Although rare, hemoptysis is a frightening and potentially life-threatening complication of cystic fibrosis (CF) that may occur during anesthesia. Anesthesia providers should be aware of the complication and know how to promptly and effectively manage it if encountered.

This case illustrates the effects of hemoptysis during anesthesia on a patient with an already compromised pulmonary status. A 26-year-old patient experienced massive hemoptysis during monitored anesthesia care for an esophagogastroduodenoscopy with percutaneous endoscopic gastrostomy tube placement. The patient's hemoglobin saturation with oxygen fell from a percentage in the low 90s to less than 10\% within seconds after placement of the scope. The patient was found to have bleeding from the right bronchial artery that was subsequently embolized.

Hemoptysis is a rare occurrence during monitored anesthesia care (MAC), but it is a potential complication of end-stage cystic fibrosis (CF). Massive hemoptysis may occur in severe cases of end-stage CF\(^1\) This is an expectoration of 200 to 600 mL of blood from the larynx, trachea, bronchi, or lungs in a 24- to 48-hour period causing acute airway obstruction. Tuberculosis and bronchiectasis are the most common causes of massive hemoptysis; bleeding neoplasms in the lungs may also be a source.\(^2\)

Immediate management of hemoptysis is directed toward airway management, oxygenation, and localizing the bleeding site. Death may result from hemoptysis due to asphyxia, aspiration of blood, and hemorrhage. Intubation with a large bore endotracheal tube (ETT) (to facilitate suctioning) and possibly a double lumen ETT (to isolate the affected lung) may be required.\(^2\)

In bronchiectasis, infection causes chronic inflammation of the lung parenchyma, which stimulates bronchial arteries to enlarge (hypertrophy) and become tortuous (angiogenesis). This is thought to create the source of bleeding in hemoptysis in the CF population.\(^3\) Bronchiectasis may be detectable by computed tomography scan as opposed to chest radiograph.\(^4\) Vitamin K deficiency, common in CF, may also contribute to hemoptysis.\(^5\)

A patient presenting to the gastrointestinal (GI) laboratory for an esophagogastroduodenoscopy (EGD) with percutaneous endoscopic gastrostomy (PEG) tube placement experienced massive hemoptysis during MAC anesthesia for the endoscopic procedure. We questioned the cause of the bleeding and whether there were signs to alert us of the potential for hemoptysis during this particular procedure.

Fatal complications of anesthesia in CF patients have only recently been published and thus far have only concerned general anesthesia, not MAC. One study of bronchial artery embolization for hemoptysis in CF reported 3 deaths with the induction of general anesthesia attributed to pulmonary hemorrhage.\(^5\) In another study, McDougall and Sherrington\(^6\) similarly describe 3 patients who experienced fatal hemoptysis during induction of general anesthesia for bronchial artery embolization procedures. Both reports claimed that positive pressure ventilation associated with induction of general anesthesia may have caused increased pressure and tension against airway walls and abnormal pulmonary arteries leading to hemorrhage. The practice of administering anesthesia for

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bronchial artery embolization can be applied to patients with CF for other procedures requiring anesthesia. Intermittent positive pressure ventilation should be avoided if possible. Alternatively, intravenous sedation, local anesthesia, and preservation of spontaneous respiration should be instituted during anesthesia whenever possible in this patient population.  

**Case Summary**  
A 26-year-old woman presented to the GI laboratory for EGD and PEG tube placement for malnutrition. She weighed 36 kg and was 155 cm tall. The patient had a history of CF with poorly controlled diabetes mellitus, dependent on Novolog insulin treatment, with frequent large swings in blood glucose levels. Before the procedure, she had been hospitalized for approximately 6 weeks for CF exacerbation and hemoptysis that required bronchial artery embolization shortly after admission. She was positive for methicillin-resistant staphylococcus aureus (MRSA) and pseudomonas aeruginosa pulmonary infection and was being treated with meropenem, tobramycin, azithromycin, and linezolid among several other CF medications. Reported drug allergies included cefaclor, iodine, and vancomycin. The EGD and PEG procedures had been attempted the previous day; however, they were aborted because an unspecified amount of blood in the oropharynx was assumed to be due to trauma from the scope device at that time.

An echocardiography from admission showed left ventricular ejection fraction of 50%, which was improved compared to earlier tests. An electrocardiogram performed 5 days earlier showed sinus tachycardia (heart rate 100/min), which was typical for this patient. Normal heart sounds were auscultated. Pulmonary function tests 1 week before the procedure revealed severe obstructive defect with a markedly reduced vital capacity: forced vital capacity (FVC), 1.28 L (institution normal, 3.33 L); forced expiratory volume (FEV<sub>1</sub>), 0.69 L (institution normal, 2.97 L); and FEV<sub>1</sub>/FVC, 54% (institution normal, 89%), which would be expected for a patient in the late stage of CF. Chest radiograph was reported to be consistent with CF changes and showed lung fields with pulmonary infiltrates, bronchiectasis, and fibrotic scarring. Laboratory tests on the morning of the procedure were as follows: hemoglobin/hematocrit, 8.8 g/dL and 26.6%, respectively; platelet count, 739 k/L; white blood cell count, 14,300; sodium, 143 mEq/L; potassium, 5.4 mEq/L; chloride, 99 mEq/L; carbon dioxide, 32 mEq/L; blood urea nitrogen, 16; creatinine, 0.36; mg/dL; albumin, 2.4 g/dL; magnesium, 1.8 mEq/L; calcium, 8.7 mg/dL. Liver function tests and coagulation studies were within normal limits. The patient's baseline arterial blood gas reading approximately 2 weeks before the procedure showed compensated respiratory acidosis with the following results: pH, 7.36; PaCO<sub>2</sub>, 86 mm Hg; Pao<sub>2</sub>, 87 mm Hg; bicarbonate, 46.9 mEq/L; base excess, 18.3 mEq/L; and oxygen saturation, 97.3% on 6 L/min oxygen per nasal cannula. Elevated PaCO<sub>2</sub> is often indicative of advanced disease.

Surgical history was remarkable for an uncomplicated cesarean delivery with spinal anesthesia 8 months earlier and Port-a-cath (Smiths Medical, St Paul, Minnesota) placement 2 years earlier with no anesthetic complications. No other pertinent history was obtained. Immediately preceding the procedure, the patient was pleasant and calm, on 4 L of oxygen by nasal cannula, with a hemoglobin saturation with oxygen reading (SpO<sub>2</sub>) of 96%, and respiratory rate of 18/min. Her pulse was 87/min with a blood pressure of 115/85 mm Hg. The patient was classified ASA class IV.

The patient was brought to the GI suite and a face tent with 10 L/min of oxygen flow was applied and connected to an end-tidal carbon dioxide detector. Midazolam, 2 mg, was administered intravenously (IV) in 2 divided doses. A propofol infusion was started at 20 µg/kg per minute, then titrated for comfort. Fentanyl, 50 µg, was administered IV in 2 divided doses for analgesia. The patient remained comfortable and maintained spontaneous ventilations. Jaw thrust assistance was implemented when mild airway obstruction occurred. The scope was passed down the esophagus with some coughing, but otherwise without incident. No bleeding was detected on the monitor, and the stomach was insufflated for PEG placement.

During insufflation, gurgling was heard in the oropharynx. Suctioning was employed, which returned blood, and the patient's SpO<sub>2</sub> quickly fell from a percentage in the low 90s to 60%. The scope was quickly removed and positive pressure ventilation by face mask was attempted, but the patient coughed and expectorated approximately 300 mL of blood. The SpO<sub>2</sub> reading decreased further from 20% to 30%, and the patient became cyanotic. End-tidal carbon dioxide reached more than 80 mm Hg, and the SpO<sub>2</sub> reading fell to less than 10%. It should be noted that SpO<sub>2</sub> may not be accurate below a reading of 70%. The patient's heart rhythm was sinus tachycardia (heart rate of 138/min), with a stable blood pressure of 154/88 mm Hg.

Emergent atraumatic oral intubation with a 7.0-mm ETT was performed after administration of 120 mg of succinylcholine. Blood was present in the airway, but a laryngoscopy grade I was noted. Positive end-tidal carbon dioxide was detected and bilateral breath sounds were auscultated. Ventilation with increasing positive pressure up to 50 cm H<sub>2</sub>O was difficult, as evidenced by minimal chest rise and failure to sufficiently increase the SpO<sub>2</sub> reading at 47%. A high concentration of sevoflurane was administered for bronchodilation in 10 L/min of 100% oxygen flow. Despite increasing closure of the adjustable pressure-limiting valve to 70 cm H<sub>2</sub>O, resistance to manual ventilation was sustained. Albuterol was admin-
istered and suctioning with a 14 French suction catheter was implemented through the ETT multiple times, which continued to return blood. Ventilation and SpO₂ slowly recovered from 60% to 70%. The patient was paralyzed with 30 mg of rocuronium, and support measures were continued while the rapid response team and pulmonary intensive care team were notified.

After the teams arrived, the patient was transported to the medical intensive care unit (MICU) for close monitoring, testing, and recovery. The patient had returned to baseline vital signs upon arrival to the MICU, with SpO₂ of 100% with ventilator support, pulse 108/min, and blood pressure 111/65 mm Hg.

Later that evening, the patient was taken to the interventional radiology department and found to have bleeding from the right bronchial artery, which was subsequently embolized. On the first day postprocedure the patient was stable, awake, and responded appropriately to questions, though she remained intubated. A PEG tube had been placed under conscious sedation at the bedside earlier that day, as this was considered the safest option because an ETT was in place. She was extubated on postprocedure day 2 without incident. Over the course of the next 9 days, the patient experienced intermittent periods of shortness of breath and was transferred out of the MICU. Her physicians were considering discharge in the next few days, but the patient subsequently developed a new pneumonia with mucus plugs and decompensated requiring emergent reintubation. Severe bradycardia followed, which resulted in a cardiac arrest. Resuscitation attempts were not successful. The family reportedly denied an autopsy.

Discussion
Cystic fibrosis is the most common, life-threatening inherited disease in the United States. It is an autosomal recessive disease occurring in 1 in 3,300 live births in the Caucasian race. The CF transmembrane conductance regulator gene is mutated, causing defective secretion of sodium, chloride, and water across the epithelia of the pancreas and sweat glands and the intestinal, biliary, reproductive, and respiratory systems. The osmotic gradient is affected, and viscous secretions result in obstructions in these organs. Excessive electrolyte loss by sweat glands, saliva, and tears provide diagnostic data; high levels of chloride in sweat is indicative of CF.

Clinical manifestations of CF involve many organ systems, most notably the respiratory system. In brief, the dehydrated mucous in the airways is unable to clear bacteria that would otherwise be expectorated. This causes frequent infections, inflammation, and subsequent destruction of the parenchyma, all of which lead to chronic obstruction of the airways. Eventually, airway remodeling and resistance lead to a decrease in ventilation/perfusion ratio resulting in hypoxemia, respiratory failure, pulmonary hypertension, and cor pulmonale. Pneumothorax, bronchospasm, and hemoptysis are potential complications of significance in anesthesia.

In addition to respiratory complications, several other clinically significant complications of CF are seen in other organ systems. Gastrointestinal tract symptoms in CF are due to obstruction mechanisms, which may lead to impaired bowel transit and decreased enzyme secretion. Meconium ileus and viscous mucus plugs in the small intestine commonly appear in neonates and are nearly pathognomonic for the disease in newborns. Pancreatic enzyme deficiency causes malabsorption and steatorrhea. Glucose intolerance is common. Portal hypertension and cirrhosis may develop with chronic biliary obstruction and malnutrition. Vitamin K deficiency may manifest as coagulopathy and increased risk for hemorrhage. Reproductive organ function may also be complicated by mucus obstruction; males experience obstruction to sperm flow, while females display cervical obstruction due to viscous secretions.

Patients with CF are surviving longer, many into their thirties, because of advances in medicine. Some treatment modalities include the following:

1. Supplemental nutrition (pancreatic enzyme supplements, multivitamin supplements, and feeding tube)
2. Corticosteroids for reactive airway disease
3. Antibiotics for infection
4. Postural drainage and chest percussion to loosen and clear secretions
5. Aerosol therapy and bronchodilators
6. Bilateral lung transplantation
7. Gene therapy with transfer of a normal CF transmembrane conductance regulator gene to the respiratory epithelium (proven technically possible)

Advances in therapeutic modalities continue to increase the life span of patients with CF, increasing the likelihood that they will be candidates for procedures requiring anesthesia. An understanding of the pathophysiology of the disease, as well as its anesthetic implications and management, is crucial to the safe administration of anesthesia in this population.

Preoperative evaluation should include a full preoperative assessment with particular attention to sputum production, limitations to physical activity, and lung sounds. Recommended tests include chest radiograph, pulmonary function tests, blood glucose level, sputum culture and sensitivity, liver function tests, serum electrolyte levels, and complete blood cell count. Additional tests may include coagulation labs, arterial blood gas levels, electrocardiogram, and echocardiography. Diligent postural drainage and chest percussion should be implemented along with adequate hydration and prophylactic antibiotic therapy for several days before the procedure. Opioids and nondepolarizing muscle relaxants should be avoided, if possible, to avoid depression of cough reflex and respiratory drive. Anticholinergics should be used with caution.
because of their drying effect on the respiratory mucosa. Ketamine, which causes increased bronchial secretions, and desflurane, which can irritate the airways, should be avoided. Preoperative vitamin K should be considered if a bleeding risk is present.

If the procedure allows, spontaneous ventilation should be maintained because of the limited respiratory reserve in CF patients. General anesthesia depresses the respiratory center and cough reflex, increases work of breathing, and decreases an already impaired mucociliary action. As mentioned previously, intermittent positive pressure ventilation associated with general anesthesia also increases pressure and tension in the walls of the airways and pulmonary arteries. Humidified oxygen with an FiO₂ greater than or equal to 0.50, frequent suctioning and avoidance of coughing is recommended regardless of anesthetic technique.

If general anesthesia is necessary, a volatile agent that does not irritate the airway is preferred to facilitate bronchodilation and decrease the requirement for muscle relaxants. A decreased dose of muscle relaxant is favored for early recovery. Ventilatory goals are to maintain adequate minute ventilation with the least amount of positive pressure possible. Auffed ETT enables necessary suctioning of the airways and control of ventilation. A laryngeal mask airway may be tolerated, although the risk of aspiration and airway obstruction from secretions should be considered.

Notable postoperative complications include atelectasis, pneumonia, respiratory depression, pneumothorax, and airway obstruction. Therefore, close postoperative monitoring in an intensive care unit is recommended for CF patients undergoing surgical procedures with anesthesia.

Conclusion
Several factors may have contributed to the development of hemoptysis in this case. The patient had a predisposition to bleeding, although her previous admission for arterial embolization should have repaired the source of bleeding. Increased thoracic pressure related to stomach insufflation was considered, but oropharyngeal bleeding occurred on the previous day without insufflation. There is a possibility that trauma from the first attempt to insert the tube caused the bleeding, since it spontaneously resolved, but the patient denied a sore throat before the current procedure. In this case, hemoptysis may have resulted from reflex coughing triggered by agitation with the scope or by blood in the oropharynx. If coughing preceded the event, it may have created additional pressure leading to rupture of the artery. Hypoxia was not a precipitating factor in this case, but it may contribute to an increased intravascular pressure related to the hypoxic pulmonary vasoconstriction phenomenon. During the episode, this patient may have experienced effects of Mueller maneuver (negative pulmonary pressures associated with inspiratory efforts against an obstructed airway), which may lead to increased tension within the pulmonary system. Furthermore, increased afterload and akinesis of the left ventricle are associated with this phenomenon and can have negative effects, such as ischemia, on patients with already marginal heart function. The generated pressure could transmit from the LV to the pulmonary vasculature, creating tension on the already weakened pulmonary vessels as described above.

Patients with CF face multiple risks when undergoing procedures requiring anesthesia. A complete understanding of the disease process is necessary for planning and performing anesthesia in the afflicted. Although rare, hemoptysis is a frightening and life-threatening complication of CF that anesthesia providers should be vigilant and prepared to promptly and effectively manage if encountered.

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