Introduction: Mounier Kuhn (MK) syndrome is a rare disorder of unknown etiology characterized by marked dilatation of trachea and bronchi secondary to atrophy of the elastic and smooth muscles of the tracheobronchial tree1. We report a patient with MK syndrome who underwent bilateral lung transplantation and outline challenges in sequential lung isolation.

Case Summary: 51-year-old male with a history of severe bronchiectasis, recurrent respiratory infections was presented for bilateral pulmonary transplantation. Preoperative workup revealed severe obstructive airway disease along with tracheomegaly (35mm) and bronchial dilations of the left and right main bronchi in excess of 26 mm meeting the basic criteria for MK syndrome2 was identified in thoracic computerized tomography fig 1. His cardiac function was normal. In view of his large size trache and bronchi, difficulty in lung isolation and possibility of requirement of cardio pulmonary bypass was discussed with the patient and the surgical team.

Anesthetic induction and intubation was uneventful, we use 10mm endotracheal tube and 25cc air was required to achieve minimal leak, intracuff pressure was monitored at regular intervals to prevent tracheal mucosal injury. Fiberoptic bronchoscopy (FOB) revealed floppy tracheobronchial wall with significant movement of folds in the wall of both bronchi. We were unsuccessful in isolating right lung with appropriate size bronchial blocker though the position was confirmed with FOB and we noticed slipping of the bronchial blocker with every breath. We decided to advance the endotracheal tube into the left main bronchus and the right lung was implanted successfully. In view of new anastomosis in the right main bronchus we were unable to place the endotracheal tube into the right main bronchus for isolating left lung. After unsuccessful attempts with bronchial blocker on the left side and in view of unstable hemodynamics and respiratory acidosis, cardio pulmonary bypass was instituted to complete left lung implantation. The rest of the procedure was uneventful.

Discussion: To our knowledge little is known about the anesthetic management in MK Syndrome for lung transplant and lung isolation. There is a concern of tracheal ischemia in patients with large trachea following intubation with larger size cuffed endotracheal tube3. Author suggested to use uncuffed tubes with throat pack to minimize the leak around endotracheal tube in tracheomegaly patients who needs mechanical ventilation, not feasible in our case4. Intubating with two medium size endotracheal tubes and placing one each into the main bronchi is an option to avoid cardiopulmonary bypass, but unfortunately there was no report of anesthetizing using two endotracheal tubes. We want to emphasize detailed communication with the surgical team and be prepared for cardiopulmonary support, for rare cases such as this, especially if they are scheduled for emergency procedures.

References: