Case Report

### Pulmonary Lobectomy in Adult Cystic Fibrosis Patients with Focal Bronchiectasis

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### Background

Current literature suggests a lobectomy is indicated in pediatric cases of focal lung bronchiectasis refractory to medical treatment, but there is only minimal data on the value of surgery in adults.

### Objective

The aim of these case studies is to demonstrate the efficacy of a lobectomy in adult cystic fibrosis (CF) patients with persistent lung disease.

### Methods

Two patients, 28 and 30 years old respectively, underwent a left upper lobectomy to remove a focal area of bronchiectasis. Both patients endured a long history of acute pulmonary exacerbations treated with antibiotics and had localized bronchiectasis secondary to CF. Pseudomonas was cultured in both patients while methicillin-resistant Staphylococcus Aureus grew in the sputum sample of patient one.

### Results

Patient number one developed a bronchopleural fistula postoperatively and required bronchial stent placement to exclude the fistula. The fistula ultimately resolved without further intervention. She also was able to be weaned off antibiotics within 2 weeks after surgery. Patient number two was on IV antibiotics for 211 of 500 days prior to admission. Postoperatively she was able to be weaned off antibiotics in 3 weeks.

### Conclusion

After lobectomy both patients were able to be weaned off antibiotics and experienced a decrease in acute exacerbations leading to hospitalization. More data is needed in the adult patients to determine whether this procedure is more beneficial than conservative medical therapy in patients with localized bronchiectasis.

### Keywords:

Cystic Fibrosis; Lobectomy; Bronchiectasis

### Abbreviations:

CF: Cystic Fibrosis; CFTR: Cystic Fibrosis Transmembrane Receptor; CT: Computerized Tomography; MRSA: Lmethicillin-Resistant Staphylococcus Aureus

### Background

Cystic fibrosis (CF) is an autosomal recessive disease that results in the complete absence and dysfunction of the cystic fibrosis transmembrane receptor (CFTR). CFTR is a chloride channel that regulates the flow of chloride ions between intracellular and extracellular spaces [1].

In the lungs, CF causes thick mucus to be overproduced by goblet cells within the epithelium. The excessive mucus production results in an increased propensity for bacterial infection in the airways. In addition, elevated levels of inflammatory cytokines, such as IL-1, IL-6, IL-8 and TNF-alpha increase the amount of inflammation in the lung [2]. These processes lead to bronchiectasis and increase patient susceptibility to infection. The common organisms found in CF patients are Burkholderia cepacia, Staphylococcus aureus, Haemophilus influenzae, and Pseudomonas aeruginosa [3].

The improvement of medical treatment for patients with CF has reduced the need for surgical management of pulmonary complications such as massive hemoptysis or recurrent pneumothorax. In rare cases, however, a patient with a focal area of consolidated lung or bronchiectasia may benefit from lobectomy. The majority of the literature has focused on the pediatric population, but chronic focal areas of destroyed lung are a problem in the adult population as well.

### Case 1

A 30-year-old female with a history of CF and chronic bronchitis had been hospitalized multiple times with episodes of recurrent pseudomonas bronchitis and CF exacerbations. During the most recent previous hospitalization she was begun on intravenous piperacillin/tazobactam, and amikacin. She underwent this treatment for 8 days but ultimately developed an allergic reaction and discontinued the antibiotics. The patient was discharged to home but was readmitted 3 weeks later with a productive cough and...
worsening shortness of breath. Computerized tomography (CT) of her chest on hospital day two showed a dense infiltrate in the left upper lobe but was relatively unremarkable otherwise (Figure 1). Sputum cultures obtained at this time were negative for acid fast bacilli but were positive for methicillin-resistant Staphylococcus aureus (MRSA) and Pseudomonas.

On hospital day five she underwent a left upper lobectomy. To remove the lobe, a muscle-sparing posterolateral thoracotomy was performed, sparing the serratus anterior muscle. There were dense adhesions of the lung to the chest wall, requiring an extra-pleural dissection to mobilize the lobe. The bronchus did not appear inflamed and was divided using a linear stapler. No buttressed flap, such as intercostal muscle or pericardial fat pad, was used to cover the bronchial stump. Estimated blood loss was 300ml.

Postoperatively she was placed on tobramycin, azithromycin and linezolid. Chest tubes were removed on postoperative day 16. On postoperative day 21, however, she experienced tachypnea and shortness of breath. Chest x-ray showed a large pneumothorax with contralateral mediastinal shift. Tube thoracostomy was performed immediately, with improvement of symptoms. Bronchoscopy revealed a bronchopleural fistula at the right upper lobe bronchial stump. A bronchial stent was placed in the main-stem bronchus to exclude the fistula. She was ultimately discharged on postoperative day 22 on oral clindamycin and linezolid for 2 weeks. She was weaned off antibiotics 2 weeks after the surgery and has not required antibiotics since.

The bronchial stent was removed 5 weeks after placement. No repeat stent was performed and the fistula resolved without any further intervention. No further pulmonary function testing was performed, as she had improved so significantly after surgery.

Case 2

A 28-year-old female with homozygous delta-F508 CF and a past medical history of failure to thrive was admitted to the hospital with recurrent episodes of pneumonia during the last 18 months and evidence of left upper lobe consolidation. She required intravenous antibiotics, using piperacillin/tazobactam and tobramycin, for 211 days out of 500 days prior to surgery for symptom control's scan of the chest on hospital day 1 showed extensive interstitial fibrosis and a large area of consolidation in the anterior aspect of the left upper lobe. She underwent an elective left upper lobectomy, also utilizing a muscle-sparing posterolateral thoracotomy. During the surgery, the patient received a dose of vancomycin, piperacillin/tazobactam and tobramycin (400mg). Estimated blood loss for the surgery was 200 ml. She had a large pneumothorax with a contralateral mediastinal shift. Tube thoracostomy was performed, as she had improved so significantly after surgery. Postoperative pulmonary function testing was not performed, but she had improved functionally substantially prior to her final exacerbation.

Discussion

CF is among the most common causes of diffuse bronchiectasis in adults in the United States. The improvement of medical treatment has virtually eliminated the need for surgical intervention. In rare cases, however, a patient with a focal area of consolidation may benefit from lobectomy. Current literature suggests that lobectomy carries a significant risk of morbidity and mortality in CF patients, especially in those with a forced expiratory volume below 40 percent. The procedure should be considered only after at least six months of unsuccessful medical management [4]. Though surgery can be technically challenging, as it was in each of our cases, lobectomy can be performed safely and complications can be treated ultimately with a successful outcome. As the lifespan of patients with CF continues to increase, it may become more common to see patients who would benefit from lobectomy.

The majority of literature published on thoracic surgery for CF has involved the pediatric population, but we have described two cases of successful lobectomies for the management of persistent lung infection in adult patients. Similar to previous studies in the pediatric age group, we offered surgery only to patients with a focal area of damaged lung and prolonged need for antibiotic treatment without clinical improvement. Our first patient required intravenous antibiotic treatment for over 200 days in a 1.5-year span but was able to be taken off antibiotics only 2 weeks after lobectomy. Our second patient experienced significant symptomatic relief and was also able to be taken off antibiotics completely. Antibiotics are typically used for pulmonary exacerbations of the disease and are discontinued when symptoms resolve. If patients are appropriately selected for surgery, then lobectomy can eliminate the need for antibiotics and significantly improve quality of life in some patients with CF.

It is important to stress that the majority of patients with CF should not have surgery to treat bronchiectasis, and that only a small fraction of patients should be considered. Select patients for surgery should have a focal area of diseased lung limited to one lobe [5], FEV greater than 40%, and should have had at least 6 months of antibiotic treatment with failure of resolution of the diseased lung [6]. During surgery bronchopleural fistula is a major risk, as the bronchus will usually be quite inflamed and the overall surgical field will be infected. For this reason, we recommend covering the bronchial stump with a vascularized flap, such as intercostal muscle. Though we did not perform an intercostal muscle flap in either case, we feel that doing so would have prevented the bronchopleural fistula in our second case and we will do so going forward in all of our future lobectomies in patients with CF.

More data is needed in the adult population to determine the efficacy of lobectomy compared to standard medical therapy for the treatment of localized bronchiectasis secondary to CF. In select patients, however, lobectomy may allow a patient to experience a significant improvement in quality of life and reduction or elimination of symptoms.
References