Anesthesia for cystic fibrosis patients
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The author reviews the potential risks and anesthetic complications encountered in surgical patients who have cystic fibrosis. She offers thorough guidelines for pre-, intra- and post-operative anesthetic management, including the selection of suitable agents and techniques.

Cystic fibrosis is an inherited disorder which produces chronic involvement of the respiratory system and gastrointestinal tract. It also adversely affects the liver and portal circulation. Cystic fibrosis is the number one genetic killer of children and young adults in the United States, and is believed to occur once in approximately every 1,500 live births. An estimated 4-5% of the population carries the defective gene but shows no symptoms of the disease.1

The steadily increasing life span of these patients has contributed both to the frequency of complications encountered by anesthetists and the need for specialized anesthetic and surgical management of such cases. Prolapse of the rectum is common among younger children having cystic fibrosis, and nasal polyps tend to occur in older children. Numerous therapeutic and diagnostic procedures must be performed on these children, including colostomy repair of rectal prolapse, excision of nasal polyps, bronchoscopy, lobectomy, pneumonectomy, tracheostomy, and splenoportal anastomoses for cirrhosis and portal hypertension.2

Pediatric patients with cystic fibrosis have particular anesthetic problems, thus, a thorough understanding of the pathophysiology of this disease is mandatory. This article provides a review of the potential problems and risks that anesthetists may encounter when managing the patient with cystic fibrosis.

Epidemiology
Cystic fibrosis, an autosomal recessive disease, predominates in the Caucasian race. Each parent of a child with cystic fibrosis is a "carrier" (heterozygote) of a single CF gene. The carrier, as such, shows no trace of the condition. A child born to parents with the CF gene has a one-in-four chance of getting the disease, a two-in-four chance of becoming a symptomless carrier, and a one-in-four chance of escaping both the carrier gene and the disease. When only one parent has the CF gene, none of the children will have the disease itself, but some may become carriers. The exact nature of the defect in the gene is not yet completely understood.1

Cystic fibrosis is a generalized disease of the exocrine glands, involving the respiratory and gastrointestinal tracts, and sweat glands. The sweat glands have increased electrolyte (sodium, chloride, and potassium) concentrations. Chloride concentrations in sweat are measured with the pilocarpine iontophoresis test to confirm the diagnosis of the disease.3

A new skin test to detect the cystic fibrosis carrier was reported in the January 1981 issue of
the New England Journal of Medicine. Researchers claim the new test, developed at Children's Hospital Medical Center in Boston, is highly accurate in determining which adults are likely to produce children who will have cystic fibrosis. The test consists of taking a tiny skin sample and measuring the amount of sodium that skin cells absorb when they are exposed to ouabain.

Respiratory involvement predominates in most cases and is the cause of death in 90% of cystic fibrosis cases. Pancreatic deficiency of varying severity produces digestive and nutritional problems in more than 80% of these patients.

**Signs and symptoms**

*Lung involvement.* The basic pathogenetic mechanisms which produce most of the signs and symptoms of cystic fibrosis involve obstruction of the ducts of the various exocrine glands by viscid secretions. In the lungs, this initially causes obstruction of the smaller airways, but as the disease progresses, larger airways become occluded. Symptoms may be coughing, excess mucus production, and difficult or rapid breathing which may begin as early as 3-4 months after birth.

Respiratory failure develops usually by the second or third decade of life. Even if they appear fairly healthy, all of these patients have severe pulmonary ventilation/perfusion (V/Q) inequality. There may be associated cyanosis, air trapping, slight hypoxemia, an increased anteroposterior chest diameter, and prolonged expiration.

All changes in the lungs produced by cystic fibrosis tend to be self-perpetuating. A vicious cycle is established; obstruction leads to infection and infection leads to more obstruction and increased infection. These changes lead to over-inflation of the lungs, and an increase in residual volume and the ratio of residual volume to total lung capacity. Airway resistance is increased and vital capacity and flow rates are depressed. Arterial oxygen is decreased early during the course of cystic fibrosis, but PaCO₂ does not rise until relatively late in the disease.

The patient may exhibit clubbing of fingers and toes. Susceptibility to fatigue, shortness of breath, fever, a barrel-shaped chest, more frequent cough and sputum production, and decreased appetite are indications of uncontrolled respiratory involvement. In advanced cases, hemoptysis and pneumothorax may occur. Chronic lung disease may effect changes in the heart (cor pulmonale).

Blood gas analysis and pulmonary function testing are helpful in assessing respiratory status.

Respiratory treatment for cystic fibrosis is essentially symptomatic and preventative. Methods include inhalation therapy and intermittent aerosol medication, bronchial drainage, physical therapy of the chest and breathing exercises. Antibiotics are frequently used to treat infections of staphylococcus aureus and pseudomonas aeruginosa, the two chief organisms causing respiratory damage.

*Gastrointestinal involvement.* The meconium in infants who have cystic fibrosis is abnormally viscid. Approximately 15% of these infants develop a form of intestinal obstruction called meconium ileus. These infants may present secondary to-deficient pancreatic function.

Lack of pancreatic digestive enzymes in the duodenum may result in abnormal stools and a failure to gain weight. Deficiency in pancreatic exocrine function also leads to malabsorption, steatorrhea, and various forms of intestinal obstruction. The pancreas is progressively replaced with fibrous tissue.

Many patients with cystic fibrosis have biliary cirrhosis which may not be recognized until late in the course of the disease. Initially it is focal, asymptomatic and may be associated only with an abnormal serum glutamic-oxaloacetic transaminase (SGOT). In approximately 2-3% of the patients, it progresses to a multifocal pattern. A few patients have extensive liver involvement resulting in portal hypertension, splenomegaly with hypersplenism, ascites and/or esophageal varices with gastrointestinal hemorrhage. Surgical shunting of blood flow may provide relief of symptoms. Diabetes mellitus occurs among patients with cystic fibrosis more frequently than in a control population.

*Sweat gland involvement.* The sweat and parotid glands produce secretions with abnormal electrolyte concentrations. There is excessive loss of electrolytes in sweat, saliva, and tears. Heavy salt loss, unless compensated, may result in heat prostration.

**Preoperative considerations**

The variety of organs involved in cystic fibrosis predisposes the patient to a number of surgical procedures that are peculiar to this disease. The cystic fibrosis patient may also undergo surgical procedures unrelated to the disease itself; hence, special attention to the patient's overall condition is of vital importance.

The anesthetist may want to keep the following potential problems in mind when administering anesthesia to a patient with cystic fibrosis.

1. The patient may have copious, extremely viscous secretions in the respiratory tract.
2. Because of the ventilation/perfusion inequality, hypoxemia may develop rapidly during anesthesia, and induction of anesthesia with inhalational agents is prolonged.

3. Because of reduced lung compliance, extremely high lung airway pressure may be required to provide adequate ventilation and prevent hypoxemia. Therefore, use of a cuffed endotracheal tube is advised.

4. Many children with advanced cystic fibrosis become severely emotionally upset. They require careful, considerate handling and much assurance.\(^6\)

Because of extensive pulmonary involvement in cystic fibrosis, the anesthetic management and the pre-and post-operative care of these patients may be difficult. Prior to surgery, a chest x-ray, complete blood count (CBC), arterial blood gas measurements, liver function tests, urinalysis and blood urea nitrogen (BUN) should be obtained. Detailed pulmonary function testing to determine function and reserve should be done if possible.

Elective surgical procedures should be avoided when a patient has a vital capacity less than 50\% of the normal capacity for the patient’s sex and height. Surgery should also be avoided when the PaCO\(_2\) is above 50 mmHg, and the FEV\(_1\)/FVC ratio is less than 65\%.\(^5\)

Emphasis on preoperative clearing of the secretions by postural drainage, liquification and suctioning, antibiotic therapy, and pulmonary physiotherapy is of great importance in preparing the patient for surgery.

Hyponatremia caused by an abnormally active sodium pump must be looked for so that sodium can be replaced prior to anesthesia.\(^8\)

Preoperative guidelines for the anesthetist include the following:

1. Carefully assess the patient’s physical status.
2. Do not give narcotic premedication because it depresses ventilation. Use diazepam to counter anxiety.
3. Adequate hydration is mandatory. Avoid withholding fluids for long periods. The patient should be offered clear liquids until four hours preoperatively.\(^6\)
4. Antibiotics should be given for two days prior to the operation and continued until the patient is able to cough well and participate actively in postural drainage.
5. Postural drainage should be instituted upon admission to the hospital, and usually involves as many as four treatments per day with all segments of the lungs treated.

6. Infants with cystic fibrosis, especially those less than a year old, have an increased risk for hemorrhagic phenomena due to vitamin K deficiency (because of decreased gastrointestinal synthesis of vitamin K and malabsorption of fatsoluble vitamins). Therefore, infants (and possibly all patients) with cystic fibrosis undergoing surgical procedures should be given vitamin K.\(^5\)

Anesthetic management

It must be kept in mind that the cardinal rules for managing these patients are the prevention of respiratory depression and an avoidance of inspissation of tracheobronchial secretions. Therefore, agents such as narcotics, nondepolarizing muscle relaxants and cholinergics should be employed only with good clinical judgment.\(^5\)

Premedication and anesthetic agents should be carefully selected. Many clinicians believe that atropine should be avoided in cystic fibrosis cases because it dries the respiratory mucosa and causes more tenacious secretions. Generally, narcotics should be avoided for premedication and during the anesthesia administration. Nondepolarizing muscle relaxants should also be avoided when possible. These drugs suppress both ventilation and the cough reflex, and their effects may last into the postoperative period.\(^5\)

Prior to induction, preoxygenation with 100% oxygen by mask for at least five minutes is beneficial. The increased ventilation-perfusion abnormalities that occur in cystic fibrosis cause the induction of anesthesia with inhalation anesthetic agents to be slower than usual. Therefore, induction may be aided by use of an intravenous agent such as thiopental, which can be followed by an inhalation agent such as halothane.\(^8\) Unless one is careful to reduce the concentration of the inhalation agent well before the end of the operation, awakening and return of respiratory control may be considerably delayed.\(^2\)

Ketamine usage in cystic fibrosis patients has an increased risk because of severe and prolonged airway irritation.\(^2\)

Endotracheal intubation is advocated unless the operative procedure is brief. Normal responses to general anesthesia include depression of the respiratory center and cough reflex, increased work of breathing, and decreased ciliary action. In patients with cystic fibrosis, these normal responses may lead to complications. Intubation provides better control of the airway and facilitates tracheal suction as does assisted ventilation.\(^5\)

Removal of mucopurulent secretions is essential with whatever anesthetic agents are used.
It cannot be stressed enough that prime requisite of the anesthetic management in these cases is that the anesthetist must be able to remove airway secretions effectively and quickly at any time.

Thus, it is particularly important to avoid the use of curved endotracheal tubes or adaptors. One should take advantage of the opportunity near the end of the anesthetic effect to clear the trachea and main bronchi while the patient is still asleep. The thick secretions are always difficult for patients to expel by themselves. These secretions become even more dangerous in the postoperative period when they are more tenacious and patients have less ability to cough them out.

Because of these respiratory problems associated with patients who have cystic fibrosis, the suggestion to use spinal anesthesia is frequently made. However, spinal anesthesia in cystic fibrosis cases carries an important risk. The cough reflex of these patients is extremely sensitive. If spinal anesthesia is accompanied by violent and prolonged coughing by the patient, the level of anesthesia may reach to the upper thorax or above, which obviously is a situation dangerous to the patient.2

Of chief concern is the prevention of irritation and coughing during induction and again as the patient awakens; continued violent coughing may produce extensive emphysema. Sedation and generous spraying of the patient's throat with 4% lidocaine are helpful, but the situation is not easily controlled and may be extremely unpleasant for the patient.3

Assisted ventilation during surgery is advised because when unassisted, the tidal volume tends to decrease during anesthesia, and controlled ventilation may cause hyperventilation and hypocarbia. This could result in hypoventilation during postoperative recovery.4

It is recommended that oxygen should constitute at least 50% of the total gas mixture to prevent arterial hypoxia. Keeping the patient in a light plane of surgical anesthesia is a safe means to assure that the patient will be awake and extubated at the end of the procedure, will be breathing spontaneously and will have full reflex activity.

During the course of the surgical procedure, extensive and frequent aspiration of secretions in the tracheobronchial tree should be performed. High humidity anesthesia systems are desirable. Before the endotracheal tube is removed, extensive aspiration should again be accomplished.

In the postoperative period, arterial blood gases can be monitored to evaluate respiration. Postural drainage should be instituted as soon as possible after the operation. Atelectasis and pneumonia are frequent postoperative complications.5

In summary, the anesthetist must recognize the potential risks and anesthetic considerations in order to manage the patient with cystic fibrosis. A thorough understanding of the epidemiology of cystic fibrosis is most important.

REFERENCES


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