Anesthetic Considerations for Bilateral Lung Transplantation in Mounier-Kuhn Syndrome

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MOUNIER-KUHN SYNDROME (MKS), or tracheobronchomegaly, is a rare disorder of unknown etiology that is characterized by marked dilation of the trachea and major bronchi, due to severe atrophy and thinning, as well as absence of elastic fibers and smooth muscular fibers of the large airways.[1] This syndrome commonly presents in men in the third or fourth decade of life. It is marked by weakness of the membranous and cartilaginous parts of the trachea and main bronchi leading to an insufficient cough, mucous retention, and tracheobronchial wall collapse during respiration. Symptoms can range from well-preserved lung function to refractory respiratory failure. There are fewer than 100 case reports in the literature with only 2 reports of successful lung transplantation for patients with this syndrome.[1–3] Herein, the authors present a case of a patient diagnosed with MKS presenting for bilateral lung transplantation and describe the one-lung ventilation (OLV) strategies that were attempted and utilized.

CASE REPORT

A 51-year-old male (87.8 kg, 178 cm) with a history of MKS and bronchiectasis presented for bilateral pulmonary transplantation. His past medical history was significant for hypertension, depression, and anxiety. He had a negative workup for cystic fibrosis and a negative evaluation for immunodeficiency. Preoperative arterial blood gas analysis showed mixed respiratory acidosis and metabolic alkalosis (pH: 7.40; $PaCO_2$:51; PaO_2: 114; HCO_3-: 41; SaO_2: 98%). Lung function tests demonstrated a severe obstructive ventilator defect with a possible concomitant restrictive ventilator defect; FEV_1: 1.04 (27% predicted), FVC: 3.05 (60% predicted). Computer tomography confirmed diffuse bronchiectasis with bronchial thickening and extensive mucous plugging. His echocardiogram revealed mild left ventricular dysfunction with mild enlargement of the right ventricle and no significant valvular dysfunction.

Preoperative tracheal computer tomography reconstruction showed the maximum tracheal diameter of 2.58 cm (sagittal) and 4 cm (transverse) while the left and right mainstem bronchus were 3.08 cm and 4.24 cm at their greatest diameter, respectively. For an adult, any diameter of the trachea (transverse), right main bronchus, and left main bronchus that exceeds 3.0 cm, 2.4 cm, and 2.3 cm, respectively, on a chest radiograph is indicative of MKS.[4] Preoperative bronchoscopy demonstrated a severely dilated and ectatic trachea and mainstem bronchi with hyperdynamic collapse with expiration (Figs 1 and 2).

This patient underwent bilateral, sequential lung transplantation via a clamshell incision. As anesthetic and surgical teams were aware of the challenges with lung isolation, the decision was made to proceed with lung isolation with back-up cardiopulmonary bypass to facilitate lung transplantation. The patient was intubated with a 10-mm single-lumen endotracheal tube (Teleflex Medical, Research Triangle Park, NC), and 25 mL of air were required to achieve minimal leak with acceptable cuff pressure (18-20 cm H_2O). Prior to intubation, available double-lumen tubes (size 39 and 41F) were tested and found not to have sufficient bronchial cuff balloon size diameter to occlude the corresponding bronchi.

A size 9-French Fuji bronchial blocker (Fuji Systems Corporation, Bunkyo-ku, Tokyo) was chosen for isolation of the right lung while the right lung was transplanted. One-lung ventilation was initiated, but because of the fish-mouth shape of the bronchi, successful lung isolation was marginal and there continued to be a small persistent leak at the apices of the Fuji blocker in the bronchi (Fig 3) although the cuff was filled with 20 mL of air, far in excess of the 8 mL recommended by the manufacturer. However, the authors thought that pressure would not be expected to transmit to the bronchial wall since a seal was not obtained. The bronchial blocker was repositioned numerous times, but complete isolation was never accomplished. The Arndt blocker also was attempted unsuccessfully to isolate the right lung for the same reasons. Then, the blocker was removed, and the endotracheal tube was advanced to the left mainstem bronchus as an isolation technique. The right lung was successfully implanted using this technique.

Due to unsuccessful attempts of right lung isolation by 2 different bronchial blockers, the authors decided not to utilize bronchial blockers to isolate the left lung for left lung transplantation. Although there had been success on the left bronchus, the authors did not feel comfortable attempting to place the endotracheal tube in the right main bronchus for concern of potentially disrupting the newly completed anastomosis of the right bronchi. It was decided that the surgeon would occlude the left main bronchus with a vascular clamp to prevent an air leak, and the left lung was transplanted using cardiopulmonary bypass. Cardiopulmonary bypass time was 57 minutes. Further course was uneventful except for persistent air leaks due to bronchial size mismatching at the anastomosis sites observed on bronchoscopy, but eventually, air leaks resolved, and he was discharged home on postoperative day 16.

DISCUSSION

Tracheobronchomegaly was first described by Mounier-Kuhn in 1932.[6] It is characterized by severe atrophy of longitudinal elastic fibers with thinning of the muscularis

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Fig 1. Bronchoscopy image of the trachea looking at the carina. Note the extensive dilation of tracheal lumen as well as distally into the mainstem bronchus. Also observe the significant amount of redundant, hyperdynamic mucosa.

mucosa, which results in dilation of the membranous and cartilaginous portion of the trachea and main bronchi.[1] The increased compliance of the walls allows the development of broad diverticulum like protrusions of redundant musculomembranous tissue between cartilaginous rings. Dynamic studies demonstrate marked flaccidity of the trachea and main bronchi. With deep inspiration, the airways can distend to great proportions and then on expiration can markedly collapse. During forced expiration, or on coughing, the trachea and main bronchial lumens can occlude completely, predisposes patients to chronic, recurrent pulmonary infections.[6] The diagnostic criteria for MKS involve chest radiography and computer tomographic scanning that reveals any diameters of the trachea, right main bronchus or left main bronchus that exceed 3.0, 2.4, or 2.3 cm, respectively.[2,5]

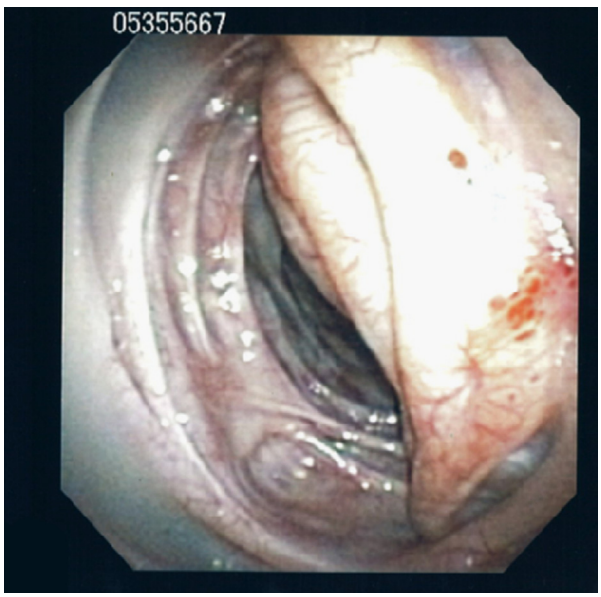

Fig 2. Bronchoscopic image of the Left mainstem bronchus. Again, notice the dilated bronchi as well as the redundant, floppy mucosa.

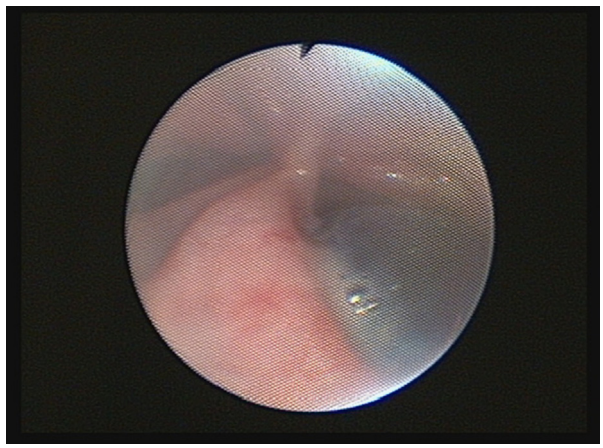

Fig 3. Bronchoscopic image during blocker placement. Note that the Fuji bronchial block is in the right mainstem bronchus, inflated with 20 mL of air, and there is still area at the 11 o'clock position of the lumen where the cuff does not provide a seal because of the oblong shape of the bronchi. Also, observe the copious secretions around the balloon.

For anesthesiologists, the major concern with MKS is airway management in terms of ability to secure an airway and to adequately ventilate the patient. For lung transplantation, in particular, lung isolation is one of the challenging tasks in MKS. In those situations alternative options such as extracorporeal membrane oxygenation or cardiopulmonary bypass should be considered to maintain oxygenation and metabolic status. Although there is no randomized trial that supports utilization of CPB, it has significant benefits in certain clinical conditions such as severe pulmonary hypertension and requirement for concomitant cardiac repair.[7] The authors had extensive discussion with the surgical team on using CPB as an alternative plan, considering some of deleterious effects such as bleeding, graft failure, and pulmonary reimplantation responses etc.[7]

Preoperatively, computed tomography should be reviewed for areas below the vocal cords that appear nondilated for positioning of the endotracheal cuff. Inability to achieve an effective seal could lead to ineffective positive-pressure ventilation yielding large airway collapse and obstruction. Further, an incomplete seal risks pulmonary aspiration while overinflation could lead to injury to the already thin and delicate tracheal walls. It was established that the cuffs on a conventional 39F or 41F double-lumen tube could not inflate sufficiently to achieve an adequate seal. To facilitate lung isolation, a single-lumen large-size endotracheal tube (ETT) and bronchial blocker were the initial plan by the authors. The ETT was placed without difficulty, and it was inflated with an excessive volume of air