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Pulmonary Complications of Cystic Fibrosis

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Goals and Objectives

Participants will be able to

- Explain the pathophysiology leading to the pulmonary complications of cystic fibrosis
- Recognize the clinical presentation of the pulmonary complications of cystic fibrosis
- Formulate a treatment plan for the pulmonary complications of cystic fibrosis

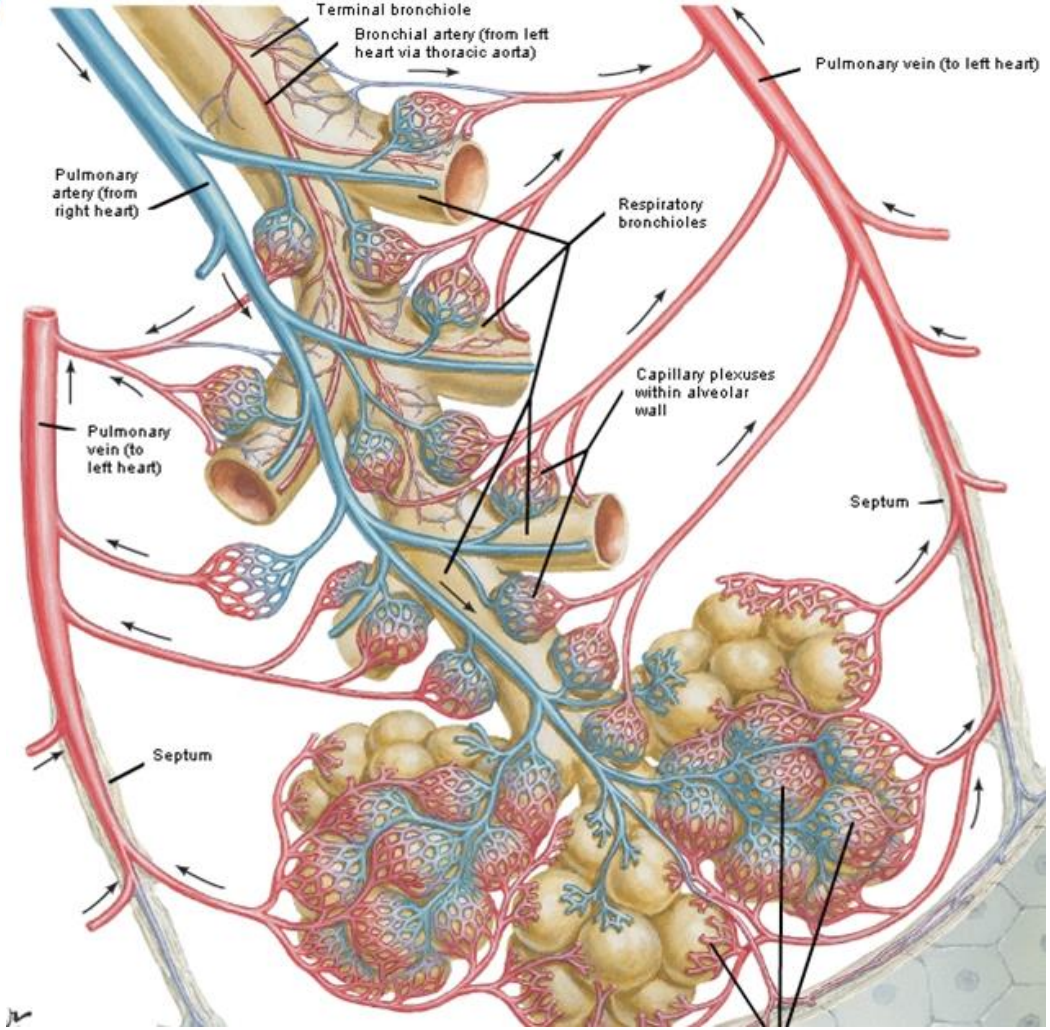
CFF Consensus Guidelines

- Am J Respir Crit Care Med Vol 182. pp 298–306, 2010
- Delphi methodology
  - PICO questions developed by CFF Pulmonary Guidelines Committee
  - 42 CF clinicians, 6 interventional radiologists, 7 transplant surgeons
  - Literature search (1985-2009)

- Questionnaire
  - Series of statements on Hem and Pntx
  - Likert scale of 0-10 (Hem) or 0-9 (Pntx) representing complete disagreement to complete agreement
  - Completed, refined, completed a second time
  - Consensus report generated
- Consensus Definitions
  - Perfect = all respondents agree
  - Very Good = Median and middle 50% (IQR) of respondents are found at one integer (e.g., median and IQR are both at 8) or 80% of respondents are within one integer of the median (e.g., median is 8, 80% of respondents are from 7–9)

- Good = 50% of respondents are within one integer of the median (e.g., median is 8, 50% of respondents are from 7–9) or 80% of respondents are within two integers of the median (e.g. median is 7, 80% of respondents are from 5–9)
- Some = 50% of respondents are within one integer of the median (e.g., median is 8, 50% of respondents are from 7–9) or 80% of respondents are within two integers of the median (e.g. median is 7, 80% of respondents are from 5–9)
- None = All other responses

– Subsequent summaries must have “Good” agreement and IQR of ≥ 5



EPIDEMIOLOGY AND PATHOPHYSIOLOGY HEMOPTYSIS

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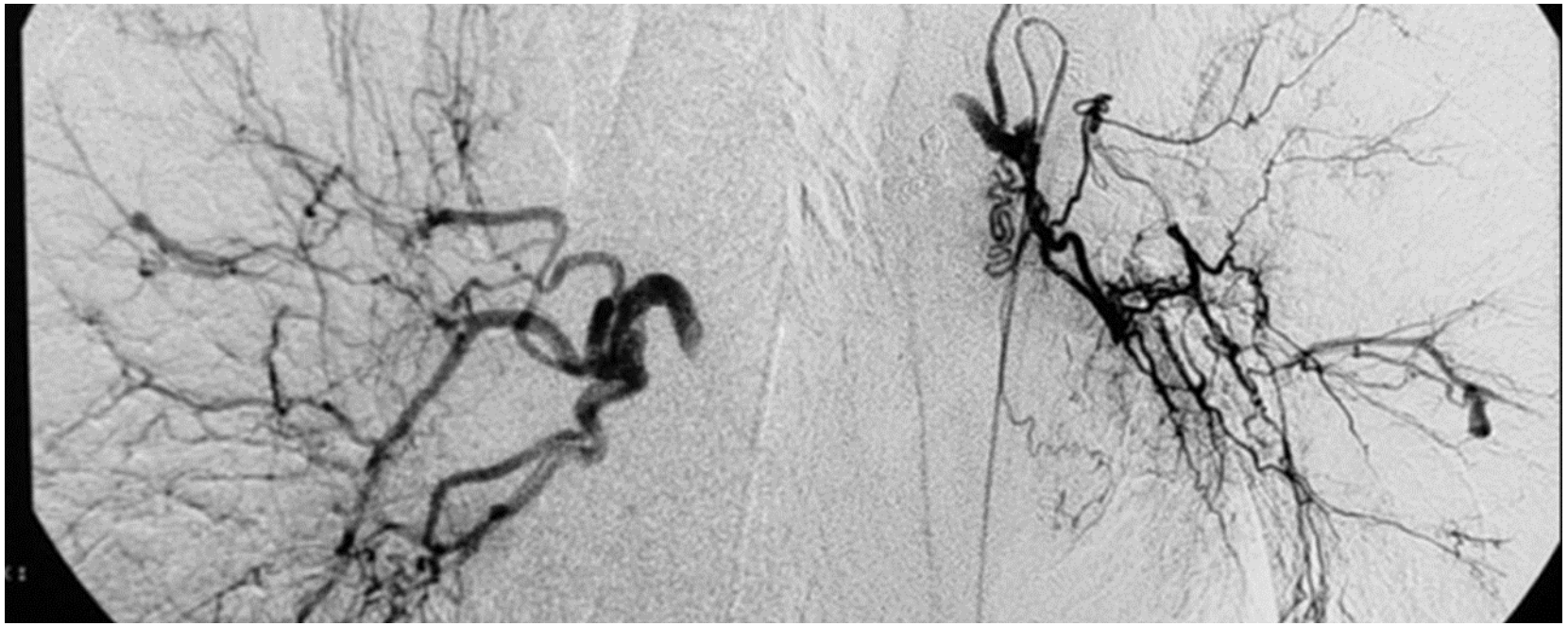
- 2008 Israeli review reported 9.1% of patients had hemoptysis in a 5-year period
- 2005 US review
  - 4.1% of all patients will suffer massive hemoptysis during their lifetime
  - average annual incidence is 0.87% (1 in 115 patients per year)
    - Median age for massive hemoptysis is 23 years
    - 75% of cases occur in patients $\geq$ 18 years of age

- Chronic inflammation leads to bronchial artery hypertrophy and angiogenesis
  - High pressure system
  - Thin-walled vessels
- Acute infection or pulmonary exacerbation often associated with hemoptysis
  - Association with SA

- Definition
  - Scant = blood streaking or < 20 ml/day
  - Mild-moderate = > 20 ml/day but < 240 ml/day
  - Massive = 240 ml/day or recurrent bleeding of substantial volumes >100 ml/day (> 500 ml/day in adults sometimes used)

DIAGNOSIS OF HEMOPTYSIS

- Coughing up blood or blood-stained mucus
- “Pseudohemoptysis”
  - Upper respiratory or GI source
  - Fictitious
- CXR and CT rarely helpful in identifying vessel involved
- Bronchial artery angiography most helpful

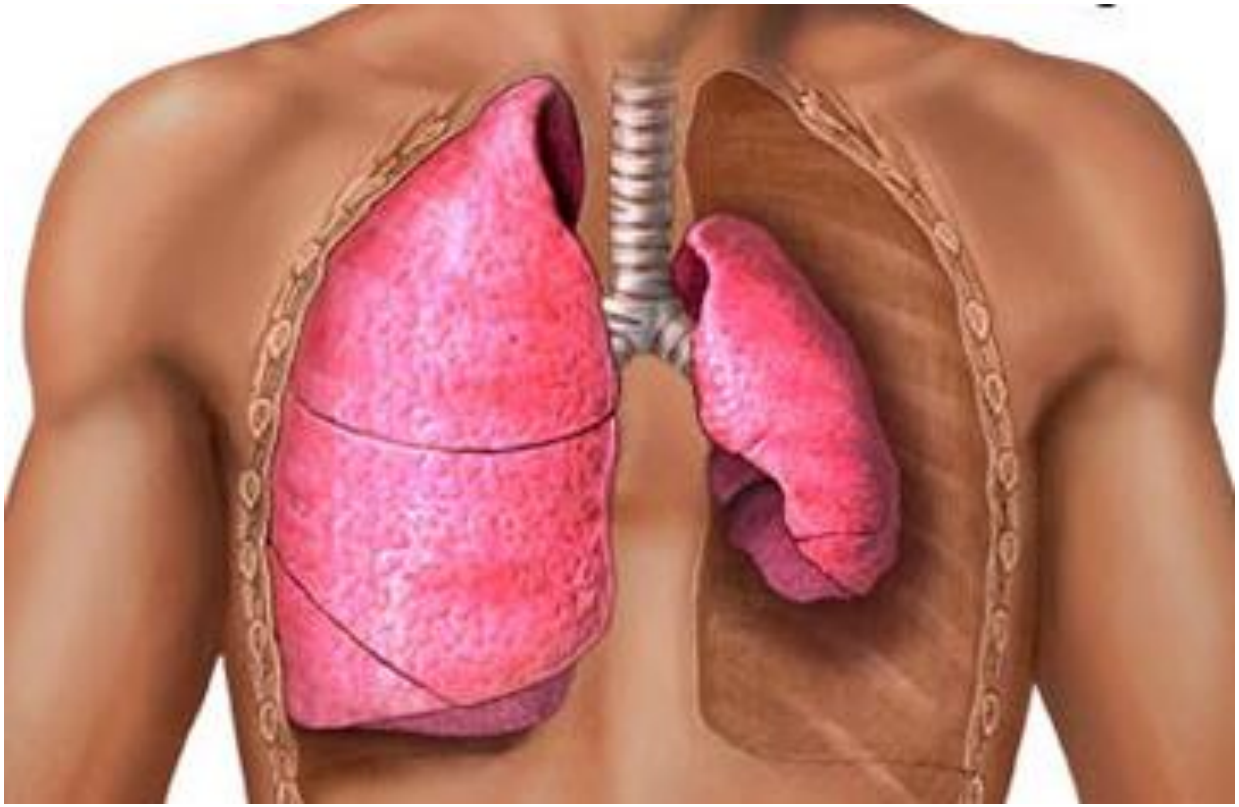


THERAPY
HEMOPTYSIS

- CFF Guidelines
  - First episode of scant, persistent scant, mild-to-moderate, or massive hemoptysis requires contact with provider
  - Massive hemoptysis requires admission
  - Mild-to-moderate or massive hemoptysis requires antibiotic therapy
  - NSAIDs should be stopped for **any** hemoptysis
  - Unstable patients with massive hemoptysis should have BAE

- Airway clearance therapy
  - Good consensus that scant hemoptysis did not require discontinuation of ACT
  - Poor consensus and no recommendation for mild-moderate hemoptysis
  - Better consensus that ACT be discontinued for massive hemoptysis
- Aerosol therapy
  - Good consensus that scant hemoptysis did not require discontinuation of aerosol therapy
  - Poor consensus and no recommendation for mild-moderate hemoptysis
  - Good consensus that hypertonic saline should be discontinued but no consensus on other therapy for massive hemoptysis

- Medical management
  - Assess CBC and coagulation studies
  - Discontinue any medications associated with anticoagulation
  - Consider treatment with vitamin K, tranexamic acid, empiric antibiotics
    - Tranexamic acid prevents plasmin-tPA complex from binding to fibrin, inhibiting fibrinolysis; Q4-8 dosing (oral or IV); can clot central lines
  - Supportive care (oxygen, fluids, transfusion)
- Bronchial artery embolization
  - Demonstrated efficacy; often limited by resources; not risk-free



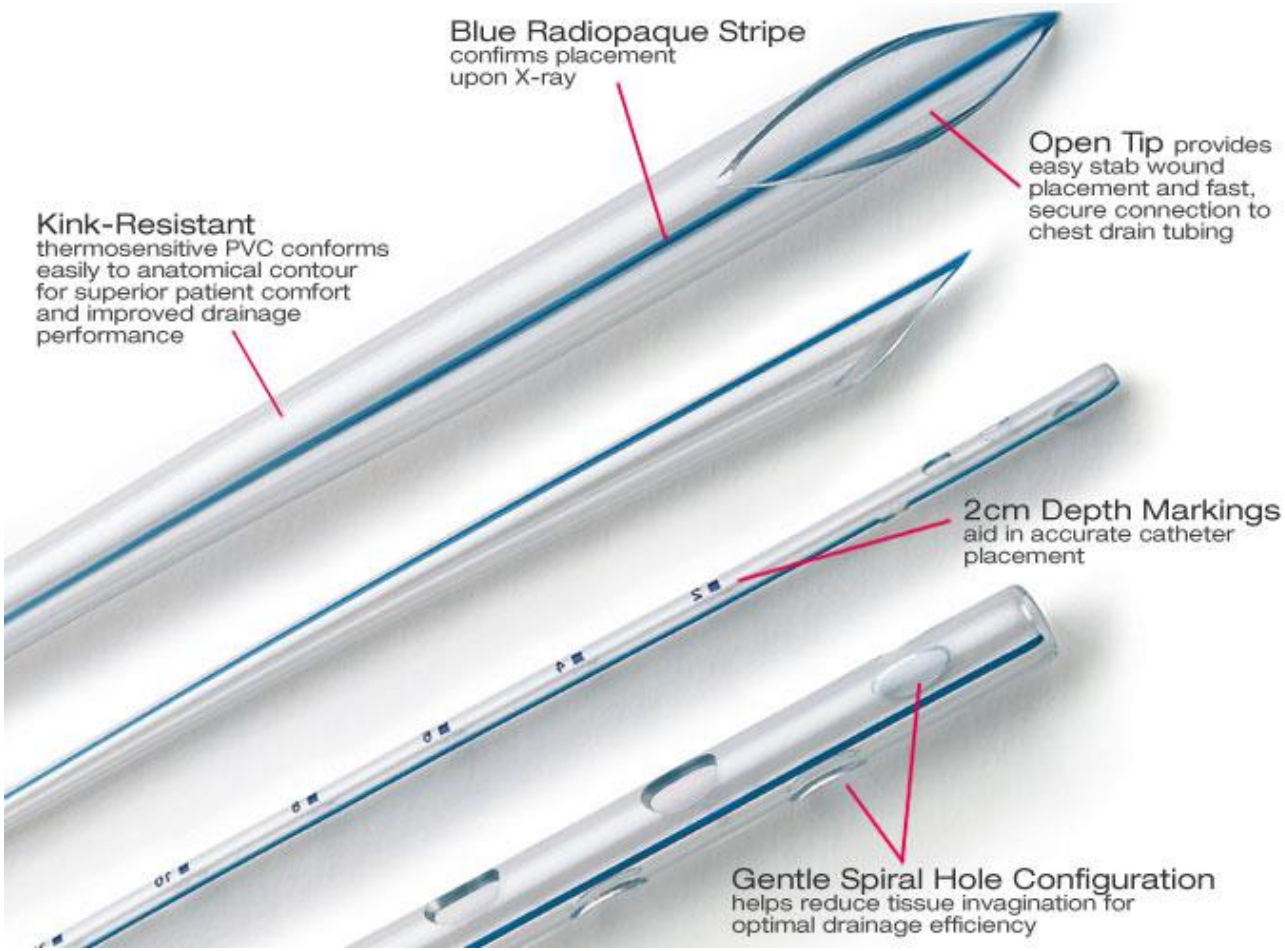
EPIDEMIOLOGY AND PATHOPHYSIOLOGY PNEUMOTHORAX

- 3.4% of individuals will experience a pneumothorax during their lifetime
  - Average annual incidence of 0.64% (1 in 167 patients per year)
  - Median age for pneumothorax was 21 years
    - 72.4% occurred in patients $\geq$ 18 years of age
  - Risk factors
    - 75% occur in patients with an FEV_1 less than 40% predicted
    - Other indicators of advanced disease (PA, Cepacia, medications, etc.)
  - 46% of patients with contralateral recurrence
  - 48.6% 2-year mortality

- Pathophysiogy
  - Air-trapping caused by obstruction
  - Alveolar pressure exceeds interstitial pressure
  - Air movement from alveolus to interstitium
  - Rupture of air into mediastinal or pleural space

DIAGNOSIS OF PNEUMOTHORAX

- Symptoms
  - Acute onset chest pain
  - Dyspnea
  - Cyanosis
- Diagnostic imaging
  - CXR
  - CT



THERAPY
PNEUMOTHORAX

- CFF Guidelines
  - Small, stable pneumothorax can be observed; large pneumothorax requires admission
  - Small unstable and **any** large pneumothorax requires a chest tube
  - **Any** recurrent pneumothorax requires surgical pleurodesis
  - BiPAP should be discontinued for **any** pneumothorax
  - Air-travel, weight lifting, and PFT are prohibited for 2 weeks for **any** pneumothorax

- Airway clearance therapy
  - Poor consensus overall except that positive pressure ACTs should generally be avoided and that absolutely should be avoided for large Pntx
- Aerosol therapy
  - Good consensus that aerosol therapy should be continued with the exception of hypertonic saline, for which there was no consensus

EVERYTHING ELSE

- Respiratory Failure
  - Epidemiology & Pathophysiology
    - Hypoxemia and hypercarbia caused by ventilation-perfusion mismatch
    - Muscle fatigue
  - Diagnosis
    - Pulse oximetry
    - Blood gas sampling
  - Therapy
    - Acute care for pulmonary exacerbation
    - Oxygen supplementation
    - Ventilatory support
    - Lung Transplant

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Thank You!

Questions?

Discussion?