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Case Reports

Left Pulmonary Artery Patch Augmentation for Lung Transplant in a Patient With Situs Inversus

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Kartagener syndrome is characterized by situs inversus and defective cilia motion, the latter of which can lead to chronic infections and respiratory failure. If lung transplant is indicated, dextrocardia can pose surgical challenges. We report a rare case of sequential bilateral lung transplant in a 58-year-old man with Kartagener syndrome whose left pulmonary artery was abnormal in length, location, and direction. After placing the donor lungs in their orthotopic position, we augmented the recipient left pulmonary artery with a bovine pericardial patch. After 17 months of follow-up, the patient was in good condition with excellent graft function. ***(Tex Heart Inst J 2021;48(1):e197112)***

Kartagener syndrome is a rare disorder characterized by situs inversus—the reverse orientation of internal organs—and defective cilia formation during embryogenesis, which can lead to lung infections and decreased function.[1] Lung transplant in patients with Kartagener syndrome requires careful consideration of anatomic differences; asymmetric hilar anatomy can complicate bronchial and vascular anastomoses. We report a rare and challenging case of sequential bilateral lung transplant in a man with Kartagener syndrome.

Case Report

A 58-year-old man with Kartagener syndrome was evaluated at our institution for bilateral lung transplant. His medical history included chronic, hypoxemic respiratory failure with end-stage lung disease secondary to bronchiectasis. Computed tomograms showed situs inversus totalis and reversed hilar structures (Fig. 1). The patient's pulmonary artery (PA) pressure was 82/49 mmHg (mean, 62 mmHg). Pulmonary function tests (PFTs) revealed a forced vital capacity (FVC) of 1.64 L (31% of predicted value), a forced expiratory volume in 1 second (FEV1) of 0.74 L (17% of predicted value), and an FEV1/FVC ratio of 45%. The patient was taking inhaled steroids, a β2-agonist, and anticholinergics. He was deemed a candidate for lung transplant and was admitted when donor lungs became available.

A bilateral anterior thoracotomy (clamshell incision) was performed across the sternum at the 4th intercostal space to expose the patient's pleural cavities. The right lung had 2 lobes, and the left lung had 3 lobes (Fig. 2). The patient did not tolerate single-lung ventilation during dissection of the right hilum; therefore, we administered heparin, cannulated the right-sided ascending aorta and right atrium, and initiated cardiopulmonary bypass.

The right superior and inferior pulmonary veins, the right PA, and the right bronchus were isolated and divided sequentially. The right superior and inferior pulmonary veins were dissected and closed with vascular staples. A vascular clamp was placed proximally over the PA. Although the angulation between the hilar structures was abnormal (Fig. 1), the right bronchial, PA, and atrial anastomoses aligned with the right donor lung; therefore, the donor lung needed no substantial structural modifications. The PA was sewn with a running 5-0 Prolene suture; the vascular clamp was then released, and the PA was flushed out through the left atrium. The left atrium

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was de-aired by removing the left atrial clamp and then sewn with a running 4-0 Prolene suture imbricating the posterior wall. The right bronchus was anastomosed approximately 2 to 3 cm from the carina and proximal to the takeoff of the upper lobe with a running 4-0 Prolene suture. The membranous and cartilaginous portions of the main bronchus were carefully aligned, and the connection was buttressed with lymphatic and pericardial tissue over the top. The right lower lobe completely expanded with ventilation.

Using the same technique, we dissected and resected the recipient left lung. The left bronchial anastomosis was performed proximal to the takeoff of the upper lobe branch. During anastomosis of the left PA, we noted that the abnormal location and direction of the native

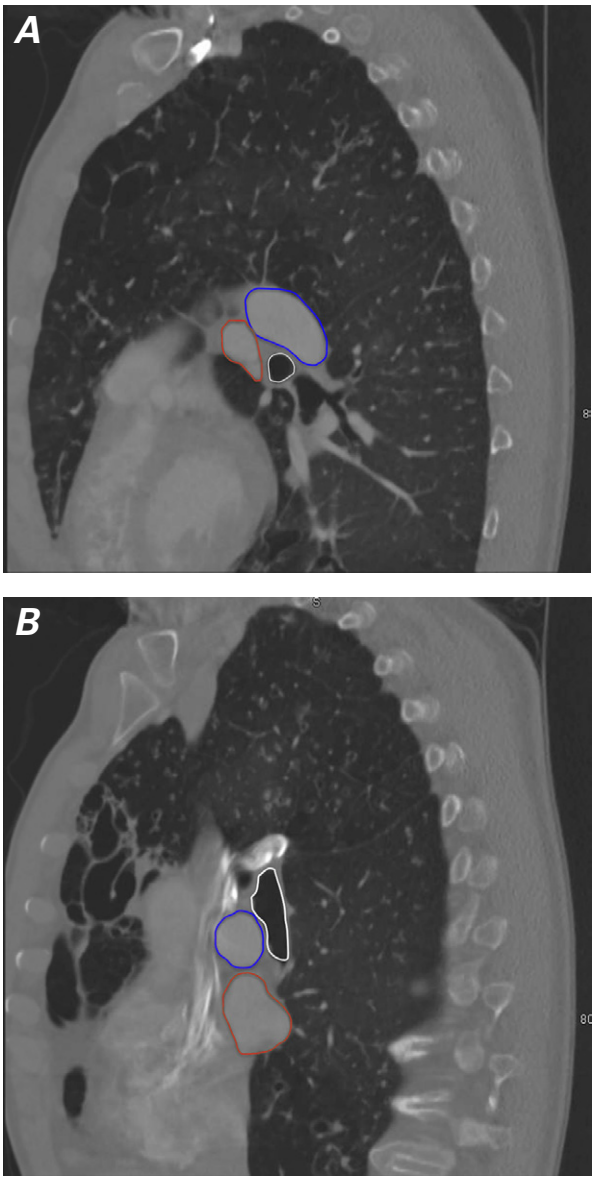

Fig. 1 *Computed tomograms of the patient's* ***A****) right and* ***B****) left hilar structures (sagittal views) show the reverse relationship of the pulmonary artery (blue outline) to its corresponding bronchus (white outline) and pulmonary vein (red outline).*

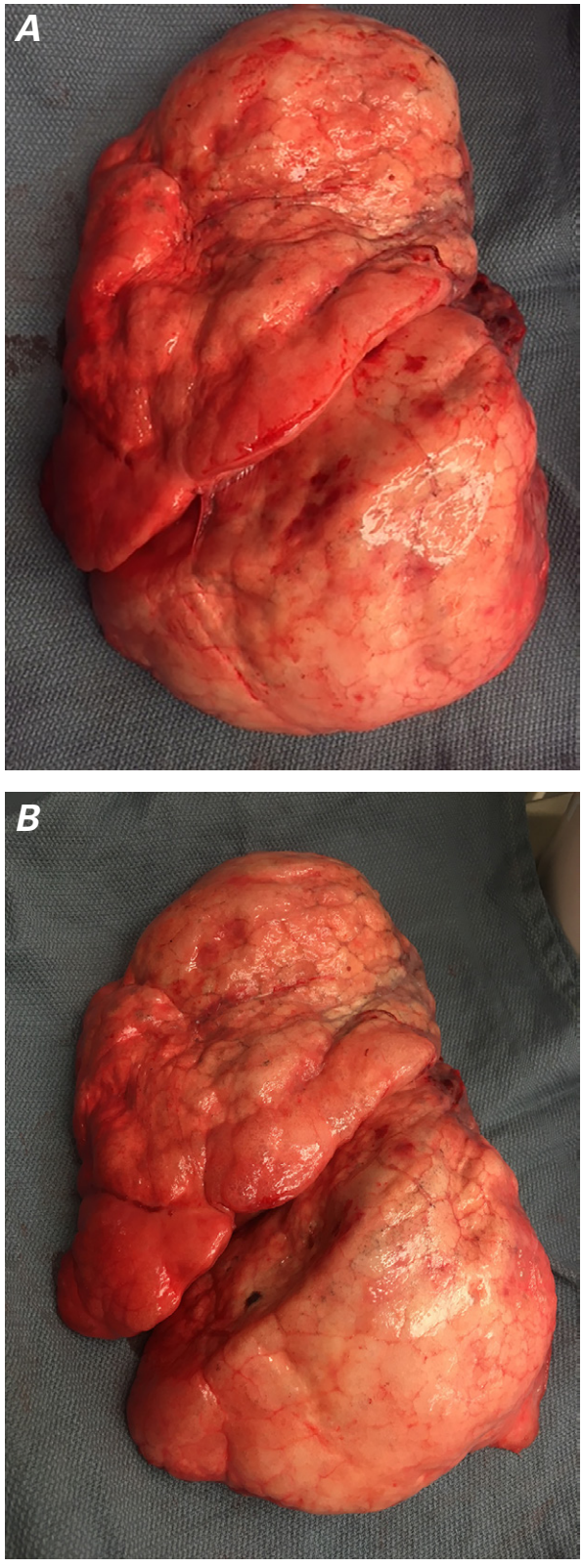

Fig. 2 *Photographs show the patient's* ***A****) bi-lobed right lung and* ***B****) tri-lobed left lung after resection.*

PA caused excessive tension, so we sewed the posterior wall in normal fashion and augmented the anterior left PA with a bovine pericardial patch, sewn on with a 5-0 Prolene suture (Fig. 3). The left atrial anastomosis aligned well and bridged the gap between the recipient and donor arteries without mechanical obstruction or tension.

The patient's postoperative clinical course was uneventful. His lung function was excellent, except for a mild air leak that necessitated tube thoracostomy for 7 days after surgery. He had no bronchial or vascular anastomotic complications. On postoperative day 30, PFTs showed an FVC of 3.78 L (69% of predicted value), FEV1 of 3.62 L (88% of predicted value), and FEV1/FVC ratio of 96%.

The patient's clinical course after discharge from the hospital was favorable and uneventful. At his 17-month follow-up visit, PFTs showed an FVC of 3.87 L (70% of predicted value), FEV1 of 3.44 L (83% of predicted value), and FEV1/FVC ratio of 89%. Chest radiographs were unremarkable (Fig. 4).

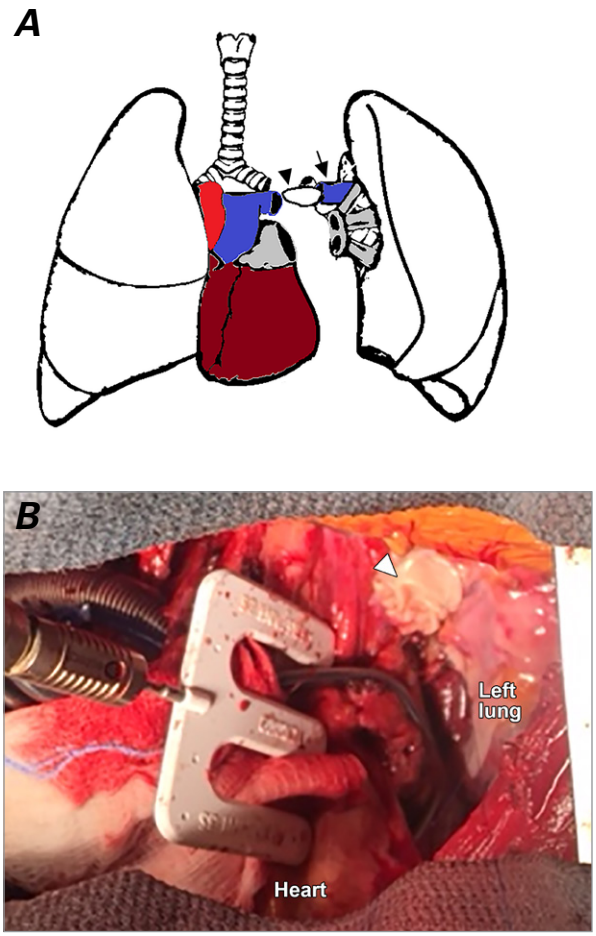

Fig. 3 ***A****) Illustration shows augmentation of the anterior left pulmonary artery (arrow) with a bovine pericardial patch (arrowhead).* ***B****) Intraoperative photograph shows the operative result in our patient, with patch in place (arrowhead).*

Discussion

Bilateral lung transplant in patients with situs inversus totalis is rarely reported in the literature and is limited to case reports and small case series.[2-4] In this case, the abnormal length and direction of the recipient left-sided PA was unusual and necessitated modifications to our standard implant technique. In normal situs, the left main PA is superior and slightly posterior to the bronchus, and the right main PA is anterior and inferior to the bronchus. The mirror-image dextrocardia changes the direction and length of the hilar structures, which may increase the risk of mechanically obstructing the pulmonary vasculature after lung transplant.[5] Furthermore,

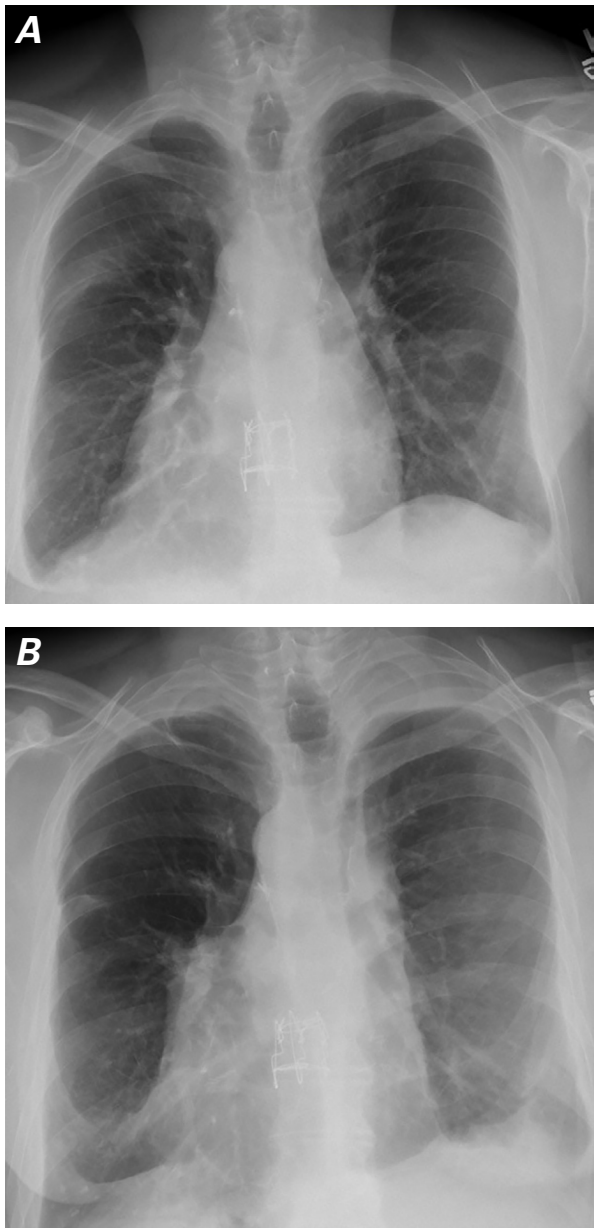

Fig. 4 *Chest radiographs show normal-appearing lungs* ***A****) 6 months and* ***B****) 17 months after transplant.*

the heart's malposition may lead to chronic compression and atelectasis of the implanted donor lung's right lower lobe, which could necessitate the lobe's removal.

In this case, we aligned the recipient right-sided hilar structures with no difficulty and no concern for anastomotic complications, and we found that the dextrocardia did not result in compression of the right lower lobe. The right donor PA and vein were also long enough to accommodate the length discrepancies—critical to ensure before cutting and anastomosing the PA. In fact, we recommend performing the venous anastomosis first and then modifying the PA anastomosis procedure as needed in such cases. The left-sided PA anastomosis in this case was under tension while we were estimating the position of the venous anastomosis, and we were concerned about flattening of the PA and potential stenosis. The recipient PA was angled inferiorly, whereas the donor PA was angled superiorly. In addition, the angulation of the left bronchial anastomosis further pushed the donor PA away from the recipient vessel. Augmenting the anterior left PA anastomosis with a bovine pericardial patch resulted in adequate vessel length and unobstructed flow.

In conclusion, sequential bilateral lung transplant in patients with situs inversus is reasonable and feasible, but only after anatomic relationships between the donor lungs and recipient hilar structures are first assessed to prevent potential anastomotic complications. Depending on the anatomy seen after dissection and pneumonectomy, the surgeon should be prepared for multiple reconstruction options.

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