A 22 year-old female with CF (F508del/G551D), chronic hypoxemia requiring supplemental oxygen, *Burkholderia cenocepacia*, chronic sinusitis, malnutrition (BMI 16) with pancreatic insufficiency, and gastroesophageal reflux presented to another facility following the acute onset of left sided chest pain and shortness of breath. She had severe obstructive lung disease (FEV$_1$ 0.71, 21% predicted), a history of two episodes of distal intestinal obstructive disease (DIOS), frequent pulmonary exacerbations requiring hospitalization which had decreased in frequency following initiation of ivacaftor, and a right spontaneous pneumothorax two years ago. At the other facility, the patient was diagnosed with a small left sided pneumothorax. She was treated with high flow oxygen and intravenous antibiotics, and was transferred to our hospital by ambulance for further care including re-evaluation for lung transplantation.

Following transfer, the patient developed increased chest pain, chest tightness and dyspnea. Her pneumothorax had increased in size and a pigtail catheter was inserted into the left chest under CT-guidance. Despite this intervention, the pneumothorax did not fully resolve and a chest tube was inserted into the left chest four days later. Following insertion of the chest tube, the pneumothorax improved radiographically but a large air leak persisted consistent with a bronchopleural fistula. At this time, a multidisciplinary discussion ensued regarding management options of long term pleural drainage: endobronchial valve placement, video-assisted thoracostomy (VATS) or open thoracotomy for pleurodesis, bullectomy, and wedge or lobe resection. The decision was made to forego any invasive procedure and to continue treatment with chest tube drainage while waiting for lung transplant. Fortunately, donor lungs became available nine days after placement of the chest tube and she underwent successful double lung transplantation. She was discharged on post-op day 40 and remains well 14.5 months after transplant.

In contrast, an 18 year-old male with CF, severe obstructive lung disease and recurrent pneumothoraces s/p VATS with surgical pleurodesis, and chronic MRSA and *Achromobacter xylosoxidans* presented to our facility one month prior with another recurrent pneumothorax and underwent right thoracotomy with bullectomy and wedge resection. His post-operative course was complicated by hemotherax and acute/chronic respiratory failure requiring tracheostomy. A chest tube for drainage remained in place until donor lungs became available two months later and he underwent successful double lung transplantation. He was discharged on post-op day 36 and is well 16 months after transplant.

**Discussion Points:**

1. Pneumothorax is a common complication of CF (3.4% of all patients will have a pneumothorax during their lifetime). Many patients will have a recurrent pneumothorax (50 – 90%) or develop a contralateral pneumothorax (46%). The main risk factor is severe obstructive lung disease. Boat et al. reported in a retrospective review that up to 15% of patients may be asymptomatic at the time of diagnosis. Pneumothorax must be considered in all CF patients with severe lung disease given its serious consequences.

2. The treatments for pneumothorax are broad, from simple observation to partial or complete lobe resection. Multiple factors have to be considered in making management decisions including the size and cause of the pneumothorax, history and treatment of prior pneumothoraces, comorbidities and pulmonary reserve of the patient, use of invasive or noninvasive ventilation, goals of care, and the patient’s current clinical status. Prospective studies regarding the management of pneumothorax in CF have not been performed, and we must rely on retrospective reviews and expert recommendations to guide our decision-making.

3. Lung transplantation may be more difficult following thoracic surgical procedures due to adhesions and increased bleeding. Although prior surgical procedures are not a contraindication to transplantation, a patient’s candidacy for lung transplantation should be considered in the decision-making regarding management of pneumothorax in patients with end stage disease or respiratory failure. Risk of morbidities and complications of surgical intervention short of transplant must be considered against the option of long term tube thoracostomy for bronchopleural fistula.

4. Although many patients with CF will present with a pneumothorax or recurrent pneumothoraces, it is important to consider the individual characteristics of a patient prior to making any decisions regarding management, and to consider the implications of pneumothorax for prognosis.

**References:**

4. Flume PA, Mogayzel PJ Jr, Robinson KA, Rosenblatt RL, Quittell L, Marshall BC; Clinical Practice Guidelines for Pulmonary Therapies Committee; Cystic Fibrosis Foundation Pulmonary Therapies Committee. Cystic fibrosis pulmonary guidelines: pulmonary complications: hemoptysis and pneumothorax. Am J Respir Crit Care Med. 2010 Aug 1;182(3):298-306.