



C47. CASE REPORTS: PEDIATRIC CYSTIC FIBROSIS, LUNG INFECTION, AND SLEEP DISORDERED BREATHING

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Management of Persistent Air Leak Using Negative Pressure Ventilation in the Pediatric Intensive Care Unit - Case Report and Review of Literature

M. Patel¹, L. Guglani¹, M. DeAlmeida², D. Simon¹; ¹Division of Pulmonology, Allergy/Immunology, Cystic Fibrosis and Sleep (PACS), Dept of Pediatrics, Emory University, Atlanta, GA, United States, ²Pediatric Intensive Care Unit, Emory University, Atlanta, GA, United States.

Corresponding author's email: mpatel1903@gmail.com

Introduction Development of pneumothorax and persistent air leak is one of the recognized complications in Cystic Fibrosis (CF) patients with advanced lung disease. Use of positive pressure ventilation in the Intensive Care Unit can prevent closure of bronchopleural fistula and lead to persistent air leak. We describe a CF patient with acute respiratory failure and persistent air leak who was successfully treated with negative pressure ventilation (NPV). **Case Report** A 16-year-old female CF patient with past history of *Mycobacterium avium-intracellulare* infection and Allergic Bronchopulmonary Aspergillosis (ABPA), was admitted for an acute pulmonary exacerbation with fever, malaise, worsening dyspnea despite treatment with oral antibiotics as outpatient. On day 3 of admission, she developed severe hemoptysis requiring bronchial artery embolization and was started on supplemental O₂ via nasal cannula post operatively. Her respiratory status deteriorated rapidly on day 12 of admission with new onset fever and Chest Computed Tomography (CT) showed rapidly progressive invasive endobronchial infection and massive cystic bronchiectasis with nodular opacities and abscesses in the parenchyma (Figure 1A). Despite expanded antibiotic and antifungal coverage, she developed left sided pneumothorax (Figure 1B) on day 22 and required intubation for positive pressure ventilation (PPV) along with chest tube placement. By day 27, she was extubated to Bilevel Positive Airway Pressure (BiPAP) support. By day 38 there was no left lung re-expansion and pneumothorax was still present despite a second chest tube being placed. At that time, patient was taken off BiPAP support and placed on NPV with Cuirass Ventilator using Continuous Negative Pressure (CNEP) mode. By day 50, her pneumothorax showed resolution and one of the chest tubes was removed followed by the removal of the second chest tube 2 days later (Figure 1C). This patient was transferred to another institution on continued NPV for listing for lung transplant due to significant decline in respiratory status. **Discussion** This is the first report highlighting the use of NPV for management of persistent air leak in a CF patient with acute respiratory failure requiring respiratory support. Because of her acute respiratory failure, she was a poor surgical candidate which precluded most surgical interventions. Continuation of conventional PPV would not have allowed the bronchopleural fistula to heal. Besides improving pulmonary blood flow and reducing the risk of iatrogenic infections, NPV provides effective respiratory support and should be considered in pediatric patients where PPV may be detrimental including in cases of persistent air leak.