Simultaneous pectus excavatum correction and lung transplantation–A case series

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Severe chest wall deformities are considered an absolute contraindication for lung transplantation. The significantly impaired chest compliance associated with pectus excavatum is thought to result in a high risk of postoperative respiratory complications and significant morbidity and mortality. We herein report our pooled institutional experience consisting of 3 patients who underwent bilateral lung transplantation and simultaneous correction of a pectus excavatum. Two of the patients were children and 1 patient had severe asymmetric pectus. All patients received a size-reduced double lung transplant and the deformity was corrected by a Nuss or modified Ravitch procedure. The perioperative course was complicated by prolonged weaning requiring tracheostomy in 2 of the 3 patients. However, long-term results were good and all 3 patients are alive in excellent clinical condition 72, 60, and 12 months after the transplantation. This case series demonstrates that patients with severe chest wall deformities should not a priori be excluded from lung transplantation, and a combined approach is feasible for selected patients.

KEYWORDS
clinical research/practice, lung transplantation/pulmonology, patient referral, surgical technique

1 | BACKGROUND

According to an International Society for Heart and Lung Transplantation (ISHLT) consensus, severe chest wall deformities are considered a contraindication for lung transplantation. One of the main concerns is the limited chest wall compliance leading to impaired postoperative mucus clearance as well as reduced long-term pulmonary function. Moreover, asymmetry of the chest and scoliosis of the spine make size-matching difficult. Although pectus excavatum (PE) is a relative rare condition, it is likely to be encountered by large lung transplant centers. We chronologically report 3 cases of patients receiving lung transplantation with simultaneous pectus correction between June 2012 and December 2018 at our transplant center.
2 | CASE PRESENTATIONS

2.1 | Case 1

A 38-year-old woman with idiopathic pulmonary fibrosis and symmetric pectus excavatum (Haller Index 5.8) was referred to our transplant center for evaluation of lung transplantation (Figure 1A). Oxygen requirement at rest was 4 L/min. Preoperative lung function test showed a reduced diffusing capacity of the lung for carbon monoxide of 52%, predicted total lung capacity (TLC) of 4.9 L, and real TLC of 2.1 L. The medical history of the patient was unremarkable except for 2 open wedge lung resections performed several years before due to recurrent pneumothoraces. The patient was listed for bilateral lung transplantation with the plan to correct the chest wall deformity during transplantation.

After a waiting time of 137 days, a 46-year-old female 165-cm-tall donor became available with a calculated TLC of 5.1 L. A bilateral sequential lung transplantation on central venoarterial extracorporeal membrane oxygenation (ECMO) support with size reduction by resection of the middle lobe and lingula was performed. The patient’s chest was opened through a clamshell incision using the fourth intercostal space. The transversal sternotomy was done at the lowest level of the sternal defect. Mobilization of the lungs proved difficult due to dense pleural adhesions. Ischemic times were 370 (left lung) and 464 minutes (right lung). At the end of the procedure, a 305-mm (12 inch) Nuss bar was inserted slightly below the transverse sternotomy and fixed with a left-sided stabilizer. The transected sternum was closed with 2 sternal wires. Based on our institutional guidelines, venoarterial ECMO was prolonged postoperatively as a prophylactic measure due to increasing pulmonary arterial pressure with preserved gas exchange at the end of the procedure.\textsuperscript{5,6} The peripheral ECMO was removed on the second posttransplant day.

The postoperative course in the intensive care unit was characterized by prolonged weaning with need for a tracheostomy, and a transient organic psychosyndrome. Six days after removal of peripheral ECMO support, sudden cardiac arrhythmia with severe hemodynamic impairment required cardiopulmonary resuscitation. Based on this event, a left-sided hemothorax developed, which required evacuation a few days after. Despite the chest compressions, no bar dislocation was observed. The patient fully recovered and was transferred to the normal ward on postoperative day (POD) 21 and was discharged from the hospital on POD 30.

The last follow-up of this patient was 6 years after transplantation. Chest radiograph and computed tomography (CT) scan show an adequate position of the Nuss bar (Figure 1B). Pulmonary function tests revealed a mild but stable restrictive pattern with a vital capacity, TLC, and FEV1 of 2.05 (54%), 4.19 (79%), and 1.99 (63%) at the last follow-up visit.

2.2 | Case 2

At the age of 14 years, a boy diagnosed with alveolar proteinosis and symmetric pectus excavatum (Haller Index 5.5) was referred to our pretransplant clinic. The patient was diagnosed with alveolar proteinosis at the age of 4 years. By the age of 12 years, the boy had developed a pattern of nonspecific interstitial pneumonia as diagnosed by lung biopsy. Due to a continuous therapy-resistant worsening of his respiratory function with significant secondary pulmonary hypertension, he was listed for lung transplantation (Figure 2A). The patient had a predicted TLC of 4.9 L. Body plethysmography for establishing the real TLC could not be performed due to poor compliance.

After a waiting time of 149 days, lungs of an 8-year-old male donor were allocated to this patient. The arterial partial pressure of oxygen (\(P_{aO_2}\)) at a fraction of inspired oxygen (\(F_{I_O_2}\)) of 1.0 was
387 mm Hg at the time of offer, and the calculated donor TLC was 5.7 L. A bilobar lung transplantation with central venoarterial ECMO support was performed using the lower lobe of the donor for the right side, and the upper lobe for the left side. The surgical access was gained through a clamshell incision at the level of the fifth intercostal space. After implantation, limited chondrotomies were performed underneath the divided sternal bone and a short 254 mm (10 inch) Nuss bar was placed for correction at the sixth intercostal space. Xiphoid de-attachment was not needed. The Nuss bar was fixed with a sternal cable system (Synthes CMF, Chester, PA, USA) without lateral stabilizers. The same cable system was used to close the sternotomy. Figure 2B shows posttransplant chest radiograph and CT scans of the patient with an adequate position of the Nuss bar.

The patient developed a *Klebsiella* spp. pneumonia; however, he quickly recovered under antibiotic therapy. He was extubated on POD 5 and was transferred to the normal ward on POD 9. The further course was uneventful, and the patient was discharged from the hospital on POD 20.

Only 4 weeks later, the patient was readmitted with a newly developed suspicious left hilar mass on CT scan and a biopsy revealed Epstein-Barr virus–associated posttransplant lymphoproliferative disease. He received rituximab and cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy with an excellent response. Five years posttransplant, the patient is still in remission and in good clinical condition without any signs of recurrent alveolar proteinosis.

2.3 | Case 3

A 9-year-old girl with alveolar proteinosis and asymmetric pectus excavatum presented at our pretransplant outpatient clinic. During the
previous months, her respiratory function had gradually worsened and she required 4 L/min oxygen at rest. The patient had been receiving additional alimentation with a percutaneous endoscopic gastrostomy tube since the age of 4 years. Preoperative CT confirmed a severe asymmetric pectus excavatum with a Haller Index of 9.5 (Figure 3A). The predicted TLC of the patient was calculated as 1.95 L. Lung function tests could not be performed due to a lack of compliance.

After a waiting list time of 315 days, an organ from an 8-year-old female (120 cm, 36 kg) brain-dead donor was allocated to the patient. The donor PaO₂ at a FiO₂ of 1.0 was 521 mm Hg, and the calculated donor TLC was 2.13 L. A sequential double lung transplantation with central venoarterial ECMO support was performed. A clamshell incision was performed using the fourth intercostal space and the retrosternal plane was fully mobilized so that the heart could medialize after implantation. After a size-reduced double lung transplantation (extra-anatomical removal of middle lobe and lingula), a modified Ravitch procedure was performed with asymmetrical chondrotomies around the sternal bone from the third rib downwards to the xiphoid appendix. In addition, two 229-mm (9 inch) Nuss bars were used to lift the sternum. The first bar was placed in an oblique position between the fourth and the fifth intercostal space (above the sternotomy), and the second bar was placed in the sixth intercostal space underneath the sternotomy. Both bars were fixed using a cable system. Ischemic times were 280 minutes (right lung) and 350 minutes (left lung).

Postoperatively, respiratory weaning was prolonged and a tracheotomy was necessary. The patient developed a 4 MRGN Acinetobacter baumannii pneumonia, which required treatment with aztreonam, ceftazidime/avibactam, and fosfomycin. The patient was finally able to be transferred to the pediatric ward on POD 24 and was discharged from the hospital in excellent condition on POD 51.

At a follow-up of 12 months, the patient has returned to school and has started to participate in school sports. A CT scan 1 year after the operation shows a good correction of the chest wall deformity (Figure 3B), and the alveolar proteinosis has not recurred.

3 | DISCUSSION

Based on an ISHLT consensus, severe chest wall deformities are considered an absolute contraindication for lung transplantation due to complications associated with limited chest wall compliance. Accordingly, only 2 case reports exist in the literature of lung transplantation with concomitant correction of a pectus excavatum. In a report published in 2018 by Aigner and colleagues, a symmetric pectus excavatum in a 38-year-old fibrosis patient was corrected after a bilateral lung transplantation with a Nuss bar, which was inserted to lift the sternum through 2 anterior thoracotomies.7 The second report was published by a Chinese group in 2019, using a similar technique in a symmetric pectus patient requiring lung transplantation for graft-versus-host disease.7 The reported series herein extends these 2 cases by reporting 3 cases including (1) 2 pediatric transplantations, (2) 1 bilobar transplantation, and (3) 2 modified Ravitch procedures instead of a simple Nuss procedure (Table 1).

All 3 patients were transplanted through a clamshell incision. This type of incision has several advantages for pectus deformities.

| Case 1 | Case 2 | Case 3 |
|-------|-------|-------|
| **Age at Tx** | 38 | 14 | 9 |
| **Height (cm)** | 165 | 153 | 123 |
| **Weight (kg)** | 51 | 36 | 19 |
| **pTLC (L)** | 4.9 | 4.9 | 1.95 |
| **rTLC (L)** | 2.1 | n.d. | n.d. |
| **Donor TLC (L)** | 5.1 | 5.7 | 2.13 |
| **Diagnosis** | Fibrosis Symmetric PE | Alveolar proteinosis Fibrosis Symmetric PE | Alveolar proteinosis Surfactant deficiency Asymmetric PE |
| **Haller index before correction** | 5.8 | 5.5 | 9.5 |
| **Type of surgery** | DLuTx Size reduction | Lobar Tx | DLuTx Size reduction |
| **ICU days** | 21 | 9 | 24 |
| **Hospitalization** | 30 | 20 | 51 |
| **Tracheostomy** | Yes | No | Yes |
| **Haller index after correction** | 2.2 | 2.7 | 3.5 |
| **Follow-up time** | 72 mo | 60 mo | 12 mo |

Abbreviations: DLuTx, Double Lung Transplantation; ICU, intensive care unit; n.d., no data; PE, pectus excavatum; pTLC, Predicted Total Lung Capacity; rTLC, Real Total Lung Capacity; Tx, transplant.
First, it provides an optimal exposure of the surgical field. Especially in patients with a small left chest cavity with a mediastinal shift towards the left, dissection of the left hilum is hardly possible by other approaches. Second, a clamshell incision facilitates repositioning of the mediastinum in cases of severe asymmetric pectus excavatum. Third, in older patients with an asymmetric pectus with a lack of "self-correction" during Valsalva maneuver, such an approach allows tension release but chondrotomies are required for a good cosmetic result.

In our opinion, accepting patients with severe chest wall deformities for transplantation is possible; however, it comes at the cost of a higher risk of perioperative pulmonary complications. The limited chest wall compliance leads to impaired mucus clearance after extubation and patients are prone to develop pneumonia. This has several implications for the perioperative treatment of such patients. Sufficient pain control is essential in order to facilitate coughing. In addition, patients should receive intensive physiotherapy with the aim of improving clearance of bronchial secretions, recruiting atelectasis, and reaching early mobilization. In case of mucus retention, it is important to opt for an early tracheostomy in such patients. This facilitates cleaning of the bronchial system but also allows prolonged positive end-expiratory pressure ventilation in an awake and mobile patient and thus reduces the risk for chronic atelectasis. Another important aspect—especially in pediatric patients—is the sizing of the Nuss bar. Using shorter Nuss bars reduces the impact on respiratory mechanics and prevents long-term complications in the form of bar displacement or chest wall deformities in patients with an ongoing growth period. Chest wall deformities were fully corrected in all 3 patients with normal Haller indices in the 1-year follow-up CT scan and no relevant dynamics thereafter.

Although all 3 herein presented patients had a successful outcome, not every lung transplant candidate with severe chest wall deformity can be accepted and patient selection is crucial. Respiratory muscular status has to be preserved and frail patients are to be avoided, which means that the herein described combined procedure is only an option for young patients. Since the perioperative course is challenging, additional complicating factors should be avoided. This includes previous major lung surgery, circumstances requiring cardiopulmonary bypass instead of ECMO, additional spinal deformities and colonization with multiresistant bacteria such as Mycobacterium abscessus or Burkholderia cenocepacia. Minor chest wall deformities do not require a concomitant correction with a Nuss bar or a Ravitch procedures. They can be addressed by repositioning of the mediastinum or by unilateral size-reductions of the donor lung.

Titan endoprostheses are at a higher risk of infections in transplanted patients.\(^9\) We therefore decided to avoid immunosuppressive induction therapy in these patients. In addition, antibiotic coverage with piperacillin/tazobactam was maintained until C-reactive protein levels fell below our laboratory threshold of 0.5 mg/dL. With a follow-up of 72, 60, and 12 months, we have not removed the bars in our 3 patients. None of them has developed any bar-related complications and due to anticipated adhesions, we believe that a removal of the bars is an unacceptable risk.

We conclude that patients with severe chest wall deformities can be accepted for lung transplantation with an acceptable risk profile. Future revisions of guidelines for selecting lung transplant candidates should take this into account.

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**DISCLOSURE**

The authors of this manuscript have no conflicts of interest to disclose as described by the American Journal of Transplantation.

**DATA AVAILABILITY STATEMENT**

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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