Cystic Fibrosis Complications

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MARCH 3, 2012
INTRODUCTION
PNEUMOTHORAX
HEMOPTYSIS
RESPIRATORY FAILURE
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
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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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
Normal CXR

PTx

Collapsed Lung tissue

Shift of heart
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
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
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

Flume et al, Chest 2005
Risk Factors for PTx

- Pseudomonas aeruginosa
- Burkholderia cepacia
- Aspergillus / ABPA
- FEV$_1$ ≤ 30% predicted
- Pancreatic insufficiency
- Tube feeds
- Medicaid
Treatment of Pneumothorax

Asymptomatic: PTx < 2 cm

Admit, observation, repeat CXR(S)
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
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
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
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
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
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
Respiratory Failure

Most common causes:

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

• Dr. Paul Mohabir
Thank you