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Lobar lung transplantation, followed by partial sternal resection and bronchial stenosis, in a patient with scoliosis

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Abstract
Scoliotic deformity represents a serious spinal disorder that influences the locomotive and cardiopulmonary systems. Some patients with severe scoliosis and end-stage lung disease are therefore denied lung transplantation. In patients with scoliosis considering lung transplantation, size match, straight back syndrome, delayed chest closure and bronchial stenosis are key issues clinicians should evaluate. Therefore, it is vital to determine donor-recipient size matches very precisely. Chest opening is a routine intraoperative primary therapeutic procedure after lung transplantation in unstable patients with oversized transplanted lungs. Postoperative bronchial stenosis occurs predominantly on the right side and is usually handled through interventional bronchoscopy and the insertion of stents. This report describes the complex case of a patient with scoliosis who underwent lobar transplantation in our center.

Keywords Scoliosis, Lung transplantation, Straight back syndrome, Delayed chest closure, Bronchial stenosis

Introduction

Scoliotic deformity represents a serious spinal disorder that influences the locomotive system as well as the cardiopulmonary system. An X-ray long film is an elementary part of the examination of patients with scoliosis, and curve measurement is based on the Cobb angle. Scoliotic curves up to 40 degrees are indications for conservative treatment (rehabilitation, orthosis). Scoliotic curves greater than 40 degrees are indications for surgical treatment [1].

There is no data on the number of patients with end-stage lung disease and scoliosis with indications or contraindications for lung transplantation (LTx). On the other hand, there is written evidence worldwide about a few successful cases of lung transplantation in patients with these combined diagnoses, which encourages other centers at least to consider this procedure in these patients. Lung transplantation in patients with severe scoliosis can cause severe pulmonary restriction and reduced chest volume due to chest wall abnormalities. Therefore, severe scoliosis is considered a technical contraindication for lung transplantation and may lead to a poor functional outcome [2].

Theoretically, two leading aspects may play a critical role in patients with respiratory diseases and thoracic deformities: challenges in accurate volume predictions

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based exclusively on height, necessitating interventions such as volume reduction surgery or delayed chest closure, as well as complexities arising from severe lung deformities, increasing the risk of an oversized lung allocated according to predicted lung capacity [3]. However, an official guideline for patients with thoracic deformities has not yet been developed and current findings are mainly based on individual case reports. For the first time in literature, we present a case report of our patient with scoliosis who underwent a lobar lung transplantation.

Case report

A 42-year-old female patient with interstitial pulmonary fibrosis and scoliosis was placed on the waiting list for a lung transplantation in February 2018. The indications for lung transplantation were marginal due to dextroscoliosis (with Cobb angle 34°), an extremely thin chest, a poor nutritional condition, with a body mass index of 16,4, and celiac disease. However, after reviewing all considerations, including the socioeconomic status of the patient, who was the mother of a 9-year-old son, highly motivated, young, and well educated, with good rehabilitation potential and no other comorbidities, our multidisciplinary transplant team decided to accept her for lung transplantation. The patient remained on the waiting list for a suitable donor for 38 days (female, brain-death donor) (Fig. 1).

Lung transplantation was performed through a clamshell thoracotomy under extracorporeal membrane oxygenation (ECMO) support. Unfortunately, the recipient's chest cavity was too small relative to the donor's lung, which required the implantation of the middle and lower right lobes and a volume reduction of the lingula on the left side. The ECMO time and total ischemic time were 250 min and 355 min, respectively.

The first X-ray was performed in the hour after lobar transplantation and showed slight pulmonary edema. The partial arterial pressure of oxygen was 10,1 kPa, corresponding to grade 3 primary graft dysfunction. Transthoracic echocardiography revealed mild diffuse hypokinesia of the nondilatated right ventricle. However, in view of progressive reperfusion-related lung edema and excess pressure in the right and left ventricles, urgent rethoracotomy was required to open the chest for prompt adjustment and to decrease vasopressor support and improve respiratory functions. The patient stabilized (hemodynamically and respiratory) slowly in the following days.

The chest wall was closed on the seventh day after LTx in the operating theater under transesophageal cardiography control, which showed good function of both heart ventricles as well as good respiratory function. After monitoring for 4 h in the intensive care unit, tachyarrhythmia, with a heart rate of 180 beats per minute, followed by cardiac arrest occurred. Transesophageal cardiography revealed mechanical oppression of the right ventricle, which forced urgent open rethoracotomy on the bed in the intensive care unit. Clamshell

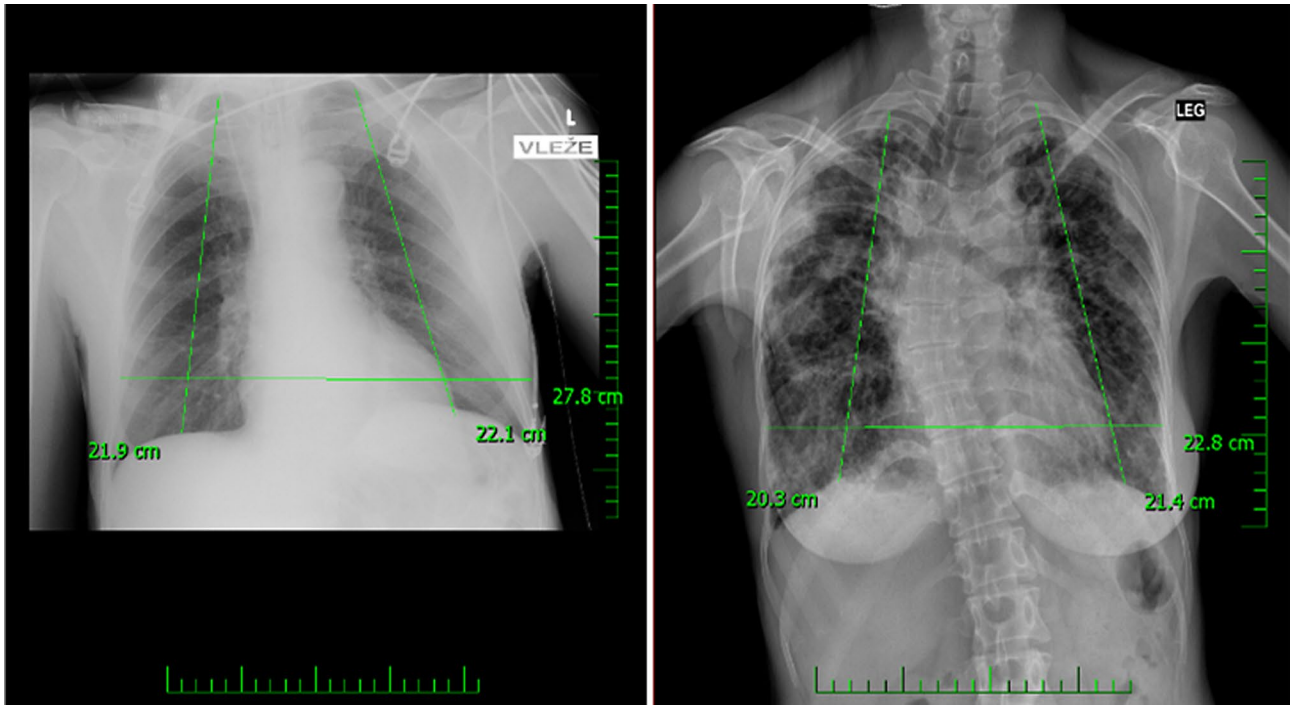

Fig. 1 Chest X-ray – donor left vs. recipient right size match

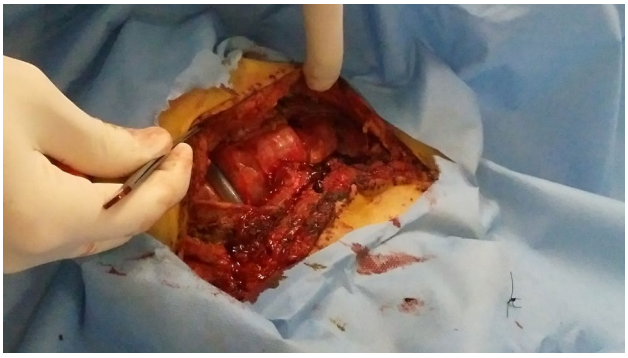

Fig. 2 Partial resection of the distal sternal bone

rethoracotomy closure was performed the same day after circulatory stabilization. However, the mechanical oppression of the chest led to continuous severe circulatory insufficiency and the introduction of an intra-aortic balloon pump. Despite repeated maneuvers, including re-rethoracotomy and closure, there was clear evidence that the distal part of the sternum was still exerting significant pressure on the entire right ventricle, causing a low cardiac output, so severe circulatory instability still progressed, we were forced to perform an emergency partial distal sternal resection (Fig. 2) as a last resort. The sternum was released at the spot of the previous transverse sternotomy and cut approximately 3 cm distally at the site of the greatest pressure on the right ventricle. After this partial resection of the distal sternum the circulation was promptly restored.

The wound was healed by negative pressure wound therapy (NPWT) using a nonadherent layer and foam connected to a vacuum device, with continuous negative pressure of 40 mmHg. The dressing was changed 3 days after the first loading and then after 6 days, with gradual tightening of the thoracotomy. In total, negative pressure therapy was performed 4 times with subsequent skin suturing. A few stiches were added each time to close the wound dynamically (Fig. 3). During the NPWT the heart was protected against NPWT bleeding by covering with a sheet (foil). Negative pressure wound therapy allowed gradual approach of the thoracotomy and closure of the skin wound within 3 weeks. Due to the significantly weak skin cover, any mechanical strengthening (with plates, wires, stitches or other temporarily interposed material) was not possible. NPWT did not interact with chest drains.

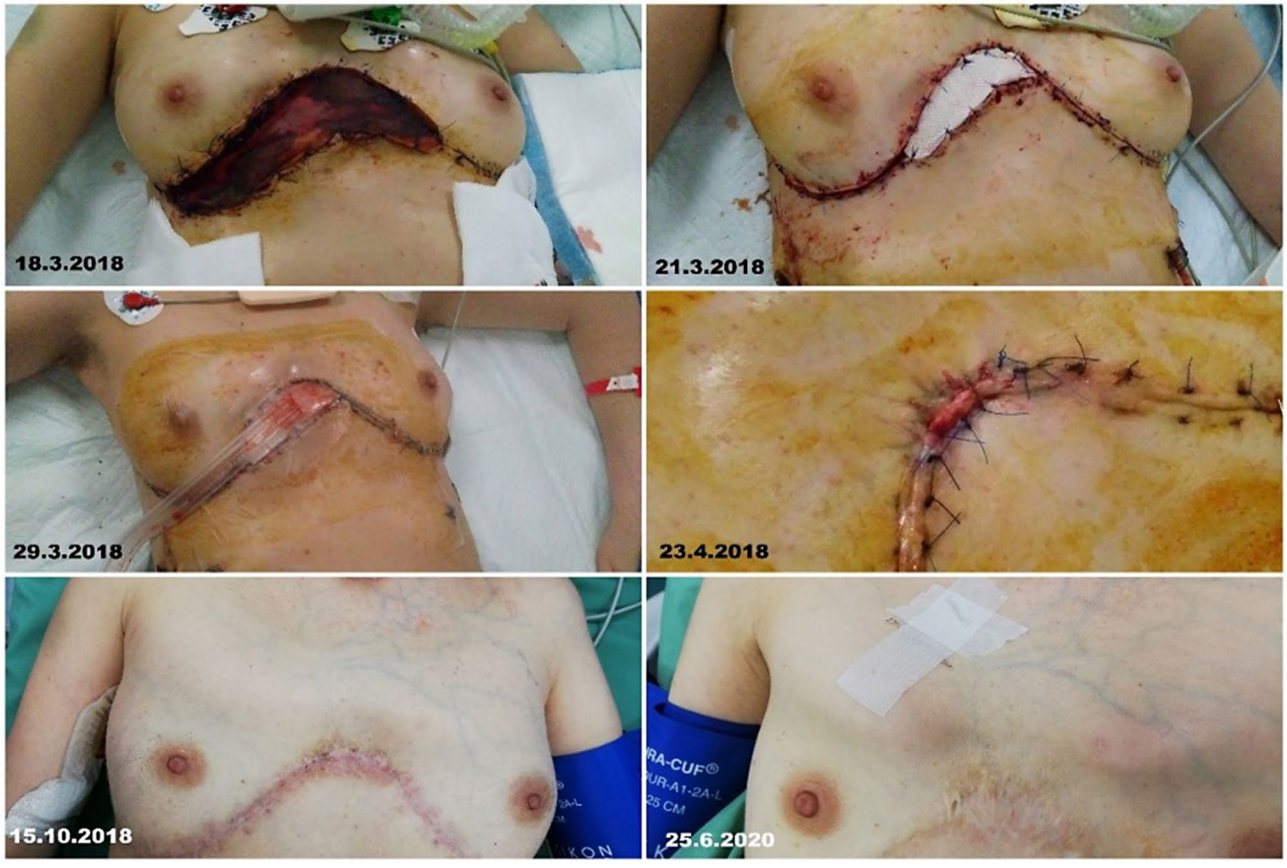

Fig. 3 Wound healing after partial distal sternal resection

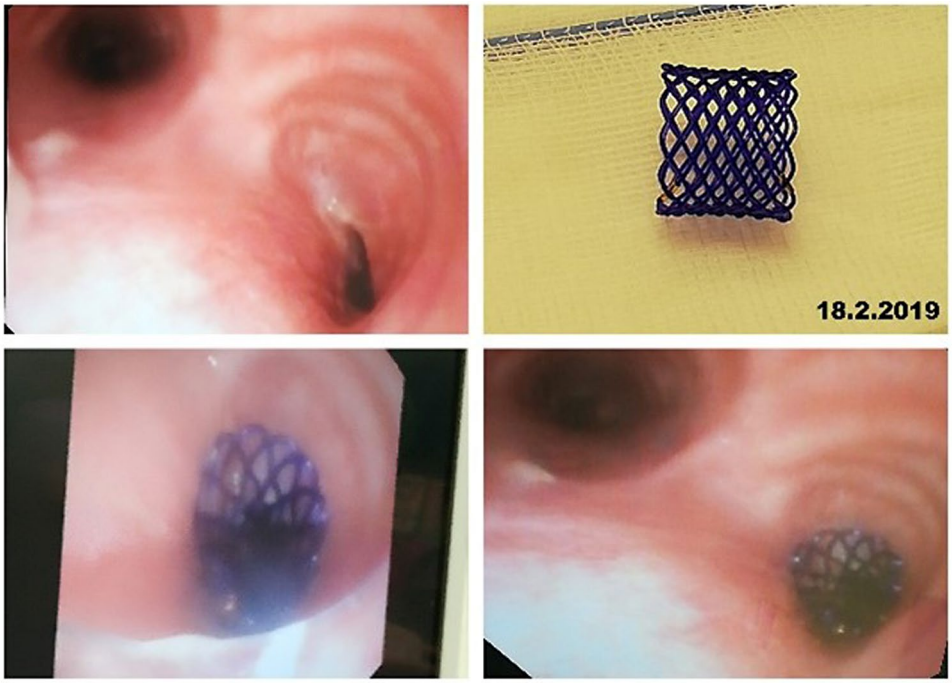

Fig. 4 Right bronchial stenosis and the inserted biodegradable stent

In the further course of hospitalization patient's recovery was accompanied by a few complications. Acute cellular rejection was one of the significant complications of her weak recovery and was treated with pulse corticosteroid therapy. Donor-specific antibodies were identified in blood samples in the third month after LTx and were treated with a regimen of plasmapheresis and intravenous immunoglobulin, followed by alemtuzumab. The first transbronchial lung cryobiopsy of this fragile patient was performed three and half months after LTx, with any histopathological signs of rejection. The postoperative course was further complicated with recurrent pulmonary infection caused mainly by multiresistant *Pseudomonas aeruginosa*, *Enterococcus faecium*, *Klebsiella pneumoniae*, *Candida glabrata*, *Corynebacterium* and *Chryseobacterium indologenes* treated according to antibiotic sensitivity. Surveillance bronchoscopy demonstrated 50% bronchial stenosis in the bronchus intermedius, which did not require any intervention at that time. She was weaned from the ventilator breathing machine five months after transplantation and stayed partly dependent on nasal oxygen therapy. She was discharged from the hospital 228 days after LTx.

Nevertheless, the question remained whether it is realistic to somehow repair the defect between the proximal and distal part of the sternum. Given the complexity of the whole case and at the same time the very intimate relationship between the sternum and the heart, let time mature.

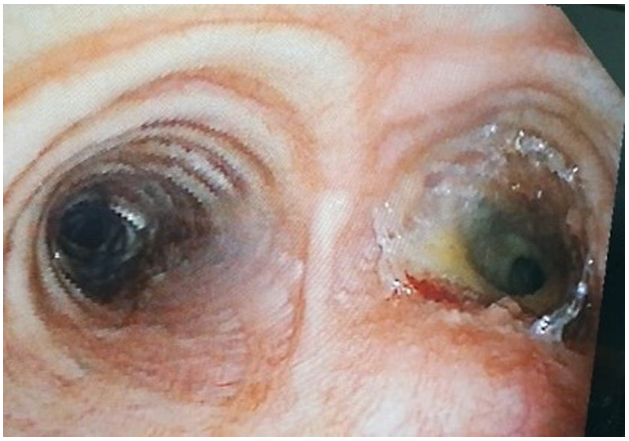

Fig. 5 Right bronchial stenosis with a previous biodegradable stent before the insertion of a new BD stent

She underwent flexible bronchoscopy six months after discharge. Bronchoscopy revealed almost complete right bronchial stenosis with bronchial obstruction. Stenosis was dilatated and treated by inserting a biodegradable (BD) stent (Fig. 4). Additional sessions have been performed every three-four month thus far, with the last insertion of a biodegradable stent in December 2022 (Fig. 5), in total of 15 inserting of BD stent, size of 20 × 15 mm within the first two session and continued with size of 25 × 15 mm. She tolerated the stent implantation well. We planned regular replacement of the stent every three months. An earlier stent exchange was performed due to expectoration of the parts of the previous

biodegradable stent. We tried twice to terminate another stent placement but each time there was a narrowing of the bronchus. After four years, the bronchus finally solidified. The patient has been without a stent for a year and the right bronchus shows only minimal flattening (Fig. 6). After five years, the chest wall defect remains unresolved. It is obvious that a sternal defect causes a mechanical breathing disorder, however, any reconstruction of the chest wall is still highly risky even with the passage of time.

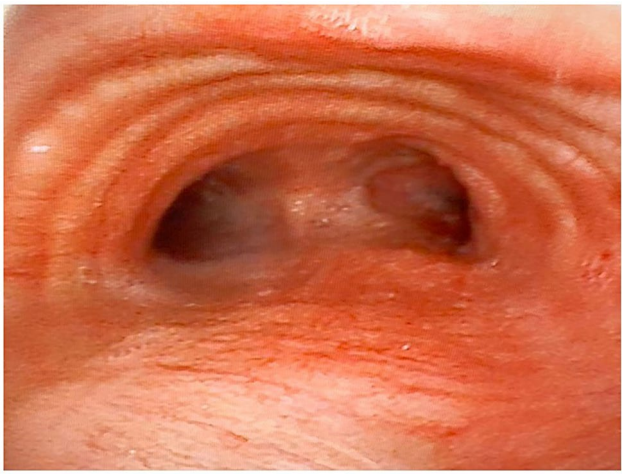

Fig. 6 Bronchoscopy after one year of no-stenting

Discussion

Skeletal thoracic deformities are known to produce functional disturbances in the cardiovascular system. Some patients with end-stage lung disease and severe scoliosis are denied lung transplantation. There is still a lack of evidence and reviews that may clarify the treatment approach in these patients.

Based on the literature and our experience, in patients with scoliosis considering lung transplantation, size match, straight back syndrome, delayed chest closure and bronchial stenosis are key issues clinicians should evaluate.

Piotrowski says that exact determination of the size of the donor lung using exact measurements on chest computed tomography makes it possible to obtain a very well-matched organ. Kyphoscoliosis and mediastinal shift require careful donor-lung sizing with computed tomography and is not dependent on typical parameters [4, 5]. It is very important to study recipient tomography scans and patient chest defect anatomy caused by scoliosis and very carefully and accurately compare the results with donor size [2]. It is essential to compare not only the anterior-posterior view but also the lateral view and the depth of both donor and recipient chest cavities. There are also many centers using donor and recipient total lung capacity (TLC) volumes for perfect size matching. In patients without any chest deformities X-ray measures correlate much more strongly with true lung volumes than height, weight, or predicted TLC [6]. But there is still no strong evidence in patients with scoliosis. In our case measurement of TLC was not used.

Straight back syndrome is the congenital loss of normal physiological mid-upper thoracic kyphosis. This thoracic deformity biomechanically decreases the distance between the spine and the sternum, compressing the internal structures, namely, the heart. As Raggi et al. stated, "the heart appears trapped in a chest cavity too small for its size, and its anatomic architecture is seemingly altered in attempt to accommodate these insufficient dimensions [7]. More rarely, the narrowed upper chest may compress the trachea, causing extrinsic tracheal obstruction. If severe enough, this requires operative repair by effectively opening up the upper chest by a variety of techniques to allow the trachea enough room to avoid compression." [8] The right ventricular outflow tract is probably more affected because of its anterior position and its proximity to the sternum. The compression of the heart between the sternum and the vertebral column gives the impression of an increase in the transverse diameter of the heart in some patients. Increasing pulmonary edema of newly transplanted lungs can worsen the reduced space in the thorax, and the weight of mainly the right lung can drag it down, which can lead to the compression of the right ventricle, mainly shortly after lung transplantation. Later, due to the oversized of the donor lungs, the narrow space between the sternum, the heart, and the kyphoscolytically deviated spine to the right, the right ventricle can be oppressed with subsequent destabilization, which is exactly what happened in our patient.

What can also be witnessed in straight back syndrome is the dilatation of the right atrium as a result of congestion in front of the compressed right ventricle (Fig. 7), which can already be seen on CT scans before transplantation.

Stabilizing patients on prolonged ECMO or with an opened chest after lung transplantation to avoid ischemia-reperfusion injury and edema and potential straight back syndrome remains an issue. If pulmonary lung function is not adequate to maintain life, chest opening might be the right choice. Delaying chest closure after lung transplantation to relieve pressure on the heart and lungs improves tidal volume without affecting airway pressure and improves biventricular function, exerting less positive end-expiratory pressure [9, 10]. Delaying chest closure releases compartment pressure over the lungs and at the same time keeps the lungs confined to a certain extent. Delaying chest closure not only allows sufficient time for improving lung function and resolving edema but also prominently avoids lung volume reduction or at

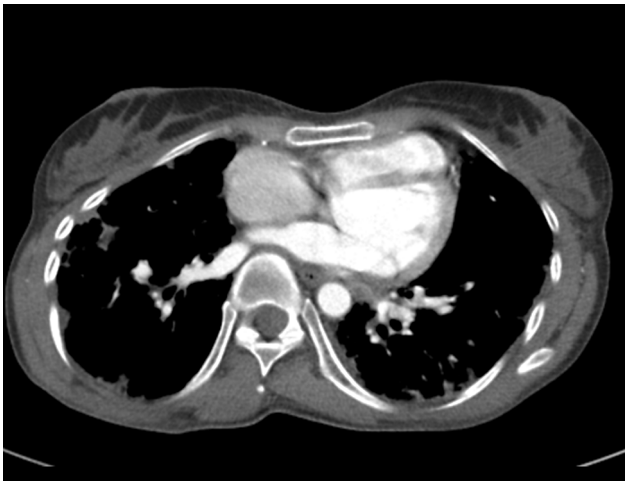

Fig. 7 Dilatation of the right atrium in front of the compressed right ventricle

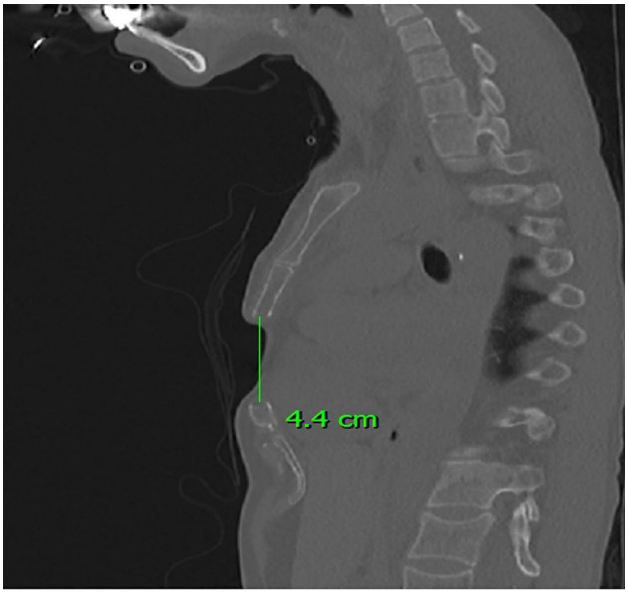

Fig. 8 Chest wall defect after five years on computer tomography scan

least the quantity of lung reduction [11]. This technique may also avoid the use of extracorporeal life support, especially with oversized grafts with primary graft dysfunction, by stabilizing hemodynamic status and improving gas exchange. Force and associates reported 5 days as the mean time to close the chest [8]. For these reasons, and especially for oversized allografts, it is often difficult to close the chest after lung transplantation. Such attempts may cause hemodynamic instability and desaturation from ventilation due to compartment-like syndrome in the chest. In our case, the problem was not in the oxygenation of the lungs, but in the size of the lungs, causing compression of the right heart, which ECMO probably would not solve.

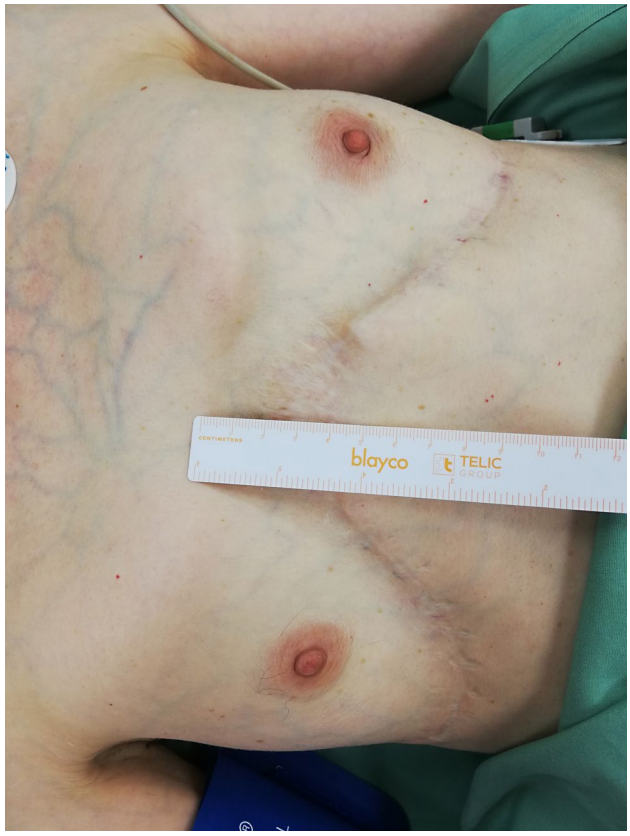

Fig. 9 Chest wall defect after five years after lobar lung transplantation

Negative-pressure wound therapy (NPWT) is the therapeutic procedure used especially in traumatic soft-tissue wounds or infections in thoracotomy or sternotomy. In our case we did not use NPWT to heal as a chronic wound, but we mainly used its ability as a dynamic suture with replacement of the vacuum cover at low (40 mmHg) negative pressure with gradual tightening of the thoracotomy. Negative pressure therapy enabled complete closure of the wound without signs of infection, but mainly temporary or even permanent protection of the heart.

During the careful follow-up of the patient over the course of several years, there was still a slight gaping of the thoracotomy, but the defect between the proximal and distal parts of the sternum did not enlarge on CT (Fig. 8) and clinically (Fig. 9). The reason is probably based on numerous adhesions between the heart and sternum, and between the lungs and the thoracotomy. Definitive reconstruction of the chest wall appears to be high risk.

Airway complications occur after lung transplantation in 2–18% of patients [12]. Bronchial stenosis has been reported as a typical complication in lung transplant recipients with scoliosis [13]. From available sources, bronchial stenosis is recorded more often in the right bronchus [13, 14]. The right bronchus can be directly compressed between the lung and vertebra. Lower

vascularization and a lack of blood leading to ischemic changes might be other reasons. Bronchial stenosis was treated with end bronchial stenting and biodegradable stents. The diameter and length of the stenotic bronchus must be exactly measured [15, 16]. The stent is self-expandable and is well tolerated in all our patients. The stent usually dissolves in three months.

The description of our experience with this patient might lead clinicians to consider scoliosis a contraindication to lung transplantation. However, this case was successful, meaningful and reflects the complexity of the lung transplantation procedure from donor procurement until postoperative care. Despite prolonged recovery, the patient is doing well with a good quality of life and is back to work.

In summary, perfect donor-recipient size matching is essential for patients with scoliosis. Use all possible methods of lung size measurement for accurate matching, such as X-ray measurement, CT volume measurement, TLC, or even nowadays the possibility to use virtual reality. In case of oversized donor´s lung and recipient scoliosis, it pays to let the chest open, wait until the pulmonary edema subsides and under echocardiography close the thoracotomy. In the follow-up period patients with scoliosis should require regular bronchoscopy for at least one year, to manage potential more frequent bronchial complications associated with these recipients.

Conclusion

Lung transplantation in patients with end-stage lung disease and severe scoliosis can be challenging regardless of potential complications. It is vital to determine donor-recipient size match accurately to avoid lung volume reduction, which may lead to additional complications. Exact measurements on chest computed tomography and comparing computer tomography images between the donor and the recipient are essentially helpful. Delaying chest closure is the method of choice for stabilizing postoperative lung ischemia-reperfusion edema, primarily in cases of very small chest cavities and oversized donor lungs. Right bronchial stenosis is often a postoperative complication. Surveillance flexible bronchoscopy may prevent the risk of bronchial deformation or complete bronchial obstruction with recurrent pulmonary infection. Lung transplantation in patients with scoliosis is feasible with an exact size match, ECMO support and experienced postoperative care.

Abbreviations

BD	Biodegradable
ECMO	Extracorporeal membrane oxygenation
LTx	Lung transplantation
NPWT	Negative pressure wound therapy
TLC	Total lung capacity

Author contributions

Svorcova wrote the main manuscript and is the corresponding author. Vachtenheim J. and Havlin J. made a corrections. Simonek J., Vajter J., * Prikrylova Z., * Kolarik J., Pozniak J., Lischke R. participated on the discussion.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Competing interests

The authors declare no competing interests.

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