CASE REPORT

Anesthetic Considerations for Bilateral Lung Transplantation in Mounier-Kuhn Syndrome

Sudhakar Subramani, MD, DNB, MMed, Brenton Freeman, DO, and Srinivasan Rajagopal, MD

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. 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; PaCO2: 51; PaO2: 114; HCO3-: 41; SaO2: 98%). Lung function tests demonstrated a severe obstructive ventilator defect with a PEF: 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 bronchi 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 H2O). 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 mucosa, which results in dilation of the membranous and cartilaginous portion of the trachea and main bronchi. 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. 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.

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. 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.

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.
to achieve a seal, warranting measurement of the cuff pressure at regular intervals to avoid mucosal injury. Bourne et al described a similar technique using a substantial volume of air in the endotracheal cuff. To minimize tracheobronchial mucosal injuries, Messahel recommended using uncuffed tubes in these patients with a throat pack to reduce air leaks and risk of aspiration, and this may not be an effective option for lung transplantation. Kim et al described a significant leak around the tracheostomy cuff in an MKS patient, and they were forced to close the tracheostomy and orally intubate again with an endotracheal tube for a prolonged mechanical ventilation patient in the intensive care unit.

Subsequently, the technique employed for lung isolation for transplantation was arduous because of the increased airway diameters. The authors believed that the cuff on the 9F Fuji blocker was large enough compared to the double-lumen bronchial cuff to occlude the airway, but the spherical shape blocker cuff could not conform to the oblong shape of the bronchi. There is a need to be aware of consequences of the large-volume cuff and mucosal injuries and to require monitoring of cuff pressure at regular intervals. Even though the shape of the Arndt blocker is more spherical, the authors encountered difficulties in achieving acceptable lung isolation. Unfortunately, the authors had no prior experience in using other bronchial blockers such as the Cohen or EZ Blocker due to non-availability in their institution. The cuff from the Cohen blocker should be able to accommodate a large volume of air, and the authors were unsure about successful lung isolation in MKS patients. In addition to imperfect bronchial sealing with the blocker, they also had difficulty in visualizing the bronchial structures through bronchoscopy due to copious secretions.

Another strategy that was attempted was left mainstem intubation of the endotracheal tube in an effort to provide one-lung ventilation. While increased volumes of air were applied to the cuff, similar problems to the Fuji blocker were encountered, as the cuff on the ETT was not large enough or adequately shaped to provide an acceptable seal to facility OLV. Interestingly, though, the diameter of the right bronchus was larger than the left bronchus by 1.2 cm; the right-side lung isolation was acceptable enough to perform transplantation due to a lack of dilation at the origin of the right mainstem bronchus compared to the left one (Fig 4). Both airway manipulation and removal of secretions from the tracheobronchial tree have to be done under bronchoscopic guidance to prevent mucosal injuries from fragile tracheobronchial tissues. Due to possible disruption of the right mainstem bronchial anastomosis by advancing ETT to the right bronchus, the anesthetic and surgical team decided to perform left-lung transplantation using CPB with clamp on the left bronchus. Initial left-lung implantation followed by right lung would have the potential to allow the team to avoid this issue.

In summary, MKS is a rare disease with significant anesthetic and airway implications impacting the safety of the patients. In addition to difficulties with lung isolation, great care had to be taken to avoid damage to the tracheal walls due to extreme fragility and, therefore, high risk for tracheal rupture. The authors would also suggest implanting the left lung first followed by the right lung to facilitate lung isolation either with an endotracheal tube or bronchial blocker. Utilizing either extracorporeal membrane oxygenation or CPB potentially can allow avoidance of hemodynamic- and major airway- and ventilation-related complications.
REFERENCES