Chapter

13

Cystic Fibrosis

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A nine-year-old girl with cystic fibrosis and recurrent pulmonary infections is scheduled to undergo repair of an umbilical hernia. Her home treatment regimen includes chest physiotherapy, albuterol, tobramycin, and nighttime humidified continuous positive airway pressure (CPAP), 8 mmHg.

Preoperatively her vital signs include: blood pressure 105/68; heart rate 86, respiratory rate 20; temperature 36.8°C SpO_2 97% on room air. Physical exam revealed a very thin young girl, cooperative with history and physical. Her respiratory effort is normal without the use of accessory muscles or retractions. She had coarse breath sounds bilaterally on auscultation. Airway exam was unremarkable.

What Genetic Factors Are Associated with Cystic Fibrosis?

Cystic fibrosis is caused by a mutation in the gene that encodes for the cystic fibrosis transmembrane regulator (CFTR) protein. This is a chloride channel that is found at the apical border of epithelial cells lining most exocrine glands. The mutation is the most lethal inherited disorder and displays an autosomal recessive pattern of inheritance.

What Are the Pulmonary Manifestations of Cystic Fibrosis?

Cystic fibrosis affects numerous organ systems although the primary cause of morbidity and mortality is related to pulmonary manifestations. Copious and thick secretions combined with mucociliary dysfunction lead to airway inflammation, atelectasis, and bacterial trapping. Bacterial overgrowth and inflammation cause biofilm formation and recurrent infection. With disease progression, airway obstruction from secretions causes further air trapping resulting in bronchiectasis. Patients are plagued by recurrent exacerbations leading to progressively reduced lung function, cor pulmonale, and hypercarbic, hypoxemic respiratory failure.

What Are the Nonpulmonary Clinical Manifestations of Cystic Fibrosis?

Common organ systems affected include: gastrointestinal, and genitourinary, endocrine, bone, skin, and reproductive systems (Table 13.1).

This Patient Is Scheduled for a First Case Start. However, the Patient's Pulmonologist Insists That the Patient Be Scheduled Later in the Morning. What Is the Basis for This Concern?

The timing of surgery is important as cystic fibrosis patients have very structured time-intensive treatment regimens that they adhere to in the morning. This often includes chest percussion, nebulized medication, postural drainage, and other pulmonary toileting strategies. A first start case may result in suboptimal preoperative preparation by the patient and their family on the morning of surgery.

What Are the Important Preoperative Studies in Patients with Cystic Fibrosis?

Pulmonary function studies may be useful in determining severity of disease. Cystic fibrosis patients tend to have obstructive disease with increased functional residual capacity (FRC), decreased forced expiratory volume (1 second) (FEV1), and decreased vital capacity. X-ray imaging of the chest may demonstrate air trapping as demonstrated by a flattened diaphragm. In some cases, emphysematous changes and upper lobe pulmonary blebs can also be identified on chest radiography.

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Table 13.1 Pathologic and clinical manifestations in cystic fibrosis by organ system

Organ system	Pathology	Clinical manifestation
Upper respiratory tract	Copious nasal secretions	Chronic sinusitis, nasal polyps
Lower respiratory tract	Mucoid secretions, mucociliary dysfunction	Recurrent infections, bronchiectasis, chronic hypoxemia
Cardiac	Cor pulmonale	Right ventricular hypertrophy
Hepatobiliary	Bile duct obstruction	Biliary cirrhosis, portal hypertension
Gastrointestinal	Intestinal secretions	Neonatal meconium ileus, recurrent intestinal obstruction, malabsorption (vitamins A,D,E,K)
Endocrine	Obstruction and fibrosis of pancreatic ducts	Impaired pancreatic exocrine function and induced diabetes
Reproductive	Abnormal cervical secretions (females) Absence of vas deferens (males)	Decreased fertility (females and males)
Integumentary	Increased chloride levels in skin	Impaired thermoregulation
Bone	Impaired calcium, vitamin D absorption, increased bone catabolism	Early onset osteoporosis

Liver studies, complete blood count (CBC), and coagulation screen may also be indicated. Malabsorption of vitamin K from the gastrointestinal tract alongside hepatobiliary disease can lead to inadequate synthesis of vitamin K dependent factors, which places patients at risk for hemorrhage. Intraoperative hemorrhage in the setting of a concurrent malabsorptive iron deficiency anemia increases the risk of an adverse event following moderate blood loss.

Serum electrolyte levels may reveal a hypochloremic, hyponatremic, metabolic alkalosis and dehydration. Pancreatic dysfunction may lead to glucose intolerance, necessitating blood glucose checks.

Finally, CO_2 retention becomes worse with disease progression. Hypercarbia and chronic hypoxia are predisposed to the development of secondary pulmonary hypertension, and in severe cases, cor pulmonale. An arterial blood gas may be necessary to evaluate baseline respiratory status. In advanced disease, cardiac evaluation may include ECG and echocardiography.

Is Premedication Indicated in This Patient?

Premedication with the patient's usual regime of inhaled medications should be confirmed with the parents. It is crucial that the patient receive bronchodilator. Anxiolysis with midazolam benefits most children as it improves compliance with anesthetic induction.

How Would You Induce Anesthesia in This Patient?

Either inhalational or intravenous induction may be considered. Inhalational induction in this patient cohort may be somewhat delayed, due to their increased FRC, small tidal volumes and V/Q mismatch resulting in a slow uptake of volatile anesthetics. Nitrous oxide should be avoided as it can expand pulmonary blebs and lead to bleb-rupture pneumothorax.

Intravenous induction with propofol, while rapid, abolishes spontaneous ventilation and may be associated with significant obstruction due to sudden loss of airway tone. This may necessitate positive pressure ventilation, placing the patient at risk for bleb-rupture pneumothorax. Addition of intravenous dexmedetomidine may help preserve spontaneous ventilation. Regardless of induction strategy, patients should be deeply anesthetized prior to airway manipulation as cystic fibrosis is

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associated with high airway reactivity and a potential to provoke bronchospasm.

What Is the Optimal Anesthetic Maintenance Strategy for CF Patients?

The optimal maintenance strategy for CF patients involves maintenance of spontaneous ventilation, use of bronchodilator therapy and minimization of postoperative respiratory depression. Inhaled agents allow for maintenance of spontaneous ventilation while simultaneously serving as a bronchodilator. If positive pressure ventilation is required, employing the lowest appropriate tidal volumes with moderate positive end-expiratory pressure (PEEP) while maintaining low airway pressures is most appropriate. This serves to minimize variations in the mean airway pressure, reducing the risk of bleb rupture.

Analgesia should be provided with a multimodal approach. Opiates increase the risk of postoperative respiratory depression. Adjunctive nonsteroidal anti-inflammatories are advised. Neuraxial and regional anesthesia should be employed when possible. Ketamine, while possessing analgesic properties due to NMDA antagonism, increases airway secretions and should be used with caution.

What Special Intraoperative Concerns Exist for This Population?

Cystic fibrosis patients often have nasal polyps, combined with a possible vitamin K deficiency; this may be associated with severe bleeding with nasal instrumentation.

These patients are at risk for dehydration with a hypochloremic, hyponatremic alkalosis. Adequate intraoperative hydration is important to correct electrolyte abnormalities and reduce the viscosity of airway secretions.

Due to chronic malabsorption, cystic fibrosis patients are often thin and vulnerable to heat loss. Postoperative shivering leads to increased oxygen consumption which combined with impaired ventilation may be a dangerous stressor to the patient.

How Would You Extubate This Patient?

During the case and immediately prior to extubation, suctioning is crucial to clear the airway of secretions. Early extubation is preferred to minimize the risk of hospital acquired ventilator associated pneumonia. The use of albuterol on emergence may be indicated. Recruitment maneuvers, with low inspiratory pressures, should be used to minimize atelectasis. Finally, extubation should not take place until the patient establishes spontaneous ventilation consistent with pre-induction tidal volumes. Use of CPAP should be considered, especially in patients on home CPAP therapy.

Early reinstatement of pulmonary physiotherapy is crucial for optimal recovery.

What Are the Risk Factors for Postoperative Mechanical Ventilation?

While few studies exist, a preoperative FEV1 of <1 L and hypoxemia are risk factors for postoperative intubation. Cor pulmonale suggests elevated pulmonary arterial pressures and advanced pulmonary disease. Open abdominal incisions significantly reduce FEV1 and empiric postoperative mechanical ventilation should be considered.

Should Paralysis Be Used in This Patient Cohort?

Paralysis should be avoided in cystic fibrosis patients when possible. It is important to minimize any agents that may cause postoperative respiratory compromise. Patients with CF have progressive loss of cartilaginous airway support and must rely on muscle tone to maintain airway patency. Neuromuscular blockade may lead to increased flow obstruction. If paralysis is used then twitch monitoring is essential. Antibiotic suppression therapy including tobramycin, an aminoglycoside, will prolong the effects of neuromuscular agents. Full reversal from blockade is necessary prior to extubation, however the cholinergic effects of neostigmine may be problematic. Sugammadex, if available, may be an appropriate alternative to acetylcholinesterase inhibitors. However, in patients requiring bronchoscopy and or biopsy, use of neuromuscular blockade may be warranted as paralysis reduces the risk of pneumothoraces, especially in post lung transplantation patients.

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Suggested Reading

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