Living-donor lobar lung transplantation for primary pulmonary hypertension in an adult

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In an effort to address the donor shortage issue, living-donor lobar lung transplantation has been performed with satisfactory intermediate survival and functional results.[1] Because a limited amount of lung tissue is transplanted, this type of operation seems to be best suited for children[2] and small adults and has been applied almost exclusively in patients with cystic fibrosis. To our knowledge, this is the first reported case of living-donor lobar lung transplantation for primary pulmonary hypertension (PPH) in an adult.

Clinical Summary

A 19-year-old female patient received a diagnosis of PPH in 1996, at the age of 14 years. Continuous intravenous infusion of epoprostenol (prostacyclin) was initiated. After an initial improvement to New York Heart Association class II, her condition began to deteriorate in 1998. Despite high-dose epoprostenol therapy (140-160 ng $\cdot$ kg^{-1} $\cdot$ min^{-1}), her status deteriorated to class III in 2000. On November 4, massive hemoptysis developed and she was urgently admitted with signs of right-sided heart failure. Right heart catheterization revealed highly elevated pulmonary arterial pressure (systolic/diastolic/mean: 80/40/58 mm Hg) and severely reduced cardiac index (1.4 L $\cdot$ min^{-1} $\cdot$ m^{-2}). High-dose intravenous inotropic therapy with diuretics was initiated. A second catheter examination on December 15 revealed minor improvement in cardiac index with persistent high pulmonary artery pressure (Table 1). Two-dimensional echocardiography demonstrated a dilated hypokinetic right ventricle in association with an enlarged right atrium and pulmonary artery, as well as massive tricuspid regurgitation. Left ventricular function was normal apart from flattening of the interventricular septum. The right ventricular ejection fraction was 14.7% by first-pass nuclear angiography. She was completely bedridden and required continuous oxygen inhalation (4 L/min).

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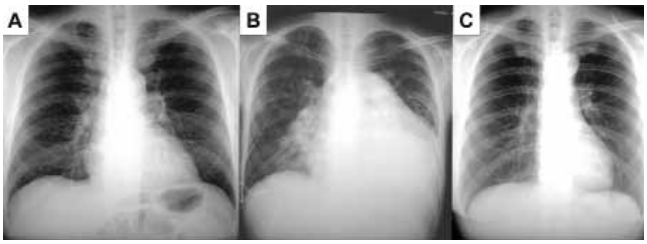

Figure 1. Preoperative chest x-ray films of the recipient and the two donors showed similarity in size. A, Right lower lobe donor; 51-year-old father. B, Recipient; 19-year-old daughter. C, Left lower lobe donor; 50-year-old mother.

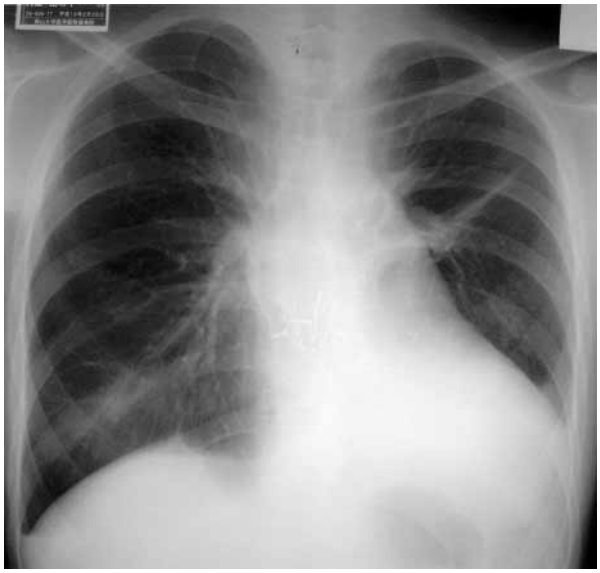

Figure 2. Chest x-ray film 50 days after living-donor lobar lung transplantation. Well-expanded grafts filled the chest cavity, leaving no detectable dead space without cardiomegaly.

On January 5, 2001, she underwent living-donor lobar lung transplantation with a right lower lobe from her father (51 years old) and a left lower lobe from her mother (50 years old). Preoperative chest x-ray films of the recipient and the two donors showed that the three were quite similar in size (Figure 1). The height and weight were 157 cm and 38.0 kg for the recipient, 172 cm and 71.0 kg for the father, and 157 cm and 55.0 kg for the mother.

TABLE I. Assessment of lung and cardiac function

	Before transplantation	After transplantation (2 mo)
PAP (mm Hg)	84/48 (58)	33/12 (19)
PCWP (mm Hg)	11	8
CVP (mm Hg)	8	0
CI ($L \cdot min^{-1} \cdot m^{-2}$)	1.8	3.2
RVEF (%)	14.7	76.6
Pao_2 (mm Hg)	82.2 (O_2 4 L/min)	96.0 (room air)
$Paco_2$ (mm Hg)	25.2	39.6

PAP, Pulmonary artery pressure; *PCWP,* pulmonary capillary wedge pressure; *CVP,* central venous pressure; *CI,* cardiac index; *RVEF,* right ventricular ejection fraction obtained by radionuclide scan; *PaO_2,* arterial oxygen tension; *$PaCO_2$,* arterial carbon dioxide tension.

Because of the recipient's poor preoperative condition, partial cardiopulmonary bypass was initiated via the femoral vessels under local anesthesia. Then, the recipient was anesthetized and intubated. The surgical and logistic aspects of the right and left donor lobectomy, the donor lobe back-table preservation technique, and the recipient bilateral pneumonectomy and bilateral lobar implantation have been previously described by Starnes's group.[1] In the immediate reperfusion period, she had pulmonary edema in the left graft and continued pulmonary hypertension. The right pulmonary arterial anastomosis was found to be severely kinked. After placement of three tacking stitches, gas exchange and pulmonary arterial pressure improved immediately.

During the early postoperative period, she was treated with various cardiovascular medications such as intravenous dopamine, dobutamine, alprostadil (prostaglandin E_1), phosphodiesterase III inhibitor, and inhaled nitric oxide. Postoperative immunosuppression was a triple drug therapy consisting of tacrolimus, mycophenolate mofetil, and prednisone. The patient was completely weaned from the respirator within 12 days. The postoperative right heart catheterization on day 50 revealed remarkable improvement in pulmonary hemodynamics (Table 1). Right ventricular ejection fraction and arterial blood gases were excellent. Echocardiographic changes included a decrease in right atrial and ventricular diameter, round left ventricular shape, and no tricuspid regurgitation. The chest x-ray film demonstrated well-expanded grafts without cardiomegaly (Figure 2). The recipient's forced vital capacity (FVC) became 1550 mL, or 49.7% of her predicted FVC. She was discharged from the hospital 60 days after transplantation.

Discussion

Significant advances in medical treatment of PPH have been achieved by continuous intravenous epoprostenol therapy in the past decade.[3] Although most patients who have improvement of their condition with epoprostenol maintain this response, sudden deterioration or life-threatening complications can occur. This patient had massive hemoptysis and was thought to have a short life expectancy without lung transplantation. The only realistic option for this patient was to receive living-donor lobar lung transplantation, because obtaining brain-dead donors is extremely difficult in Japan. There were obvious concerns regarding whether pulmonary hypertension would develop in the two lobes receiving the patient's entire cardiac output. Starnes and associates[4] have reported successful living-donor lobar lung transplantation in 4 patients with PPH; however, all of them were younger than 15 years. This patient was an adult: therefore, preoperative chest x-ray films indicated that the recipient and the two donors were quite similar in size. The father's FVC was 4360 mL and the mother's 2970 mL. Given that the right lower lobe consists of 5 segments and the left lower lobe of 4, total FVC of the 2 grafts was estimated to be 1773 mL ([4360 mL × 5]/19 + [2970 mL × 4]/19), or 56.8% of the recipient's predicted FVC (3118 mL). We thought that 56.8% was within an acceptable range since single lung transplantation had been performed successfully for patients with PPH.[5] Pulmonary arterial pressure became nearly normal within 2 months, validating the functional capacity of the two adult lobes to handle the cardiac output of the adult recipient with PPH.

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