Cystic Fibrosis Perioperative Database Review

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Summary

CF is the most common fatal genetic disorder and most common cause of chronic lung disease in children and young adults

CF patients often undergo surgical procedures to address the effects of CF

Airway, Head and Neck, and Gastrointestinal Endoscopy were the three most common procedure categories

There is scant literature regarding perioperative management of pediatric CF patients

Background

Cystic fibrosis (CF) is an inherited autosomal recessive disease affecting mostly Caucasians. Approximately 1 in 30 Caucasian patients carry the CF mutation. CF is the most common fatal genetic disorder and most common cause of chronic lung disease in children and young adults. A mutation on the long arm of chromosome 7 causes CF, resulting in chloride channel dysfunction which impacts multiple organ systems (e.g. respiratory, gastrointestinal, etc.).

CF patients undergo a variety of surgical procedures to address the effects of the disease process: endoscopic sinus surgery, central line access placement, gastroscope tube insertion, chest imaging, bronchoscopy, and liver and lung transplantation. Despite the common nature of this disease, there exist very few perioperative studies that examine the ideal preoperative optimization, effects of intraoperative anesthetic management, and post-operative outcomes of pediatric patients who have CF.

Methods

We performed a retrospective review of de-identified perioperative data to ascertain the demographic distribution of the CF surgical population at our institution. We accessed the anesthesia information management system database and extracted de-identified demographic data from a period that ranged from January 1, 2000 until June 30, 2012 and consisting of the following patient data fields: age, weight, American Society of Anesthesiologists Physical Status (ASA PS), procedure name, and procedure type.

Results

The surgical procedures performed on CF patients at our institution were equally distributed across both males (297/594) and females (297/594). Patient ages at time of surgery ranged from 4 days to 24 years, with half of all cases occurring in children ages 11 and younger. The distribution of patient ages appears bimodal with both teenagers and children under 2 years old undergoing the most procedures. The weight of the patients generally correlated with patient age.

Thirty-two different categories of surgical procedures were identified. The most common categories were Airway, excluding Tonsillectomy and Adenoidectomy (111 cases), Head and Neck (65 cases), Gastrointestinal Endoscopy (60 cases), Interventional Radiology (58 cases), and Intra-abdominal excluding hernias (54 cases).

The most common surgical procedures for CF patients were in the following categories: airway (excluding tonsillectomy and adenoidectomy), head and neck, gastrointestinal endoscopy, interventional radiology, and intra-abdominal (excluding hernias).

Discussion

Our findings are similar to those of prior reports in adults. This review is the first step in a study that will merge the local and national CF databases with our anesthesia information management system database. We plan to elucidate the preoperative optimization of CF patients and study the effects of intraoperative anesthetic management on CF patients. Post-operative outcomes will also be examined.

Additional future plans include using the comprehensive retrospective review to design prospective studies to determine the ideal optimization plan of pediatric CF patients who are scheduled to undergo general anesthesia.

References


Figure 1. Age distribution of CF patients undergoing surgical procedures during the past 12 years at our institution

Figure 2. Most common surgery types for CF patients undergoing surgical procedures over the past 12 years at CHOP

Figure 3. Cystic Fibrosis - Most Common Surgery Types

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