Lesson S42: PreAnesthetic Assessment of the Patient with Cystic Fibrosis

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Read this article, reflect on the information presented, then go online and complete the lesson post-test and course evaluation before the termination date below. (CME credit is not valid past this date.) You must achieve a score of 80% or better to earn CME credit.

TIME TO COMPLETE ACTIVITY: 2 hours
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TERMINATION DATE: April 30th, 2016

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Professional Gaps

Most anesthesiologists have a general knowledge of the management of patients with cystic fibrosis (CF) but may not be aware of the recent advances in antibiotic treatment. Anesthesiologists have the responsibility to administer antimicrobials and, as such, it is important that this group of health care workers have a clear understanding of the pathophysiology and treatment options.

Learning Objectives

At the completion of the activity, the reader will be able to:

1. Plan a perianesthetic approach for a patient with CF
2. Cite the incidence of the disease in different populations
3. Explain the function of cystic fibrosis transmembrane conductance regulator (CFTR) protein.
4. Outline the pattern of inheritance for CF
5. Identify common pathogens of pulmonary infection in CF
6. List the organ systems most usually affected
7. Tabulate the signs and symptoms of CF
8. Describe the pathology commonly seen in CF
9. Identify endocrine anomalies associated with CF
10. List commonly performed lab tests for patients with CF

Case

A 15 year old young man presented to the emergency room with severe abdominal pain of 48 hours duration. He had been diagnosed with cystic fibrosis shortly after birth. His past history was complicated by frequent respiratory infections for which he had received multiple courses of antibiotics. He was also diabetic, maintained on regular insulin. CT scan of the abdomen revealed intestinal obstruction and he was scheduled for immediate exploration.
Introduction

Cystic fibrosis is a monogenic, autosomal recessive disorder, which ultimately leads to multisystem organ dysfunction. The disorder is prevalent in about 30,000 Americans, males and females equally. As compared to 20 years ago, most infants born with CF now reach adulthood, making surgical procedures performed in this patient group more common.

The disease is a heterogeneous disorder caused by widespread dysfunction of exocrine glands and involving salivary, sweat, digestive, and pulmonary secretions. Normally, secretions from these glands are thin and slippery. In CF, a defective gene causes the body's secretions to become thick and sticky. Instead of acting as a lubricant, the secretions may obstruct tubes, ducts and passageways - especially in the pancreas and lungs. Lung pathology and respiratory failure are the two most serious consequences of CF. The disorder remains the most common lethal hereditary disorder in the Caucasian population, and in the United States it is the major cause of chronic debilitating pulmonary disease and pancreatic exocrine deficiency during the first three decades of life.1–3

The broad, variable, and sometimes confusing array of clinical manifestations of cystic fibrosis include obstructive pulmonary disease, pancreatic insufficiency, abnormally high sweat electrolyte concentrations, nasal polyps, infertility, gastrointestinal obstruction and pansinusitis. In late stages, lung, pancreas and liver transplantation may be required.

Until quite recently CF was a genetic mystery, and most people with the disease died before they reached the teen years. Isolation of the genetic basis of CF has opened the door to earlier detection. A CF gene, also called the cystic fibrosis transmembrane conductance regulator (CFTR) gene, was identified and cloned in 1989, setting the stage for gene therapy.4 Because of this finding and improved and more consistent treatments for the disease, many people with CF now live into their 40s and have fuller and more comfortable lives. There is still no cure. Mutations of the gene may be at least 3,000 years old, even though the first accurate description was made in 1988. Since that time, research has identified at least 1400 additional mutations.5–9 Cystic fibrosis represents the first genetic disorder elucidated strictly by the process of reverse genetics.

Each year approximately 3,200 Caucasian babies are born in the United States with CF. The disease is much less common among children of African and Asian descent. Two-thirds of infants born with CF are diagnosed in the first year of life.

Early Descriptions

Anderson was the first to describe the characteristic CF of the pancreas and to correlate it with the lung and intestinal disease prominent in CF.10 She hypothesized that CF was a recessive disease and used pancreatic enzyme replacement to treat affected children. She later proposed an autosomal-recessive pattern of inheritance. The generalized dysfunction of exocrine glands and the inability to clear secretions was suggested as a pathogenic mechanism of CF in 1945 by Farber.11 He introduced the term mucoviscidosis. Excessive salt loss in the sweat of children with CF was demonstrated a few years later and subsequently, the pilocarpine iontophoresis sweat test was described by Gibson and Cooke in 1959.12
Genetic Implications

In CF, a defective gene alters a protein that regulates the normal movement of salt (sodium chloride) in and out of cells resulting in thick, sticky secretions in the respiratory and digestive tracts, as well as in the reproductive system. It also causes increased salt in sweat on the skin. The prevalence of the disease varies with the ethnic origin of a population and is highest in individuals of northern and central European descent. The disease appears to have the same clinical picture worldwide. CF is most common among Caucasians and Ashkenazi Jews; one in 25 people of European descent carry one gene for CF. Life expectancy depends largely on access to health care but has been gradually increasing.

Although genetic screening for most common mutations allows detection of nearly 90 percent of CF carriers, it is not possible to detect all mutations which limits the prospect of general population screening. At present, DNA screening technologies are used for those with a positive family history.

Pathophysiology

Fundamental pathophysiologic findings in CF include the following:

- abnormal ion concentrations in the secretions from serous glands, with especially increased sodium and chloride content in sweat
- decreased water content and increased viscosity of secretions from mucus glands
- failure to clear secretions
- obstruction leading to glandular destruction
- a propensity for chronic respiratory tract colonization and infection by specific groups of bacteria.

The first two observations may be explained by abnormal cAMP-regulated chloride channel activity in CF epithelium whereas infections are secondary developments.

Ion Transport Abnormalities

A common biophysical characteristic of affected CF epithelial cells is a higher transepithelial electrical potential difference than that seen in normal epithelium.\(^{13}\) The transepithelial electrical potential difference reflects the rate of active ion transport and resistance to ion flow across the epithelium. Apical membranes of various cystic fibrosis epithelia are impermeable to the chloride ion. Since the various types of affected epithelia perform different functions in terms of electrolyte and water transport, this basic defect of chloride impermeability produces diverse effects.

The epithelium of the sweat duct and of other serous ducts is salt-absorbing. In the CF sweat duct, however, chloride is not reabsorbed because of epithelial cell membrane impermeability to chloride, and thus excessive amounts of salt are lost in the sweat. Airway epithelium normally secretes chloride and, secondarily, sodium and water onto the epithelial surface to maintain hydration. Chloride transfer into the airway lumen is limited because of underlying chloride impermeability and reabsorption of sodium from the airway surface into the cell is increased, leading to decreased salt and water content in airway secretions.\(^{14}\) Viscous, dehydrated airway secretions result.

Defective chloride transport in pancreatic ductal epithelium causes inadequate secretion of sodium bicarbonate and water into the pancreatic duct, retention of pancreatic enzymes, and destruction of
organ tissue. Production of sticky, dehydrated material in the intestines, liver, gallbladder, and genitourinary tract due to failure to secrete salt and water causes obstruction.

**Infection**

For persons with CF, the respiratory tract is the most likely site of chronic infection. The histologic picture of lung tissue of patients with CF is normal at birth.\(^1\) Endobronchial colonization begins during the first 2 years of life and initially involves the mucociliary apparatus (i.e., the cilia, a protective mucus layer, and an airway surface liquid (ASL) layer) with minimal parenchymal involvement. A synopsis of clinical and pathological observations in patients with cystic fibrosis, primary ciliary dyskinesia, asthma, and chronic bronchitis indicates that abnormalities in each compartment of the mucociliary apparatus can compromise mucus clearance and cause chronic airway disease.\(^1\) Subsequently, persistent colonization and associated peribronchial inflammation result in bronchiectasis and increased parenchymal involvement, with micro-abscess formation and focal hemorrhagic pneumonia.\(^1\)

*Pseudomonas aeruginosa* is the most common pathogen isolated in CF with colonization rates exceeding 70 percent in most reports. *Staphylococcus aureus* is frequently the initial colonizing organism, later replaced by *P. aeruginosa*. Increased rates of colonization with *P. cepacia* have been reported.\(^1\) Other organisms found less frequently include *Haemophilus influenzae*, *Escherichia coli*, *Klebsiella* species, *Proteus* species, *Serratia* species, *Actinobacillus* species, *P. fluorescens*, and *P. maltophilia*. These pathogens are usually present only transiently and are commonly supplanted by *P. aeruginosa*.

**Clinical Manifestations**

Abnormal signs and symptoms of the respiratory tract, gastrointestinal tract, and genitourinary system are seen most often. Because of multi-organ involvement, CF mimics a number of other clinical entities.

Most patients are diagnosed with cystic fibrosis intrapartum or during childhood. Typically, they present with respiratory tract symptoms such as persistent cough and/or refractory pulmonary infiltrates within the first year or two of life. Other common early gastrointestinal presentations include meconium ileus in approximately 10 percent of patients within the first days of life and subsequent steatorrhea with failure to thrive during infancy.\(^1\) In nearly 10 percent of cases with CF, however, the diagnosis is not established until adolescence or young adulthood.\(^1\)

Symptoms of CF vary, depending on the severity of the disease. For example, one child may have respiratory problems but not digestive problems, though another child may have both. Symptoms of CF may also vary with age.

The signs and symptoms of CF in children and young adults are outlined in Table 1.

<table>
<thead>
<tr>
<th>Table 1. Signs and symptoms of cystic fibrosis.</th>
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<tbody>
<tr>
<td>A salty taste to the skin</td>
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<tr>
<td>Intestinal obstruction</td>
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<tr>
<td>Foul smelling, greasy stools</td>
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<tr>
<td>Failure to thrive</td>
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<tr>
<td>Thick sputum</td>
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<tr>
<td>Chronic coughing or wheezing</td>
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<tr>
<td>Frequent chest and sinus infections, recurrent pneumonia or bronchitis</td>
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<tr>
<td>Nasal polyps</td>
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<tr>
<td>Clubbing of the fingers and toes</td>
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<tr>
<td>Intussusception in children older than age 4</td>
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<tr>
<td>Rectal prolapse</td>
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</table>
Respiratory Tract

Upper respiratory tract involvement is almost universal in cystic fibrosis. The most frequent complications are chronic respiratory infections, including pneumonia, bronchitis, and bronchiectasis. Hyperactive mucus-secreting glands that produce increased volumes of upper airway secretions, as well as edema and hypertrophy of the mucous membranes, cause chronic nasal congestion and rhinorrhea. Nasal polyps are found in 15 – 20 percent.\(^{16}\) They tend to be multiple and bilateral and often require surgery. Recurrence is common.

Progressive bronchiectasis presents in most patients over 18 months. The pulmonary course is characterized by periods of relative clinical stability, interspersed with exacerbations, typically triggered by acute infections. Progressive loss of lung function results in respiratory failure.

The earliest symptom of CF lung disease is chronic cough, initially, intermittent, dry, and associated with acute respiratory tract infection. Later it persists beyond the period of infection and becomes continuous. With time the cough becomes productive, particularly with pulmonary exacerbations, and then paroxysmal with associated gagging, choking, and vomiting, frequently worse at night and in the morning. Wheezing, rales, and rhonchi become prominent. Hyperinflation of the lungs may be noted early in the course of the disease. Atelectasis, pneumothorax, and hemoptysis are later, common complications. Pneumothorax occurs as a result of rupture of apical subpleural blebs. Life-threatening hemoptysis is a result of bleeding from eroded bronchial arteries and carries a high recurrence rate and poor prognosis.

Other clinical features of pulmonary involvement include a barrel-chest deformity, use of accessory muscles of respiration, growth retardation, hypertrophic pulmonary osteoarthropathy, digital clubbing, decreased exercise tolerance, and in end-stage lung disease, pulmonary hypertension, cor pulmonale, and respiratory failure with cyanosis.

Although antibiotics can decrease the frequency and severity of attacks, the bacteria are never completely eradicated. Growth of *Aspergillus fumigatus* in sputum may be found in up to 50 percent, and 10 percent may exhibit the syndrome of allergic bronchopulmonary aspergillosis.\(^{15}\)

Gastrointestinal Tract

Early gastrointestinal symptoms suggest the diagnosis of CF in infants and young children. Meconium ileus classically presents as intestinal obstruction with abdominal distention, failure to pass stool, and vomiting within 48 hours of birth in an infant who appears otherwise well.\(^{17}\) Approximately 50 percent of cases of meconium ileus are complicated by volvulus, atresia, and/or meconium peritonitis.\(^{17}\) Infants with cystic fibrosis are at higher risk of distal intestinal obstruction later in life.\(^{17}\) Differential diagnosis includes acute appendicitis. Another occasional cause of intestinal obstruction is intussusception.\(^{17}\) Rectal prolapse is found in approximately 20 percent.\(^{17}\) Factors associated with rectal prolapse include increased intraabdominal pressure due to distended bowel and coughing, poor muscle tone, and loss of perirectal fat that normally supports the rectum.

Exocrine pancreatic dysfunction is present in 90 percent. The deficiency of pancreatic enzymes manifests as fat and protein indigestion and results in production of frequent, pale, bulky, and foul-smelling stools. Absorption of the fat-soluble vitamins A, D, E and K is prevented. Lack of vitamin D is associated with secondary hyperparathyroidism, reduced bone mineral content, and delayed bone
maturation. Vitamin E deficiency may result in increased red blood cell destruction and neuroaxial dystrophy. Vitamin K deficiency can lead to severe bleeding as a result of hypoprothrombinemia and inadequate levels of clotting factors II, VII, IX, and X. Untreated patients develop steatorrhea, azotorrhea, and growth failure. Chronic diarrhea leads to malnutrition and further vitamin deficiency.

Endocrine pancreatic function is preserved in most patients with CF until the second or third decade of life, when frank diabetes mellitus develops (type 1 insulin dependent).\textsuperscript{18} Focal biliary cirrhosis is due to bile duct occlusion and presents as hyperbilirubinemia, ascites, and peripheral edema or massive hematemesis caused by esophageal varices.

**Renal System**

Despite earlier reports of specific renal involvement, a recent 40 year single center experience did not identify any CF specific disease or increased prevalence of renal stones.\textsuperscript{19}

**Diagnosis and Assessment**

CF may be diagnosed by several categories of testing including, newborn screening, and sweat or genetic testing.\textsuperscript{20} As of 2006 in the United States, 10 percent of cases are diagnosed shortly after birth as part of newborn screening programs. The newborn screen initially measures for raised blood concentration of immunoreactive trypsinogen. A sweat test confirms the CF diagnosis. In many cases, a parent makes the diagnosis because the infant tastes salty. Due to false positives, CF screening in newborns can be controversial and thus most states and countries do not screen routinely. Therefore, most individuals are diagnosed after symptoms develop.

Sweat testing involves iontophoretic application of a medication that stimulates sweating (pilocarpine). The resultant sweat is collected on filter paper or in a capillary tube (at least 100mg is required) and analyzed for abnormally increased amounts of sodium and chloride. Chloride sweat concentrations exceeding 60mEq/L in children and 70mEq/L in adults are considered diagnostic (normal is <30mEq/L). The sweat test is not useful in diagnosing CF heterozygotes. It does not indicate the severity or prognosis of the disease and may not be useful in newborns that produce little or no sweat.

Patients with CF have less salivary thiocyanate and hypothiocynite.\textsuperscript{21} CF can also be diagnosed by identification of mutations in the CFTR gene. Other tests (x-rays, CT scans, pancreatic and liver function, sputum analyses, and pulmonary function tests) identify complications of CF and monitor disease progression.

**Therapy**

Treatment of CF involves a comprehensive, multidisciplinary, intensive care program that is implemented in a nationwide network of specialized centers supported by the Cystic Fibrosis Foundation.\textsuperscript{22} Therapeutic goals that were formulated 40 years ago remain valid today: prevention and control of pulmonary infections, promotion of mucus drainage, and provision of adequate nutrition.

The fundamental objectives of a therapeutic plan are to maintain prolonged periods of stability and to intervene early with aggressive treatment of clinical exacerbations. (See Table 2.) Targets for therapy are the lungs, GI tract, reproductive organs (including assisted reproductive technology and psychological support).
Pulmonary Therapy

Antibiotics remain the mainstay of therapy. Although the complete sterilization of respiratory secretions is almost never accomplished, treatment aims to lower the numbers of bacteria, reduce the intensity of endobronchial infection, and delay progressive lung damage. An early and aggressive antibiotic and reduction of inflammation are the rule, and there is evidence that such treatment strategy may deter the onset of chronic colonization.\(^23\)

Antibiotic regimens are usually directed against the most commonly encountered organisms such as \(P.\ aeruginosa\), \(S.\ aureus\), and \(H.\ influenzae\). A typical intravenous antibiotic regimen consists of two agents, such as the combination of an aminoglycoside with a third-generation cephalosporin and/or anti-\(Pseudomonas\) penicillin, and/or monocyclic \(\beta\)-lactam. Also, macrolide antibiotics, while not directly effective against \(P.\ aeruginosa\), may have indirect actions against these bacteria.

The quinolones remain effective oral agents against \(P.\ aeruginosa\) pulmonary infection, but clinical usefulness is limited by rapid emergence of resistant organisms. Aerosolized antibiotics such as Aztreonam lysine are important adjuncts especially in the long-term therapy of chronic, resistant \(P.\ aeruginosa\) infections.\(^23\) However, concentrations of inhaled antibiotic delivered to the small airways are highly patient specific and varies throughout the bronchial tree.\(^24\) Dosing may have to be increased \(S.\ aureus\) and \(H.\ influenzae\) colonizations and infections are usually controlled with oral antibiotics, but intravenous semi-synthetic penicillin may be used to treat severe \(S.\ aureus\) infections. As most CF patients are on one or more antibiotics at all times, resistance develops quickly. The most frequently used antibiotics are listed in Table 3.

Eradication of methicillin resistant \(Staphylococcus\ aureus\) has been shown to be possible in some non-randomized trials.\(^25\) Further research is ongoing.

Table 2. Management and treatment of cystic fibrosis.

<table>
<thead>
<tr>
<th>Treatment</th>
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<tbody>
<tr>
<td>Antibiotics</td>
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<tr>
<td>Mucus thinning drugs</td>
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<tr>
<td>Bronchodilators</td>
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<tr>
<td>Bronchial airway drainage</td>
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<tr>
<td>Oral enzymes</td>
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<tr>
<td>Optimal nutrition</td>
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</table>
### Table 3. Commonly used antibiotics.

<table>
<thead>
<tr>
<th>Route</th>
<th>Organisms</th>
<th>Agents</th>
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<tbody>
<tr>
<td><strong>ORAL</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td><em>S. aureus</em></td>
<td>Cloxacillin&lt;br&gt;Cefaclor&lt;br&gt;Clindamycin&lt;br&gt;Erythromycin&lt;br&gt;Amoxicillin/clavulanate</td>
</tr>
<tr>
<td></td>
<td><em>H.influenzae</em></td>
<td>Amoxicillin&lt;br&gt;Trimethoprim-sulfamethoxazole&lt;br&gt;Chloramphenicol</td>
</tr>
<tr>
<td></td>
<td><em>P. aeruginosa</em></td>
<td>Ciprofloxacin</td>
</tr>
<tr>
<td><strong>INTRAVENOUS</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td><em>S. aureus</em></td>
<td>Oxacillin</td>
</tr>
<tr>
<td></td>
<td><em>P.aeruginosa</em></td>
<td>Gentamicin&lt;br&gt;Tobramycin&lt;br&gt;Amikacin&lt;br&gt;Netilmicin&lt;br&gt;Carbenicillin&lt;br&gt;Imipenem/cilastatin&lt;br&gt;Vancomycin&lt;br&gt;Piperacillin</td>
</tr>
<tr>
<td></td>
<td><em>P. cepacia</em></td>
<td>Ceftazidime</td>
</tr>
<tr>
<td><strong>AEROSOL</strong></td>
<td><em>P.aeruginosa</em></td>
<td>Gentamicin&lt;br&gt;Tobramycin&lt;br&gt;Carbenicillin</td>
</tr>
</tbody>
</table>

Chest physiotherapy remains an important part of the comprehensive treatment program. Daily regimens of postural drainage, manual or mechanical percussion, vibration, and assisted coughing promote increased clearance of bronchial mucus secretions. Physical activity programs of forced expiratory exercises and positive expiratory pressure breathing are adjuncts. Bronchodilator treatment is followed by chest physiotherapy, and then an aerosolized antibiotic is administered. Alternatively, a β2-agonist and/or cromolyn sodium can be nebulized with antibiotics. Bronchodilator treatment should be initiated with caution, however, because some patients may respond with a paradoxical decrease in expiratory flow rates and decreased PaO₂, due to increased airway collapse during expiration.

Gene therapy for cystic fibrosis lung disease is a focus of investigation. Ivacaftor is a gene-specific CF transmembrane conductance regulator potentiator that augments in vivo chloride transport in CFTR mutations that affect channel gating. Anecdotal reports of clinical improvement are encouraging.
Gastrointestinal Therapy

Patients with CF are malnourished because the pancreatic enzymes needed for digestion do not reach the small intestine. As a result they may need 50 to 100 percent more calories. Pancreatic enzyme replacement, adequate nutrition, and vitamin and mineral supplementation are indicated.

Glucose intolerance and clinically significant diabetes mellitus - occasionally seen in the second and third decades - are managed with dietary adjustments and small doses of insulin.

Preanesthetic Assessment

The clinical spectrum of patients with CF presenting for surgery ranges from an asymptomatic individual scheduled for a minor elective procedure to a moribund patient for emergency thoracotomy.

Preanesthetic assessment of the patient with CF should be directed at the most commonly affected organs and systems. The severity of pulmonary and cardiac compromise and the extent of liver involvement should be assessed by history, physical examination, and additional studies as indicated. Information elicited during the preoperative interview should include the duration of pulmonary disease, the frequency and severity of exacerbations, the quality and quantity of recent sputum production, the degree of exercise intolerance, and current medical treatment. Past surgical and anesthetic history should be discussed and available medical records reviewed.

Physical examination should focus on general nutritional status, signs of respiratory distress, respiratory rate, abnormal breath sounds, and evidence of right-sided heart failure, such as peripheral edema and hepatomegaly. Cyanosis and digital clubbing should be noted. If nasal intubation is planned, the presence of nasal polyps should be excluded. Signs of liver disease should be sought.

Laboratory data should include complete blood count, electrolyte panel, blood sugar, and coagulation profile. Baseline liver function tests should document the extent of hepatic involvement. Chest x-ray should be scrutinized for evidence of active pulmonary disease and cardiomegaly. Preoperative ECG has been recommended in all patients with cystic fibrosis. Recent spirometry tests should be reviewed to quantify the degree of pulmonary dysfunction, and arterial blood gas analysis should be obtained to determine the preoperative values of pO₂ and pCO₂.

Anesthetic Plan

The anesthetic plan for patients with CF scheduled for elective surgical procedure depends upon the extent of surgery and the age of the patient. The patient should be in optimal medical condition consistent with the stage of pulmonary disease. Parenteral vitamin K preparations are administered to patients who are not on oral vitamin K supplements. Premedication with oral benzodiazepines is safe, but opioids are avoided because of the respiratory depressant effect. Aprepitant has been used as an antiemetic. Although use of atropine has not been associated with any increase in pulmonary complications, its administration is controversial because of possible further inspissation of secretions.

Regional anesthesia is preferred for appropriate procedures, but general anesthesia can be safely administered; coagulopathy should be excluded prior to regional anesthesia. If general anesthesia (GA)
is selected, intravenous induction is preferred because pronounced V/Q mismatch may prolong inhalation induction in patients with advanced pulmonary disease. Ketamine should be avoided as it may increase bronchial secretions. Endotracheal intubation and controlled ventilation are recommended.\textsuperscript{28} Patients with mild to moderate disease do not experience significant deterioration in central or peripheral airway function following GA.\textsuperscript{29} Maintenance of general anesthesia with an inhalation agent allows the use of higher concentrations of oxygen, causes bronchodilation, and decreases the responsiveness of hyper-reactive airways. Adequate hydration and humidification of inspired gases, as well as frequent tracheal suction, are important in preventing intraoperative inspissation of secretions. Fully awake tracheal extubation is performed after appropriate criteria are met and thorough tracheal suction is accomplished.

**Management of the Case**

The patient’s lung function was assessed and considered to be optimized. Plans were made for early postoperative respiratory therapy and mucolytic therapy. A pain consult was called and a plan initiated, starting with preemptive analgesia using celebrex, gabapentin and acetaminophen. After ensuring that no nasal polyps were present, a nasogastric tube was placed. A smooth intravenous induction was achieved and the trachea successfully intubated. Anesthesia was continued with sevoflurane. An intussusception was diagnosed and required a partial colectomy. There were no complications and the patient was discharged on the 4\textsuperscript{th} postoperative day.

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REFERENCES


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Post-test

1. The prevalence of CF in the American population is:
   a. Not known
   b. 350,000 persons
   c. 30,000 persons
   d. Greatest among white males

2. The underlying defect in CF epithelium includes abnormal transport of:
   a. Potassium
   b. Glucose
   c. Sugar
   d. Chloride

3. The most common bacteria isolated from the sputum of patients with CF related pulmonary disease is:
   a. *E. coli*
   b. *M. tuberculosis*
   c. *S. aureus*
   d. *P. aeruginosa*

4. Clinical manifestations of CF are LEAST likely to be seen in the:
   a. Brain
   b. Nose
   c. Lungs
   d. Pancreas

5. Regarding inheritance of CF
   a. A dominant pattern is frequently described
   b. An autosomal recessive pattern predominates
   c. All carriers die before adulthood
   d. Only females are carriers
6. **The cystic fibrosis gene**
   a. Has undergone at least 1400 mutations
   b. Is located on chromosome 26
   c. Is a relatively recent mutation
   d. Rarely mutates

7. **The sweat test:**
   a. Is useful in diagnosing CF heterozygotes
   b. Indicates severity of disease
   c. Involves the iontophoresis of pilocarpine into the skin
   d. Always correctly diagnosis CF in infants

8. **A major pathophysiologic finding in CF is:**
   a. Increased sodium and chloride in sweat
   b. Increased water content of mucus glands
   c. Glandular hypertrophy
   d. Normal ion concentrations in serous glands

9. **Medical therapy of CF includes:**
   a. Pancreatic enzyme preparations
   b. Antibiotics
   c. Mucolytic agents
   d. All of the above

10. **Regarding dietary requirements for patients with CF:**
   a. Regular diet suffices
    b. High calorie is preferable
    c. Glucose intolerance is common in children
    d. Mineral supplementation is contraindicated