Cystic Fibrosis & Bronchiectasis

Cystic fibrosis is a fatal autosomal recessive disease carried on chromosome 7; although it is manifested as pancreatic, hepatic, pulmonary, GI, and reproductive abnormalities, 90% of the reported morbidity and mortality is pulmonary; the pulmonary pathologic process is severe obstructive disease with bronchiectasis, emphysema, and ultimately, terminal respiratory failure; bronchiectasis is a chronic suppurative disease of the airways characterized by localized, irreversible dilation of bronchi d/t destructive inflammation.

ANESTHETIC CONSIDERATIONS:

- End stage respiratory disease:
  - Severe obstructive / restrictive disease & difficulties maintaining oxygenation
  - Chronic colonizing infections (esp. pseudomonas)
  - Increased perioperative complications including cardio-respiratory collapse
  - Pulmonary HTN / cor pulmonale
  - Potential for massive hemoptysis
- Specific to CF:
  - Pancreatic insufficiency: DM, glucose intolerance
  - Malabsorption, malnutrition – vitamin K deficiency (coagulopathy)
  - Hepatobiliary dysfunction (cirrhosis & portal HTN)
  - GERD & constipation (aspiration risk)
  - Depression
- Operative: related to end-organ damage → cholecystectomy, lung resection / transplant; obstetrics
- Perioperative considerations:
  - Aggressive respiratory therapy
  - Potential requirement for OLV
  - Potential post-op ICU ventilation

ANESTHETIC GOALS:

- Minimize perioperative pulmonary complications:
  - Lung protective ventilation strategies: minimize PIPs, frequent clearance of secretions, low PEEP, humidified circuit
  - Avoid triggers of bronchospasm
  - Rapid wean from ventilator if possible
  - Aggressive respiratory therapy
  - Postoperative pain control & splinting
- Minimize lung contamination (hemoptysis or abscess)
- Avoidance of factors that cause pHTN

HISTORY

- Evidence of infection:
  - Increased cough / sputum
  - Decreased exercise tolerance
  - Fever
- DM control
- Assess extent of GI pathology: generalized cachexia (weight gain < 4.5 kg in pregnancy associated with poor outcome), nutrition, mechanical ileus, GERD, hepatic dysfunction, cholelithiasis, pancreatic insufficiency, hepatic fibrosis, intestinal obstruction
- Assess for cor pulmonale: dyspnea, cough, orthopnea, cyanosis
- Treatment: bronchodilators, corticosteroids, mucolytic agents, NSAIDs (may slow respiratory deterioration), antibiotics

PHYSICAL

- **GENERAL** – full set of vitals, including room air SpO_2; fever, tachypnea, accessory muscle use, change in auscultatory exam (evidence of infection)
- **HEENT** – airway, nasal polyps, sinus drainage (sinusitis)
- **CVS** – cor pulmonale → tachypnea, rales, rhonchi, wheezing, clubbing, cyanosis, peripheral edema, raised JVP, S3/S4; evidence of pHTN (loud P2, parasternal heave)
- **RESP** – hyperinflation, poor ventilation, cyanosis, clubbing, rales, rhonchi, cough wheeze (bronchiectasis, atelectasis, pneumonitis, bronchospasm)
- **GI** – jaundice
- **MSK** - cachexia

INVESTIGATIONS

- **Labs**
  - CBC for increased WBC & anemia of chronic disease
  - E-lytes, BS, BUN, Cr, LFTs, coagulation studies
- **Imaging**
  - CXR and comparison to previous films vs. CT of chest (better idea of disease progression)
  - ECG & ECHO if any evidence of pHTN
- **Special**
  - Baseline PFTs & ABG w/ comparison to previous hospitalizations & noted effect of bronchodilators

OPTIMIZATION

- Consultation w/ respirology & RT / PT services while ensuring adequate perioperative care (ICU / acute step-down unit)
- Pediatrics consultation
- Ensure aggressive preoperative respiratory therapy:
  - Bronchodilators
  - Incentive spirometry
  - Postural drainage & chest physiotherapy - ensure continues up until OR (i.e. do in morning prior to procedure)
  - C&S directed antibiotics - decrease sputum production & incidence of hemoptysis

- Vitamin K for coagulopathy
- Aspiration prophylaxis
- Controversial:
    - Consider avoiding respiratory depressants
    - Anticholinergics have not been shown to significantly increase tenacity of mucous secretions

ANESTHETIC OPTIONS

- Guided by surgical and patient considerations
- Local or regional when possible is reasonable (no evidence)
- Concerns:
    - Coagulopathy & RA
    - Cardiorespiratory reserves & sedation
    - Postoperative pain control & ventilation weaning

ANESTHETIC SETUP

- **Drugs**
    - Epinephrine (1 amp 1:1000 in 250 cc NS mini-bag = 4 mcg/cc drawn up in 10 cc syringes = 40 mcg doses of epinephrine for ETT flushing for hemoptysis)
- **Equipment**
    - Standard CAS monitors + temp + nerve stimulator
    - +/- Art-line
    - Largest ETT possible for adequate suctioning w/ red-rubber suction
    - Availability of DLT if OLV needed
    - If severe RLD or evidence of pHTN, then PAC or TEE can be of use in directing management of intraoperative events
    - Consider pulmonary vasodilators for severe patients

MANAGEMENT OF ANESTHESIA

- **Induction**
    - Guided by procedure and severity of CF
    - Avoid ketamine d/t increased secretions
    - Avoid nasal intubation d/t sinusitis
    - Bronchiectasis – isolation of bronchiectatic area of lung with DLT
        - DLT / OLV in CF patients complicated by: hypoxia (both lungs are dysfunctional, PTX to non-dependent lung)
- **Maintenance**
    - Aggressive suctioning throughout case aids ventilation
    - IH agents will work slower in severe disease but will decrease NMB use & help w/ bronchospasm
    - Avoid triggers of pHTN
    - Be aware of PTX risk
    - Avoid triggers of bronchospasm
    - PEEP and short expiratory times increase risk of auto PEEP and PTX
    - Avoid high ventilation pressures (PTX)
- **Emergence**
    - Use adjuvants for pain control: will increase probability of extubation at end of case - acetaminophen, NSAIDs, LA wound infiltration, RA, remifentanil infusion to extubate

DISPOSITION & MONITORING

- Think about ICU postoperatively
- Continue aggressive respiratory therapy - may require postoperative ventilation
- Regional anesthesia for postoperative pain control may help optimize respiratory mechanics postoperatively

COMPLICATIONS

- Massive hemoptysis (see Seminar):
    - Treat coagulopathies - Vitamin K if hepatobiliary dysfunction
    - Consider taking to radiology for embolization after stabilizing as best as possible (DLT & prone positioning) if > 200 mL/h
- PTX
- Post induction cardio-respiratory collapse (end-stage patients presenting for lung transplant) - multifactorial “Circle the Drain” syndrome:
    - Severe lung disease w/ V/Q mismatching causes hypoxemia
    - Hypoxemia initiates exacerbation of pHTN cascade (HPV, acidosis, bronchoconstriction)
    - Hypoxemia & pHTN cause complete RV failure
- Exacerbated bronchiectasis: postural drainage, humidified O_2, hypertonic NS nebs
- Failure to wean:
    - Awake, warm and comfortable
    - Chest physiotherapy & secretions suctioned
    - Ensure full reversal on NMB esp. if on aminoglycoside antibiotics

PATHOPHYSIOLOGY

- CF = autosomal recessive disorder of exocrine glands primarily affecting the pulmonary and gastrointestinal systems
- Epidemiology:
    - 1:2500 births
    - Up to 1:25 Caucasians are carriers
    - 33% survival to > 30 yrs currently
- Pathophysiology:

- Chromosome 7 mutation causes defective chloride ion transporter in epithelial cells in the lungs, pancreas, liver, gastrointestinal tract, and reproductive organs leading to decrease transport of Na & water causing thick, viscous secretions, luminal obstruction & scarring / destruction of exocrine glands
- Secretions coupled w/ decreased ciliary activity lead to pneumonia, wheezing, and bronchiectasis
- Chronic changes include granulation tissue w/ increased vascularity
- PFTs are characterized by increased residual volume and airway resistance w/ decreased VC and expiratory flow rates (mixed obstructive / RLD)
- Malabsorption syndrome may lead to dehydration and e-lyte abnormalities

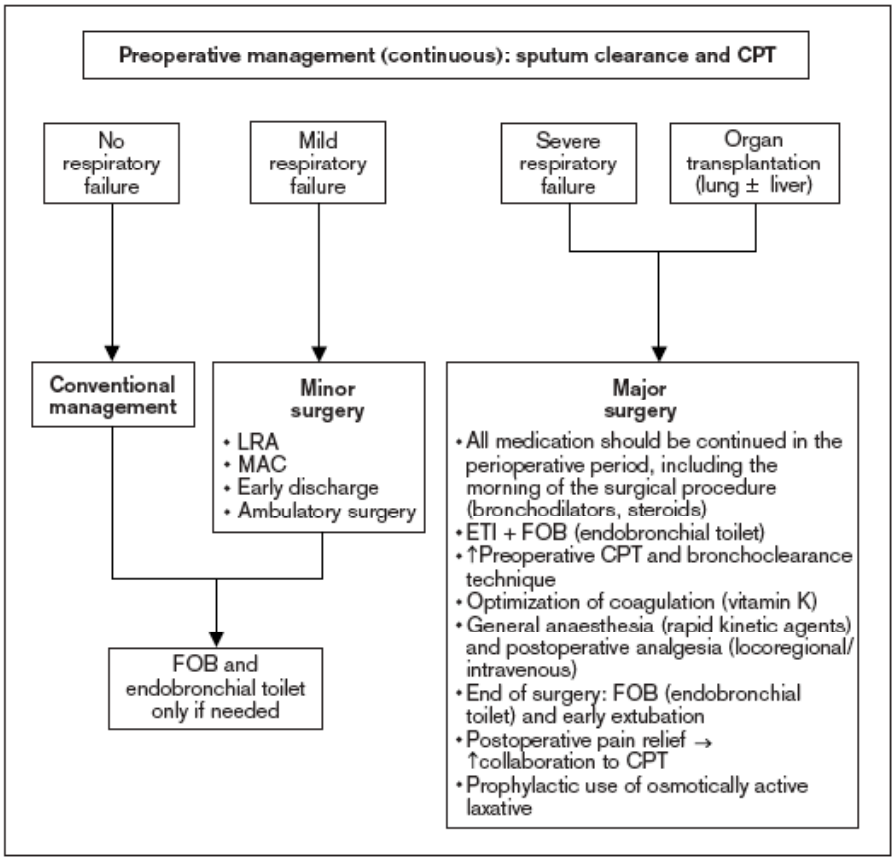
- Diagnosis = analysis of chloride content of sweat (Cl^- > 80 mEq/L)
- Management includes aggressive respiratory physiotherapy, antibiotics, and digestive enzyme replacement
- Bronchiectasis = chronic suppurative disease of the airways characterized by localized, irreversible dilation of bronchi caused by a destructive inflammatory process
  - Patients have an increased susceptibility to recurrent bacterial infections - most commonly Pseudomonas (nearly impossible to eradicate)
  - Major complication = hemoptysis
  - Etiologies include viral, bacterial or TB pulmonary infection followed by recurrent "chest colds;" recurrent aspiration; immunodeficiencies
  - Risk factors = hereditary defects in ciliary mucosal clearance: CF, Kartagener's syndrome, alpha-1 antitrypsin deficiency, bronchial cartilage deficiency (Williams-Campbell)
- Common Surgical Presentations:
  - Nasal polypectomy
  - VAD
  - Bronchoscopies
  - Bowel obstruction - laparotomy
  - End-organ dysfunction: cholecystectomy, liver & lung transplant
  - Obstetrical cases

Table 1. The most frequent indications for anaesthesia in cystic fibrosis

Neonates	Children/teenagers	Adults
Meconium ileus	Nasal polpectomy	Oesophageal varices
Meconium peritonitis	Intravenous access	Recurrent pneumothorax
Intestinal atresia	Ear/nose/throat surgery	Cholecystectomy
		Lung (liver) transplantation

Figure 1. Algorithm for anaesthesia management in patient with cystic fibrosis

CPT, chest physiotherapy; ETI, endotracheal intubation; FOB, fibreoptic bronchoscopy; LRA, loco-regional anesthesia; ↑, increase; MAC, monitored anaesthesia care.



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- Savage. Prone, head down for Pulmonary Hemorrhage. BJA 89(1): 184-188 (2002). - case report (one of many on medline, but only one I could get e-text for)