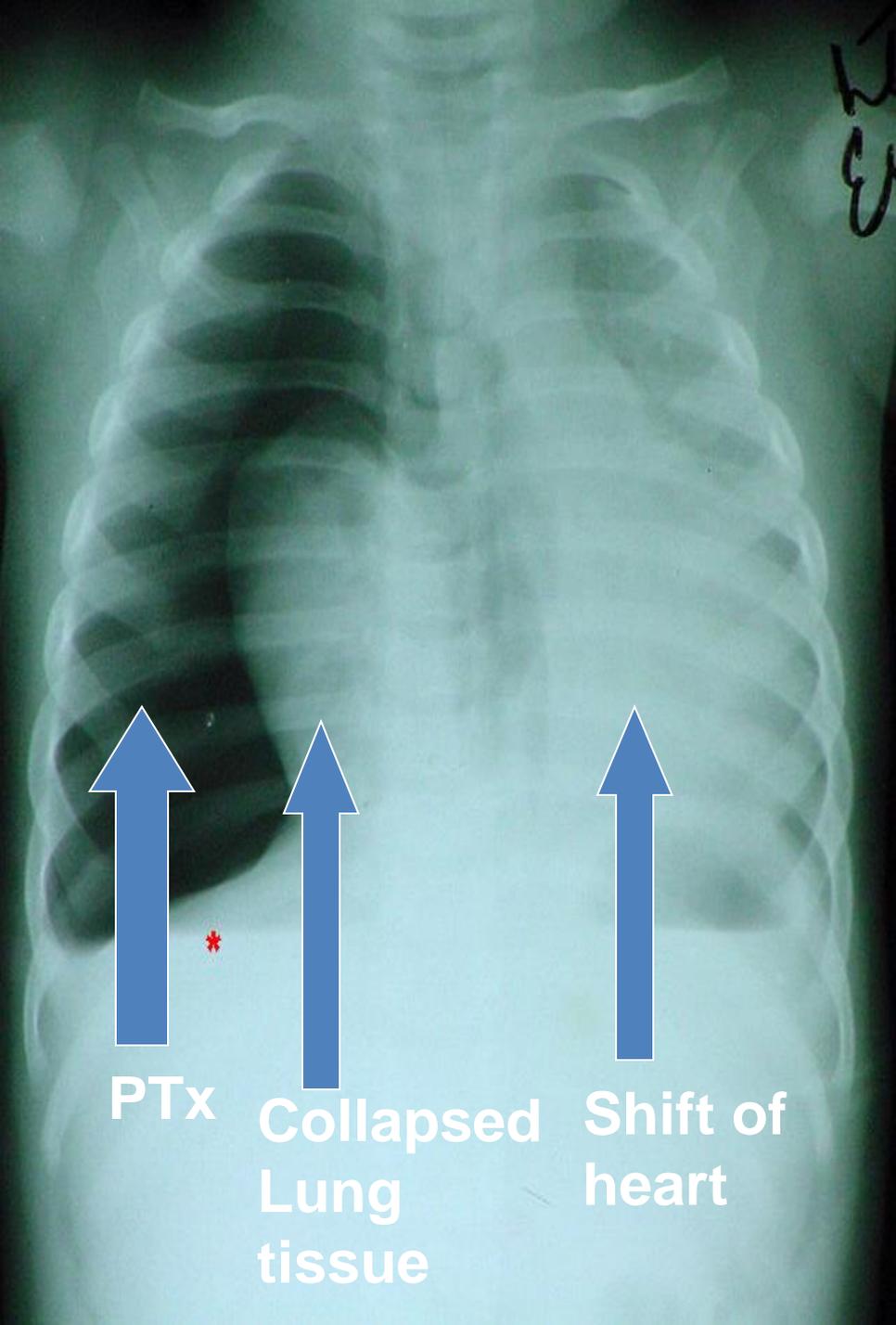
Chest wall

Pleural space

www.nlm.nih.gov, 2007



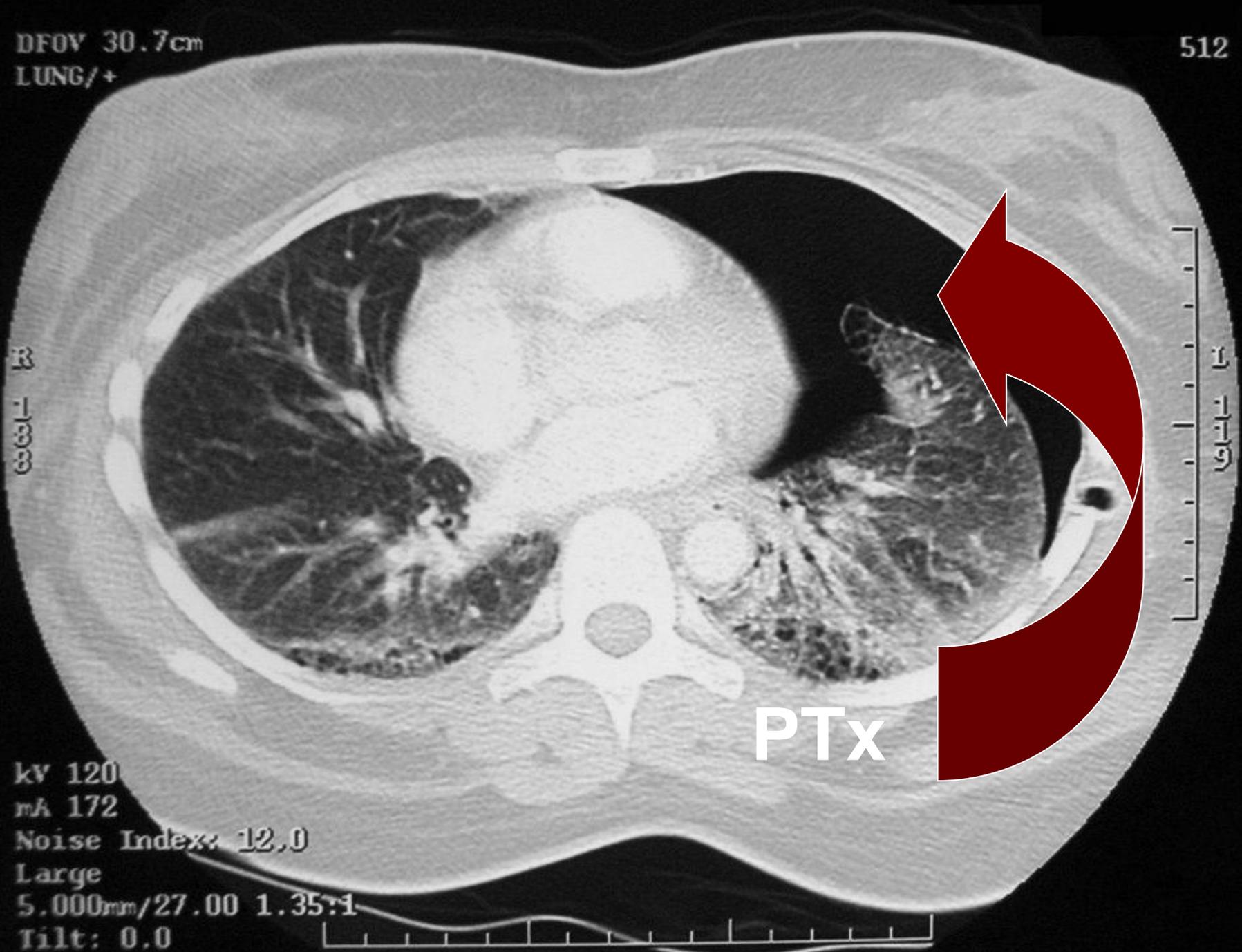
Normal CXR



PTx
Collapsed Lung tissue
Shift of heart

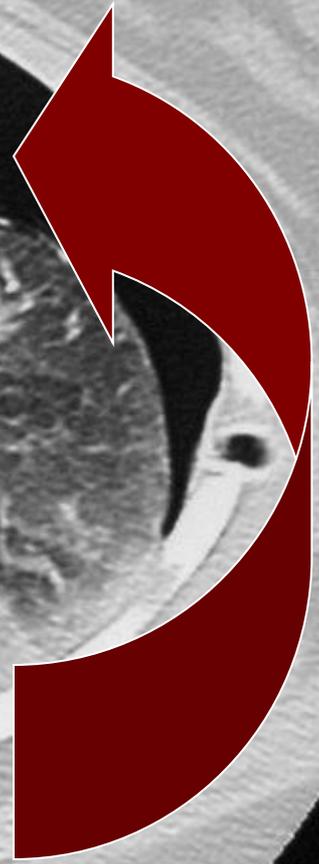
DFOV 30.7cm
LUNG/+

512



kV 120
mA 172
Noise Index: 12.0
Large
5.000mm/27.00 1.35:1
Tilt: 0.0

PTx



What causes a Pneumothorax?

- ✚ May be related to chronic inflammation of the airways
- ✚ Destruction and rupture of small lung units allowing air to leave the lung into the pleural space

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Signs and symptoms

- ✚ Asymptomatic (incidental finding on routine CXR or CT)
- ✚ Various degrees of shortness of breath
- ✚ Chest pain or pressure
- ✚ Normal to decreased oxygen saturation
- ✚ Increased heart rate
- ✚ Low blood pressure
- ✚ Respiratory failure
- ✚ Cardiac arrest

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Pneumothorax

- Commonly reported in CF – 1 in 167 patients each year
- CFF registry 1990-1999
 - 3.4% of total population experienced at least one pneumothorax
 - 82% with one event
 - 18% with greater than one event
 - 72.4% patients with first PTx are > 18 old
 - Average age of 1st PTx 21.9 +/- 9.1 yrs
 - Average age of PTx did not change over the years

Risk Factors for PTx

- ✚ Pseudomonas aeruginosa
- ✚ Burkholderia cepacia
- ✚ Aspergillus / ABPA
- ✚ $FEV_1 \leq 30\%$ predicted
- ✚ Pancreatic insufficiency
- ✚ Tube feeds
- ✚ Medicaid

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Treatment of Pneumothorax

Asymptomatic: PTx < 2 cm

🏠 Admit, observation, repeat CXR(S)

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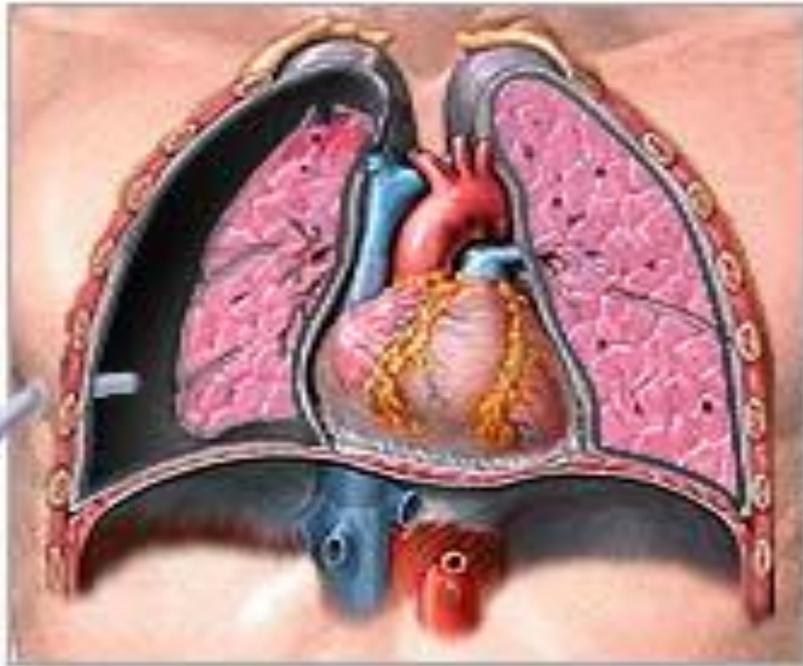


Mildly symptomatic: PTx < 2 cm and <50y
Needle aspiration

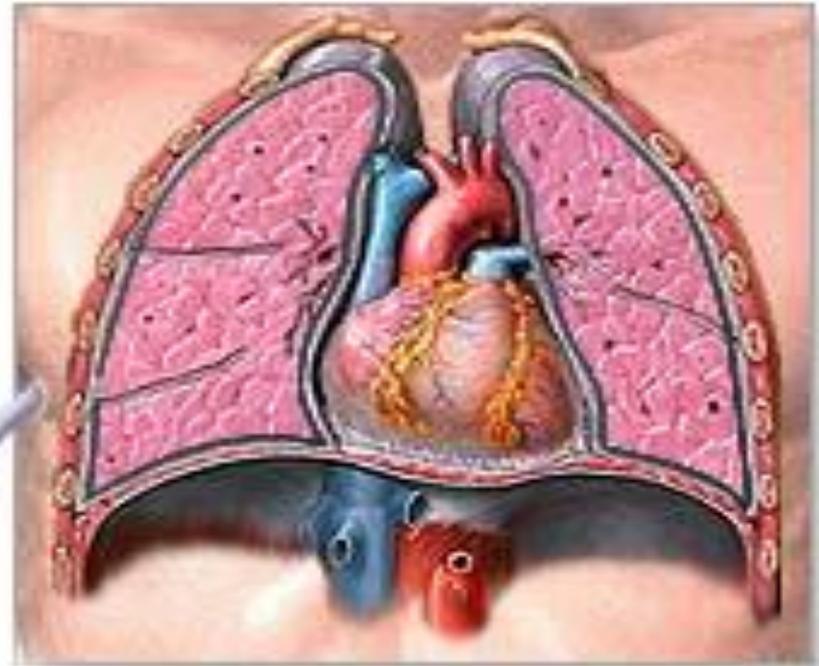


Symptomatic: PTx > 2 cm, and >50y

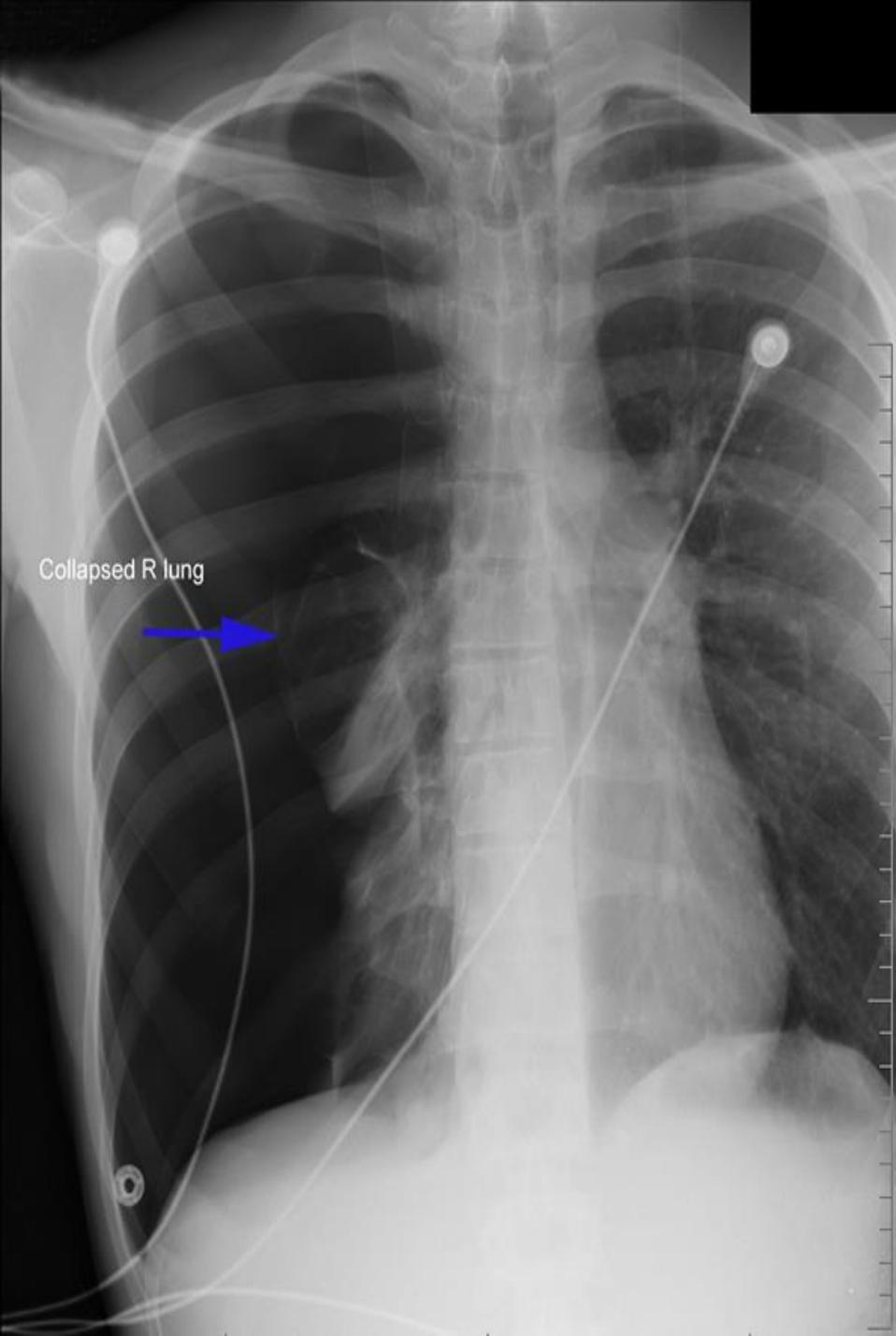
Chest tube



Pneumothorax



Re-expanded lung



Persistent Pneumothorax

- ✚ Consultation with thoracic surgeon
- ✚ Caution in thoracic surgical procedures as they may make lung transplantation surgery complicated

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Prognosis

Mortality after pneumothorax

 48.6% at 2 years

 75% at 8 years

***** Higher mortality associated with lower FEV₁**

***** ?? Early referral for Lung Transplantation despite
FEV₁ ≥ 30% predicted**

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What is hemoptysis?

✚ Expectoration of blood from the respiratory tract

✚ **Massive Hemoptysis:**

✚ Coughing greater than 250ml of blood over 24 hours or recurrent bleeding with >100 ml/ day over several days

• **Non-Massive Hemoptysis:**

– Any quantity of blood less than mentioned above

Most common cause of Hemoptysis in CF patients

- ✚ Chronic airway inflammation affects adjacent arterial supply
- ✚ Tortuous vessels
- ✚ Hypervascularity
- ✚ Weakened vessel walls

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Signs and symptoms

- ✚ Expectoration of blood
- ✚ Increasing shortness of breath
- ✚ Increasing heart rate
- ✚ Massive blood loss leading to low blood pressure
- ✚ Respiratory failure
- ✚ Cardiac arrest

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Hemoptysis

- 4.1% of CF patients experience at least one episode of massive hemoptysis
 - 74% will only have one episode
 - 26% with greater than one episode
- Majority (75%) of patients with first episode occurring after age 18

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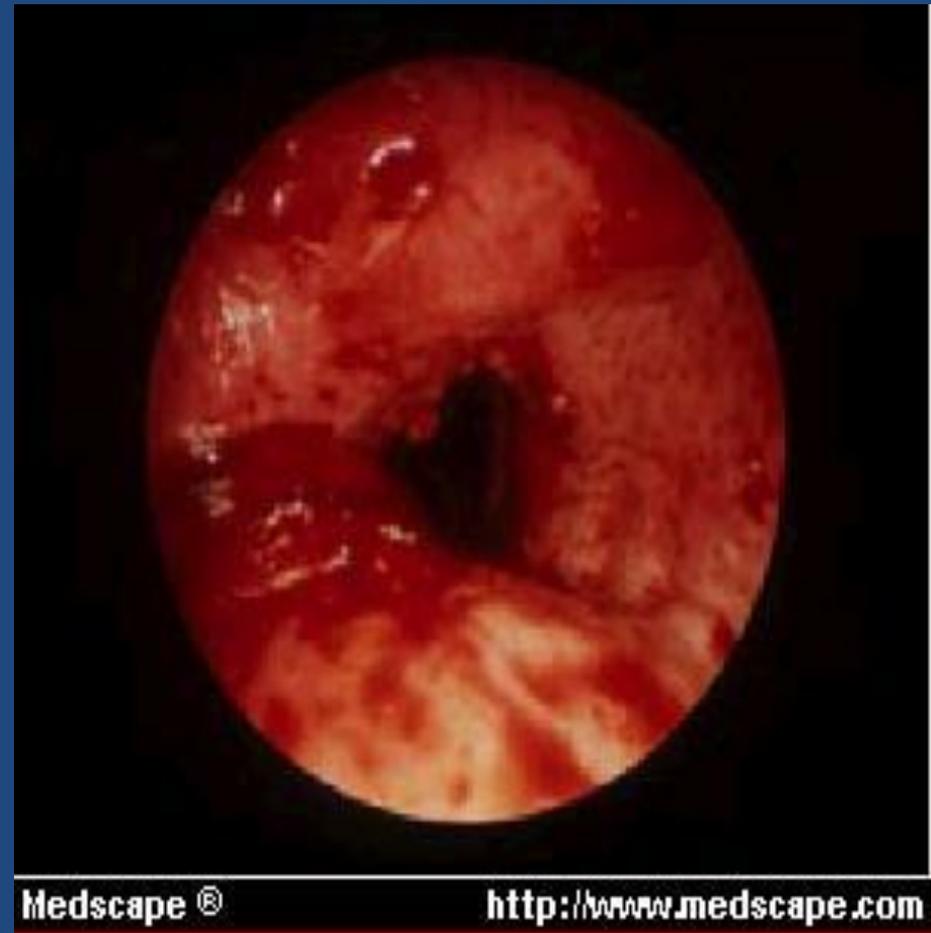
Risk factors for Massive hemoptysis

- FEV1 < 30% predicted
 - Pseudomonas aeruginosa*
 - Staphylococcus aureus*
 - Burkholderia cepacia*
 - Tube feeds
 - Cirrhosis
- ***lower risk observed in those patients receiving TOBI and pulmozyme

Treatment of hemoptysis

✚ Bronchoscopy:

- ✚ Attempts to visualize the region of bleeding
- ✚ Not very effective as view might be obscured by blood

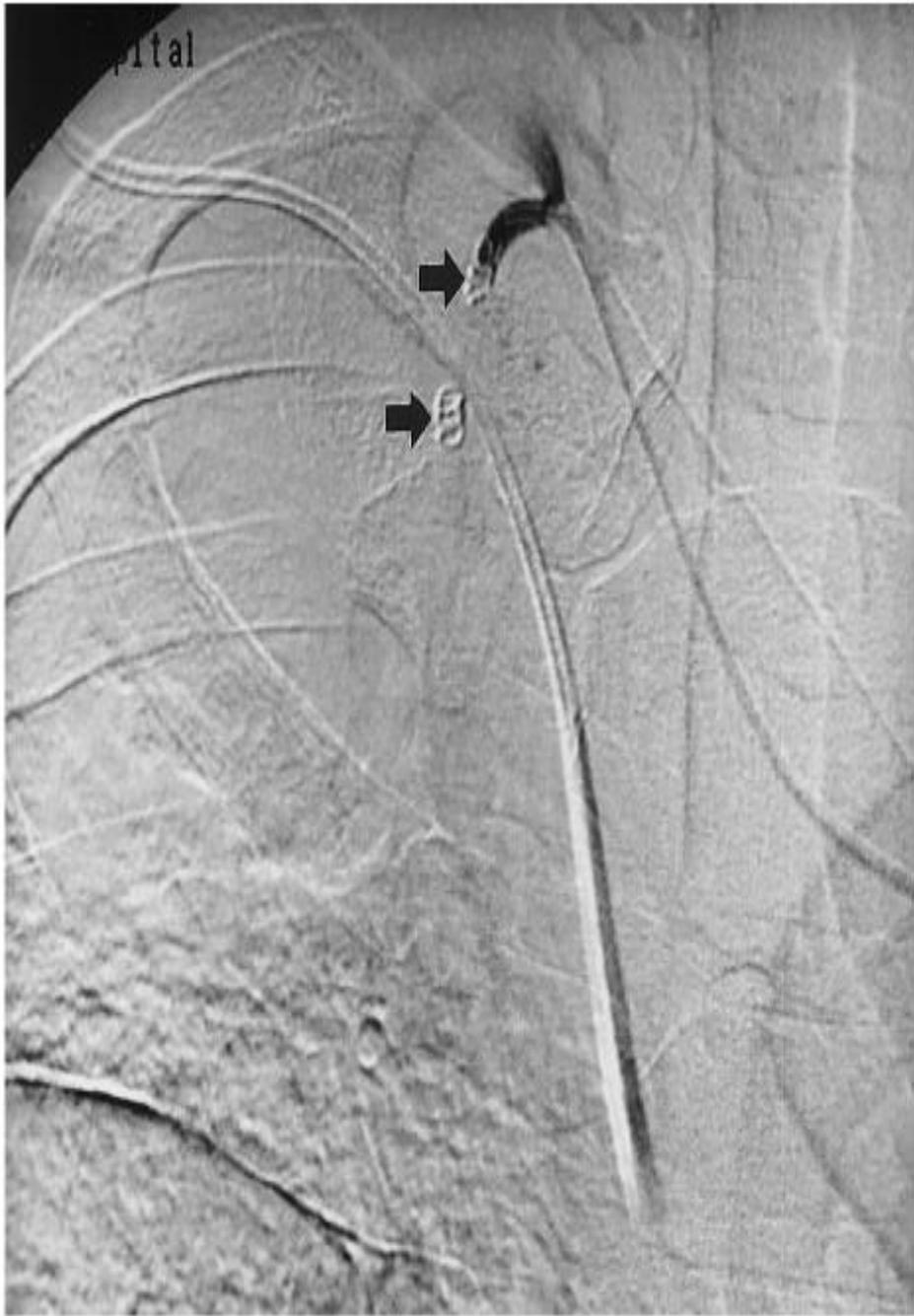


Bronchial artery embolization (BAE)

- ✚ First reported embolization 1973
- ✚ Recommended procedure of choice in massive hemoptysis
- ✚ Studies demonstrating efficacy in non-major hemoptysis
- ✚ Controls hemoptysis in approximately 90%
- ✚ 10-52% may require repeat BAE

BAE

- ✚ Catheter inserted into groin, femoral artery
- ✚ Contrast dye is injected into the arterial system
- ✚ Polyvinyl alcohol particles are injected into potential bleeding sites to occlude the blood supply
- ✚ Coils also used to occlude the bronchial arterial supply



a

b

Adverse effects

- ✚ Chest pain

- ✚ Difficulty swallowing

- ✚ Bronchial necrosis/damage

- ✚ Paraplegia: injury to spinal artery

- ✚ Respiratory failure

 - ✚ Recent evidence suggests that this may relate to low FEV1 and lung irritation from bleeding and not from procedure

Prognosis after massive hemoptysis

- Reported 44% mortality within one year
- Mean age of hemoptysis 26.4 +/- 9 years
- ***majority of these patients with FEV< 30% predicted

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Respiratory Failure

- Inadequate gas exchange
 - Low oxygen levels (hypoxemic resp. failure)
 - High carbon monoxide (hypercarbic resp. failure)
- Signs and Symptoms
 - rapid respiratory rate
 - breathlessness
 - tachycardia
 - low oxygen levels
 - confusion
 - loss of consciousness
 - respiratory arrest

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Respiratory Failure



Most common causes:

- 1) Pulmonary infectious exacerbation
- 2) Massive hemoptysis
- 3) Pneumothorax
- 4) other

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Respiratory Failure

- Previously most adult admissions to the ICU with high mortality
- Better survival attributed to:
 - Better antibiotics
 - Faster response time to critically ill inpatients
 - Use of non-invasive ventilation

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Non-invasive ventilation (NIV)

- ✚ May decrease need for mechanical ventilation
- ✚ Improves survival
- ✚ Bridge to lung transplantation
- ✚ For use in mild to moderate respiratory distress



Treatment

- Oxygen
- Mechanical ventilation (intubation or non invasive)
- Bronchodilators
- Antibiotics
- Treatment of precipitant (Pneumothorax, hemoptysis, etc.)
- Supportive care: nutrition, prevention of complications

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Respiratory failure and ICU mechanical ventilation

- ✚ Very high mortality
 - ✚ Associated with low FEV1
 - ✚ Discussion regarding end-of-life should be initiated
- ✚ Eligible patients may have improved survival with lung transplantation

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Summary

- ✚ Cystic fibrosis with improved survival
- ✚ Better strategies in treating critically ill patients
- ✚ Pneumothorax and Massive hemoptysis with better outcomes if treated early; recurrence is not uncommon
- ✚ Non-invasive ventilation improves survival, particularly if FEV1 > 30%.
- ✚ Respiratory distress requiring mechanical ventilation has a high mortality

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Aknowledgements

- Dr. Paul Mohabir

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Thank you

