CASE REPORTS

SEQUENTIAL DOUBLE-LUNG TRANSPLANTATION IN KARTAGENER SYNDROME: A CASE REPORT

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Abstract

Kartagener syndrome (KS) is a rare congenital disorder, characterized by sinusitis, bronchiectasis and situs inversus. Lung transplantation is an effective treatment for end-stage lung failure, but dextrocardia and differences between hilar structures and pulmonary lobes require adjustments to conventional surgical technique.

We present a case of a double-lung transplant without extracorporeal oxygenation in a 48-year-old male patient with KS. Through a Clamshell incision, right-left rotation was identified. To provide an end-to-end arterial and bronchial anastomosis, longer donor PA's and right main bronchus were preserved. Wedge resection of right inferior lobe was unnecessary and there was no left residual cardiac chamber.

Patient was discharged 32 days after surgery without complications.

Despite being anatomically challenging, lung transplant was done successfully without the need for plastic maneuvers or extracorporeal circulation. This reinforces the idea that it should be an option in end-stage lung disease.

Keywords: *Kartagener Syndrome; Primary ciliary dyskinesia; situs inversus; double-lung transplant*

INTRODUCTION

Kartagener's syndrome is a rare genetic disorder, with an incidence of 1 in 20,000-30,000 live births. [1]

KS is a subset of a larger group of ciliary motility disorders called primary ciliary dyskinesias (PCD) [2]. An ineffective beat of cilia results in impaired mucociliary clearance with chronic inflammation and subsequent development of bronchiectasis. [2,3]

Approximately 50% of PCD patients manifest the clinical triad of KS: situs inversus, chronic sinusitis and bronchiectasis. [3,4]. As in other PCD's, patients with KS can express other clinical signs and symptoms related to cilia dysmotility, such as ear infections and infertility [5].

There are no targeted therapies. Treatment involves the use of chronic suppressive antibiotics, inhaled hyperosmolar agents, macrolides as anti-inflammatory agents and bronchodilators. Chest physiotherapy and cardiovascular exercise are recommended to promote airway clearance [2,6].

Eventually, recurrent infections will lead to end-stage lung disease, being lung transplantation an effective treatment for pulmonary disease [4].

According to the United Network for Organ Sharing, 12 double lung transplants were performed for KS from 1987-2015. 1-year survival was comparable with others causes [7].

Since 2015, few cases have been reported, emphasizing the lack of information in this field. [4,8,9]

Asymmetric hilar anatomy may increase bronchial and vascular anastomoses complexity, with some centers having made important alterations to the usual technique [10–12]. Alternatively, others made simple adjustments, mainly regarding the length of pulmonary arteries (PA) [4,8,13].

We report a case of double-lung transplantation without the use of cardiopulmonary bypass in a patient with KS.

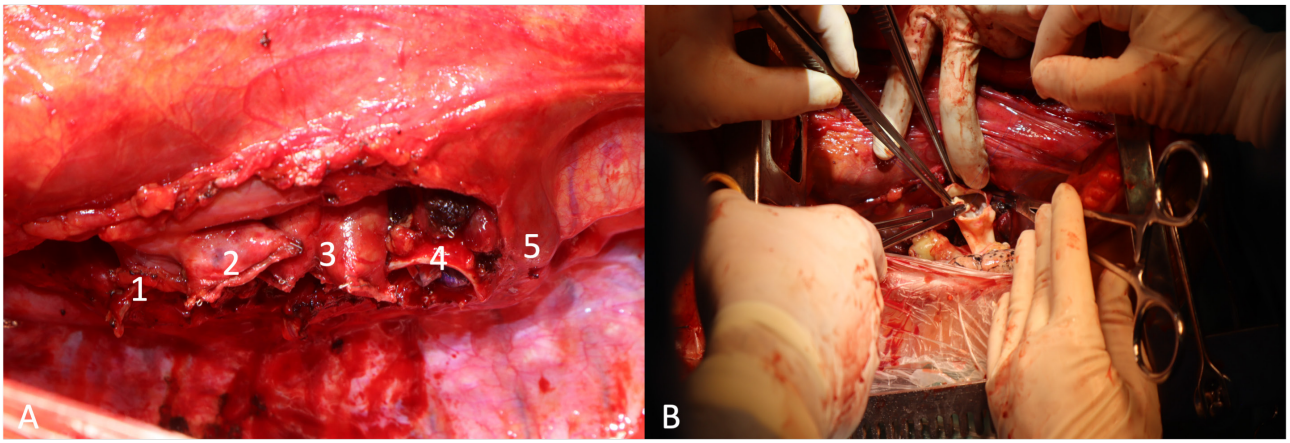

Figure 1 *Left pulmonary hilum (A) and implantation of left lung preserving a longer pulmonary artery (B). 1. Inferior pulmonary vein; 2. Superior pulmonary vein; 3. Pulmonary artery; 4. Bronchus; 5. Azygos arch.*

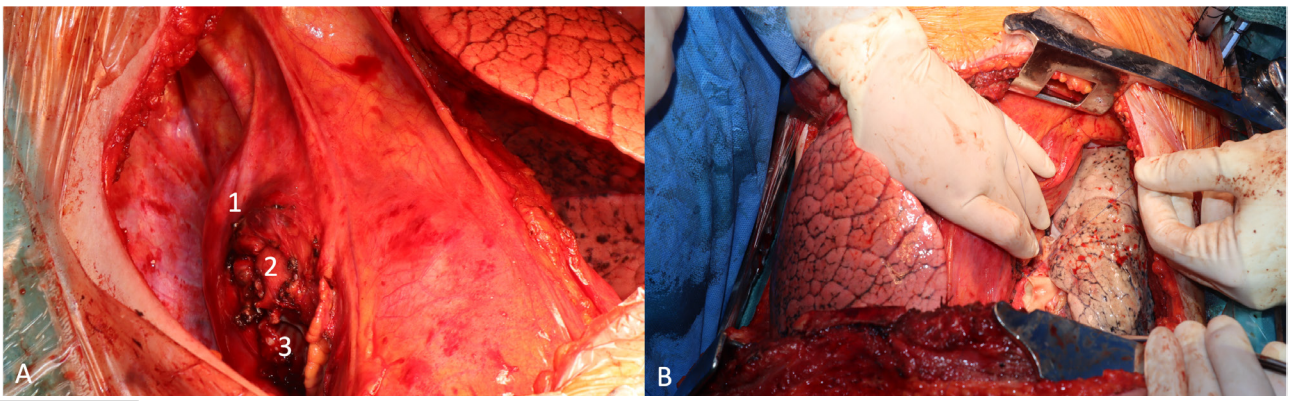

Figure 2 *Right pulmonary hilum (A) and implantation of right lung preserving a longer pulmonary artery (B). 1. Aortic arch; 2. Pulmonary artery; 3. Bronchus.*

CASE REPORT

We report a 48-year-old male with history of KS (Table 1.), with history of multiple respiratory infections and progressive declining functional status. When first evaluated for lung transplant, he had severe respiratory failure (Table 2.) and needed supplemental O2.

He remained clinically stable for 2 years, when a matching donor was found. The donor was 1,75cm (5,74ft), had a TLC of 6,9L and his PaO2 was 430mmHg using test conditions.

Donor lung procurement was performed in a regular technique.

In the recipient, an endobronchial blocker was used for ventilation. Through a clamshell incision in the fourth intercostal space, left-right structure inversion was identified.

Pulmonary ventilation/perfusion (V/Q) scan showed lung V/Q of 44% in left lung, so left pneumectomy was performed first. Left pulmonary hilus had a right configuration, with the bronchus being in a posterior position in relation to the PA, in contrast to donor bronchus, which had an inferior position (Figure 1.) Regular end-to-end anastomosis of the bronchus and PA were performed (Figure 1.). No technical alterations were made in the auricular cuff. Left lung ischemic time was 4h12min.

Right pneumonectomy was performed without the need for extracorporeal oxygenation. Recipient right pulmonary hilum had a left hilum configuration: PA in a superior position in relation to the bronchus, in contrast to donor PA, which had an anterior position (Figure 2.). End-to-end bronchus and PA anastomosis were performed (Figure 2.). No technical alterations were made in the auricular cuff. Right lung ischemic time was 6h22min.

Patient was extubated 14h after surgery.

No complications were observed during early post-operative period, without signs of acute rejection. Patient was discharged 32 days after surgery, clinically stable and without supplemental oxygen.

Spirometry parameters one month after discharge are shown in table 1.

Two years after transplant, patient remains stable without supplemental oxygen, complying with the appropriate

Table 1 **Recipient characteristics**

	Recipient
Age	49-years-old
Hight	1,69m
Weigh	69kg
BMI	24,2kg/m^2
Blood Type	A Rh+

Table 2 **Patient pulmonary function tests results before and after lung transplant.**

	T0	T1m	T2y
FEV1	1,1L (32%)	2,56L (73%)	3.26L (95%)
DLCO	3,7L (38%)	6,10L (72%)	*
TLC	7,32L	6L	*

*Not measured.
T0 – Before lung transplant;
T1m – 1 month after transplant;
T2y – 2 years after transplant

therapeutic regimen and prophylaxis. Bronchoscopy and spirometry are regularly done. Latest bronchoscopy showed no signs of rejection, and his spirometry shows improvements in FEV1 to 3.26L (95% of predicted).

DISCUSSION

Lung transplantation in situs inversus can be a surgical and anesthetic challenge.

Most authors suggest preservation of recipient's PA length as a crucial step for the procedure. That way, and end-to-end anastomosis seems to be facilitated, avoiding arterial kinking, and overcoming the differences between arterial e bronchus positioning. There appears to be no concern regarding airway and auricular cuff anastomosis, both being done in a regular way [4,8,9].

In this case, anatomical differences between left and right pulmonary hilum, and the relationship between PA and bronchus in both sides were considered. Preserving longer PAs allowed us to overcome those difficulties, making end-to-end anastomosis possible.

Another significant concept pertains to right lower lobe (RLL) position, creating an anatomical conflict with the heart, which could result in restricted lung expansion or heart compression. Some authors propose RLL lobectomy or large wedge resection to accommodate the position of the heart in the right hemithorax [11]. In our case, after lungs implantation, dextrocardia did not cause RLL compression, obviating the necessity for lung resection. Furthermore, no left-sided cardiac empty pleural space was noted.

Additionally, due to variations main bronchi configuration and length, careful consideration was given to double lung ventilation. Initial attempts using a left-sided double lumen tube posed challenges for selective ventilation, requiring the use of an endobronchial blocker. Literature suggests that employing a right-sided double lumen tube might facilitate lung exclusion from the start. [4,8].

The use of ECMO remains contentious. Some centers endorse its immediate use, particularly important during right-sided anastomosis, when there are hemodynamic challenges during heart retraction for adequate exposure. [8] In our case, ECMO support was unnecessary, and the patient was hemodynamically stable throughout the procedure.

Post-operatively, patient stayed in the hospital for 32 days, consistent to the average length of stay for patients submitted to double-lung transplant in our center. In this period, motor and respiratory rehabilitation are crucial for a full recovery. Therefore, patient underwent regular respiratory and motor physiotherapy.

CONCLUSION

We report a successfully performed double lung transplant in a patient with Kartagener's syndrome without the need of extracorporeal oxygenation. ECMO indications should remain similar to conventional lung transplantation protocols.

Despite major anatomical asymmetry between donor and recipient lungs, minimal changes were made to the standard surgical technique.

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