# ANESTHESIA FOR PATIENTS WITH CYSTIC FIBROSIS (CF)

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| **Surgical Specialty:** | Other |
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## Background:

- General Considerations[1]
  - Characterized by obstruction and infection of the airways (bronchiolitis) as well as nutritional malabsorption due to impaired sodium and chloride transport across epithelial cells
  - The rate of progression of lung disease is the primary determinant of morbidity and mortality
  - Autosomal recessive inheritance pattern
    - 1:3200 Caucasian births
  - Caused by a mutation in the CF gene which codes for a transmembrane conductance regulator (CFTR) protein
    - CFTR protein is a chloride ion channel expressed in the epithelial cells of the airways, pancreas, liver, GI tract, sweat glands, and GU system
    - Loss of chloride transport disrupts the chloride and sodium balance causing the production of an abnormally thick layer of mucous.
- Patient Considerations[1]
  - Disease Specific Considerations
    - Characterized by thickened secretions
    - Treatment consists of pancreatic enzyme replacement, antibiotics, anti-inflammatory agents, bronchodilators, oxygen, and chest physiotherapy
    - The development of cystic fibrosis modulators, which restore CFTR folding and function, has led to improved quality of life as well as increased life expectancy
    - Lung transplant is the only treatment option for end-stage lung disease
  - Associated Comorbidities/Syndromes:
    - **Pulmonary:**
      - Typical signs and symptoms include chronic productive cough, lung hyper-expansion on CXR, obstructive pattern on PFTs
      - Bullae formation increases the risk of spontaneous pneumothorax
      - Higher incidence of bacterial and viral infections: chronic pulmonary carriage of antibiotic-resistant organisms (ex. Pseudomonas aeruginosa, Aspergillus fumigatus)
      - Bronchial hyperreactivity increases the risk of bronchospasm
      - Sinus infections, chronic nasal congestion, nasal polyp, hemoptysis

- **Endocrine and Growth:**
  - Pancreatic insufficiency[2]
  - CF-related diabetes mellitus (Caused by destruction and fibrosis of pancreatic parenchyma, clinically apparent in adulthood)
- **Gastrointestinal:**
  - Meconium ileus (Can be presenting symptom in neonates)
  - Recurrent small bowel obstruction (Distal Intestinal Obstructions Syndrome: DIOS)
  - Cholecystitis, cholelithiasis, and focal biliary cirrhosis (due to thickened bile)
  - Fat-soluble vitamin (ADEK) deficiencies[3]
  - Plasma cholinesterase deficiency due to hepatic involvement (prolonged duration of succinylcholine)
- **Genitourinary:**
  - Nephrolithiasis
  - Infertility[2]
- **Behavioral and Psychiatric:**
  - Depression and anxiety[2]
- **Hematologic:**
  - Increased bleeding risk due to hepatic involvement and vitamin K deficiency
- **Cardiovascular:**
  - Increased risk for pulmonary hypertension and right heart failure (cor pulmonale) due to chronically elevated pulmonary vascular resistance[2]
- **Musculoskeletal:**
  - Premature osteoporosis

## Anesthetic Planning:

- Pre-Anesthetic Evaluation[1]
  - Care for patients with advanced CF can be complex
    - Often, CF patients have tiered treatment plans recommended by their pulmonologist for exacerbations or in preparation for a procedure.
    - Review pulmonology records, and discuss recommendations with the pulmonologist.
  - Evaluate the severity of the disease
    - Frequency and severity of exacerbations and infections
    - Volume of sputum
  - Pre-operative optimization
    - Daily pulmonary clearance techniques, bronchodilators, mucolytics, antibiotics
    - Consider scheduling the case later in the day to allow time for preoperative airway clearance interventions.
    - May need to delay the case 4-6 weeks after a recent respiratory infection
  - Ensure glycemic control
    - Perioperative environment can cause stress and hyperglycemia

    - Additional labs/tests that may be indicated during work-up (based on disease severity)
        - Electrolyte panel
        - Glucose
        - Coagulation profile (if considering neuraxial)
        - PFTs (with FEV1 being the best indicator of disease severity) can predict the need for post-operative ventilation, especially for intrathoracic and large intra-abdominal cases.
        - Low threshold to order ECG/TTE if concerned about right heart dysfunction from chronic hypoxemia
        - Baseline ABG if available (High alveolar-arterial oxygen gradient and high baseline paCO2 indicate severe disease)
    - Discussions to have with surgeon/family
        - Code Status
        - ECMO Candidacy (if the patient is critically ill or undergoing a high-risk procedure)
        - Possible need for postoperative mechanical ventilation
- Specific or Unique Room Set-up Requirements
    - Airway
        - Focus on rapid restoration of baseline pulmonary status
        - Supraglottic devices are acceptable for minor procedures
        - ETT may be needed to manage secretions
        - Humidify inspiratory gases
        - Frequent suctioning may be needed to prevent mucous plugs
    - Drugs/Infusions
        - Strategies for pain control should aim to improve post-operative respiratory status by employing non-opiate, multimodal adjuncts, and regional or neuraxial techniques.
        - Nephrotoxic medications (ie NSAIDS) are contraindicated in patients with advanced disease.
    - Monitors
        - Standard American Society of Anesthesiologists monitors
        - Train of four monitoring to ensure complete neuromuscular blockade reversal
        - Consider arterial line if indicated by case severity and need for ABG monitoring. CF patients may poorly tolerate surgeries that otherwise might not require frequent ventilation and acid-base monitoring (ex. cases with longer laparoscopic times or necessitating large-volume resuscitation)
    - PICU Bed Availability (if indicated)
        - May be needed based on the severity of the disease
    - Other Indicated Resources (ex. ECMO standby, respiratory therapy, physical therapy)

## Intraoperative Considerations:

- General[1]
    - Undergo a range of procedures—nasal polypectomy, gastrostomy tube placement, cholecystectomy, vascular access, lung biopsies, other thoracic surgeries, and lung transplantation.
- Induction
    - The decision to use mask versus IV induction is patient-dependent
    - Sufficiently preoxygenate prior to induction

    - Consider limiting nitrous oxide
    - Achieve adequate plane of anesthesia prior to laryngoscopy to avoid bronchospasm.
- Positioning
    - Give special care to padding. Avoid hyperflexion and extension of extremities due to increased incidence of osteoporosis.
- Maintenance[1]
    - Focus on rapid restoration of baseline pulmonary status.
        - Maintain spontaneous ventilation with adequate ventilation
        - Avoid paralytic and long-acting sedating medications if possible
        - Use opioid-sparing techniques
    - Frequent suctioning to prevent mucous plugs
    - Avoid desflurane (increases airway reactivity). Consider sevoflurane (potent bronchodilator) or propofol for maintenance.
    - Check glucoses intraoperatively
- Hemodynamic/Physiologic goals
    - Maintain spontaneous ventilation when able. Allow adequate expiratory time if using controlled ventilation, and avoid high airway pressures in an effort to prevent pneumothorax and increased RV afterload.
- Surgical Considerations
    - Increased bleeding risk for major procedures (type and cross, vitamin K administration) [2]
- Emergence/Disposition
    - Early extubation preferred
    - Ensure complete neuromuscular blockade reversal to prevent atelectasis[4]
    - Suction, recruit lungs, and give bronchodilators prior to airway removal
    - Lower threshold for ICU admission if respiratory issues are encountered during the case
- Post-op Care
    - Pain control with opioid-sparing techniques (including regional/neuraxial if appropriate)
    - Aggressive pulmonary therapy (incentive spirometry, chest physiotherapy, bronchodilators, appropriate antibiotics)
    - Encourage early ambulation (physical therapy)
    - Provide PONV prophylaxis, early enteral nutrition, and glucose checks
    - Consider 24-hour admission for patients with advanced disease

## Case-Specific Complications/Pitfalls

- CF is the leading cause of lung transplantation in children[1]
- Major comorbidities include cirrhosis, right heart failure secondary to pulmonary hypertension, insulin-dependent diabetes mellitus, and increased bleeding risk. All of these should be screened for and treated appropriately[2]
- CF patients should have a lower threshold for admission and are often not appropriate for ambulatory surgery centers.
- Coordinate communication with the patient's pulmonologist, respiratory therapy, physical therapy, ECMO, ICU team, and surgery team.

## References


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Created: Date; Last revised: 5/22/25