Living-donor lobar lung transplantation for lymphangioleiomyomatosis

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Abstract

Living-donor lobar lung transplantation seems to be best suited for children and small adults because only two lobes are transplanted. However, the amount of tolerable size discrepancy between donors and recipients is currently unknown. We report two cases of lymphangioleiomyomatosis with hyperinflation successfully treated with living-donor lobar lung transplantation in spite of large size disparity.

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1. Introduction

Lymphangioleiomyomatosis (LAM), a rare disease affecting women of childbearing age, is characterized by cystic destruction of the lung in the presence of hyperinflation on a chest X-ray. Single or bilateral lung transplantation is considered as valuable therapeutic modalities in patients with end-stage pulmonary LAM [1]. Living-donor lobar lung transplantation (LDLLT) seems to be best suited for children and small adults because limited amount of lung tissue is transplanted, and has been applied most exclusively to cystic fibrosis [2]. The use of significantly undersized grafts is potentially problematic because it could result in poor ventilation due to remaining large dead space [3]. We report two cases of LAM with hyperinflation successfully treated with LDLLT in spite of large size disparity.

2. Case reports

2.1. Patient 1

When a 23-year-old female had dyspnea and right pneumothorax, LAM was diagnosed by thoracoscopic lung biopsy in April 2000. Despite treatment with progesterone, she had developed multiple episodes of bilateral pneumothorax. Her forced vital capacity (FVC) and forced expiratory volume in 1 s were 710 and 220 ml, respectively. She was completely bed-ridden and required oxygen inhalation continuously (5 l/min) with arterial oxygen tension of 80.7 mmHg and arterial carbon dioxide tension of 76.2 mmHg. On October 18, 2000, she underwent LDLLT with a right lower lobe from her brother (27 years) and a left lower lobe from her mother (51 years). Preoperative chest X-rays of the recipient and the two donors showed that the recipient had the largest lung among the three (Fig. 1). The height and weight were 148 cm, 36 kg for the patient, 160 cm, 55 kg for the brother and 150 cm, 60 kg for the mother. The surgical and logistic aspects of the right and left donor lobectomy and bilateral lobar implantation have been previously described by Starnes’s group [2]. The patient was separated from cardiopulmonary bypass easily. The initial oxygen tension was 446.8 mmHg with the patient breathing 100% oxygen and the systolic pulmonary artery pressure was 47.0 mmHg after discontinuation of cardiopulmonary bypass. Her postoperative chest X-ray demonstrated well-expanded grafts without detectable dead space (Fig. 2). The patient was completely weaned from the respirator within 3 days. She became oxygen free at 1 month with arterial oxygen tension of 100.4 mmHg and arterial carbon dioxide tension of 36.8 mmHg. She has had no sign of rejection or infection. Thirty months postoperatively, she is in excellent physical

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condition with a FVC of 1710 ml (60.4% of predicted), forced expiratory volume in 1 s of 1560 ml (57.6% of predicted).

2.2. Patient 2

In January 2002, a 30-year-old female with LAM began having shortness of breath on exercise. Hyperinflation was remarkable on her chest X-ray. She developed intractable bilateral pneumothorax and became completely ventilator dependent on June 15. On July 1, 2002, she underwent LDLLT with a right lower lobe from her husband (33 years) and a left lower lobe from her mother (54 years) under cardiopulmonary bypass. The height and weight were 155 cm, 42 kg for the patient, 184 cm, 72 kg for the husband and 154 cm, 63 kg for the mother. Although she had the largest lung among the three on chest X-ray, postoperative chest X-ray showed no detectable dead space. The patient was completely weaned from the respirator within 13 days and became oxygen free at 3 weeks. Ten months postoperatively, she is in excellent physical condition with a FVC of 1820 ml (62.2% of predicted), forced expiratory volume in 1 s of 1210 ml (43.6% of predicted).

3. Discussion

LDLLT was developed by Starnes’ group [2] as an alternative to cadaveric lung transplantation. The policy of our program has been to limit LDLLT to critically ill patients who are unlikely to survive the long wait for cadaveric lungs. Because only two lobes are transplanted, this procedure seems to be best suited for children and small adults, and has been applied most exclusively to cystic fibrosis. Recently, the indications have successfully expanded to include recipients with primary pulmonary hypertension [4,5], pulmonary fibrosis [4], and bronchiolitis obliterans [5,6]. However, question remains whether LDLLT can be applied to the adult patient with hyperinflated lungs. Excessively small grafts may cause high pulmonary artery pressure, resulting in lung edema. A pleural space problem may increase the risk of empyema. Overexpansion of the donor lobes may contribute obstructive physiology by early closure of small airways [3].

We have previously proposed a formula to estimate the graft FVC based on the donor’s measured FVC and the number of pulmonary segments implanted [7]. When the total FVC of the two grafts was more than 50% of the predicted FVC of the recipient, we accepted the size disparity regardless of the recipient’s diagnosis. The total FVC of the two grafts was 54.2% of the recipient’s predicted FVC in case 1 and it was 58.1% in case 2.

As shown in figures, recipient’s adaptation ability to small grafts was remarkable. Both patients did not have any
space problems after LDLLT. The suction of the chest drainage tubes started at 10 cmH₂O and decreased to water seal within a few hours, thus avoiding overinflation of the small grafts. Although the amount of tolerable size discrepancy between donors and recipients is currently unknown, this report demonstrates that LDLLT can be applied to selected patients with hyperinflated lungs.

References


