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SUCCESSFUL ENDOSCOPIC MANAGEMENT OF SWYER–JAMES–MACLEOD SYNDROME: A CASE REPORT

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Swyer–James–MacLeod syndrome is a rare disease characterized by emphysematous transformation of an entire lung or lobe. Traditionally, the main treatment method has been surgical resection of the affected lung or lobe to reduce compression of adjacent healthy lung tissue and improve vital lung capacity. This article presents a clinical case of successful endoscopic treatment in a patient with emphysematous transformation of the entire lung, who was referred to the transplant center as a potential candidate for lung transplantation.

Keywords: interventional bronchoscopy, thoracic surgery, Swyer–James–MacLeod syndrome, panlobular emphysema, endoscopy, valve bronchoblockade.

INTRODUCTION

Swyer–James–MacLeod syndrome was first reported in 1953 by P.R. Swyer and G.S. James as "unilateral pulmonary emphysema". A year later, John MacLeod reported a similar case, describing it as "abnormal transparency of one lung" [1]. MacLeod suggested that the underlying cause was obliterative unilateral bronchiolitis, developing as a consequence of recurrent respiratory infections during childhood. However, current literature generally attributes the condition to a congenital or developmental defect, characterized by hypoplasia of the pulmonary artery and small bronchi [1, 2].

The disease may remain asymptomatic until adulthood or present with nonspecific respiratory symptoms, including productive cough, exertional dyspnea, hemoptysis, reduced exercise tolerance, and recurrent pulmonary infections. The disease can be unilateral or bilateral, but it typically affects a single lobe or lung. Despite these possible manifestations, most cases remain clinically silent for years and are often diagnosed incidentally in adulthood [3, 4].

Due to similar clinical manifestations, MacLeod syndrome is frequently misdiagnosed as chronic obstructive pulmonary disease, bronchial asthma, pneumothorax, or pulmonary embolism. The condition should be suspected in patients with presumed asthma who do not respond to conventional therapy [5, 6]. In childhood, the disease usually has minimal clinical impact, but in adulthood, it may progress and sometimes require surgical intervention [7, 8].

Diagnosis is most often suspected based on imaging studies. Chest radiography typically reveals hyperlucency and hyperinflation of one lobe or lung, accompanied by reduced volume of the contralateral lung (Fig. 1). Spirometry shows obstruction, while lung perfusion scintigraphy shows a noticeable reduction in perfusion of the affected lung, with normal or increased perfusion on the opposite side [9, 10].

The primary surgical approach involves resection of the affected lung or lobe to enhance ventilation of the remaining functional alveolar tissue. Despite decades of surgical experience in managing bullous emphysema of various etiologies, lung volume reduction remains the most commonly employed treatment strategy for this condition [10].

In 1990, a group of Russian surgeons successfully introduced an alternative technique – transthoracic occlusion of the bronchus supplying the affected lung, allowing preservation of pulmonary parenchyma. However, no subsequent reports describing its use have appeared in either Russian or international literature [11, 12].

CLINICAL CASE

A 24-year-old female patient (patient X) presented for the first time with respiratory failure during physical exertion. Chest X-ray initially suggested right-sided pneumothorax, and she was hospitalized locally; however, this diagnosis was not confirmed. Subsequent CT scan revealed total bullous emphysema of the right lung§ and mediastinal pulmonary hernia.

Her medical history was notable for frequent respiratory infections during childhood. Following additional investigations and exclusion of other pathologies, she was diagnosed with McLeod syndrome. After discharge, the patient continued to experience recurrent frequent colds, and from October 2019, she began to experience shortness of breath at rest.

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A thoracic surgeon evaluated her and found that surgical intervention was contraindicated due to a significant decrease in external respiratory function, recommending consultation with a transplant specialist. In January 2020, she underwent remote consultation at Shumakov National Medical Research Center of Transplantology and Artificial Organs, where hospitalization at the transplant center was advised to determine further management.

At the Shumakov Center, examination findings were as follows: forced expiratory volume in 1 second (FEV_1): 1.03 L (25%), forced vital capacity (FVC): 0.82 L (23%), maximal inspiratory pressure (MIP): 79%, maximal mid-expiratory flow rate ($MMEF_{25–75}$): 0.69 L/s (17%), echocardiography: no signs of pulmonary hypertension, perfusion scintigraphy: normal perfusion volume in the left lung. CT findings revealed severe bullous emphysema of the right lung with mediastinal shift and compression of the left lung (Fig. 2).

Based on the results of a comprehensive examination, it was determined that, despite severe respiratory failure, the patient's overall physical condition remained satisfactory. Consequently, a decision was made to insert an endobronchial valve in the upper lobe of the affected lung with the aim of reducing hyperinflation and alleviating

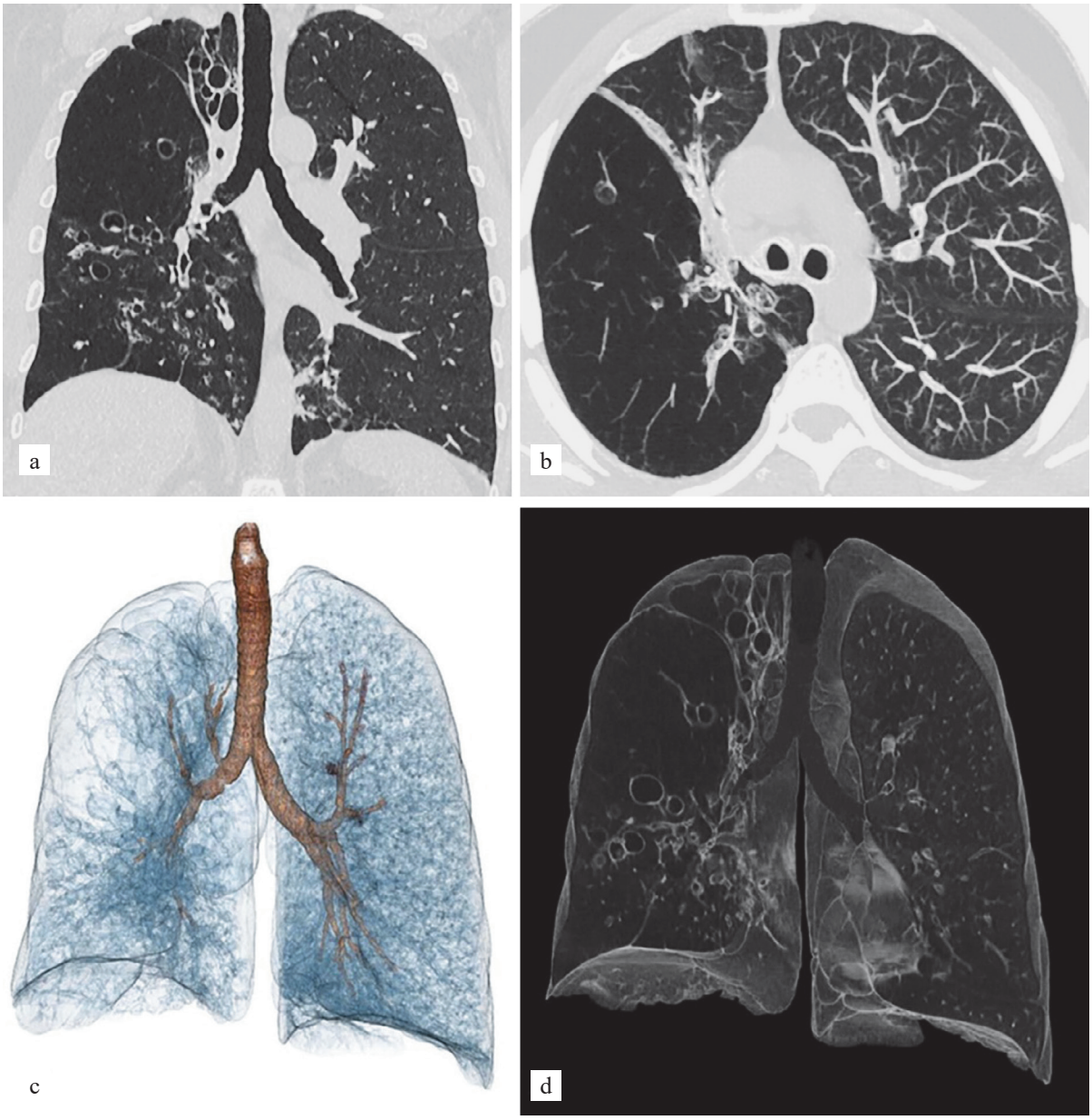

Fig. 1. Computed tomography findings in an adult patient with Swyer–James–MacLeod syndrome: a, frontal projection; b, axial maximum-intensity projection; c, d, volume-rendered chest CT images show diffusely decreased attenuation of the right lung with signs of marked hypoperfusion and bronchiectasis, some with mucous plugging. There is volume reduction of the right lung, mainly in its upper lobe, with asymmetry of the pulmonary arteries, which are preserved in the left lung. (Source: L.P. Gomes de Farias et al. Swyer–James–MacLeod Syndrome: The Hyperlucent Lung. Radiology: Cardiothoracic Imaging)

compression of the contralateral (healthy) left lung. The procedure was carried out under general anesthesia.

One month after the insertion, a positive clinical response was observed, characterized by improved exercise tolerance and reduced dyspnea. It was concluded that there were no current indications for lung transplantation, and outpatient follow-up was continued.

Four months after insertion of the endobronchial valve, the patient reported a deterioration of her condition, returning to the pre-intervention level. Computed tomography revealed proximal migration of the bronchial blocker, resulting in recurrent hyperinflation of the upper lobe of the right lung with secondary compression of the left lung. The displaced valve was removed, and a repeat endobronchial valve insertion was performed – this time targeting both the intermediate and upper lobes of the right lung (Fig. 3).

Five days after the second intervention, despite some reduction in respiratory insufficiency, the patient's condition remained unfavorable. A sharp displacement of the mediastinum secondary to subtotal atelectasis led to severe retrosternal pain, although there were no signs of cardiovascular compromise.

Given the ineffectiveness of repeated insertion of endobronchial valves, a decision was made to perform circumferential argon plasma coagulation to induce fibrotic stricture formation, thereby decreasing ventilation and reducing hyperinflation of the affected lung. The procedure was performed under high-frequency ventilation (HFV) using an electrosurgical coagulator (ERBE VIO3, PRESICE mode).

In the postoperative period, the patient experienced an exacerbation of chronic bronchitis, which was managed with antibiotic therapy.

Two weeks after the intervention, the patient showed a decrease in respiratory insufficiency. During outpatient follow-up at 3, 6, and 12 months, there was sustained improvement in respiratory function, with significant regression of respiratory failure, increased exercise tolerance, and no episodes of acute bronchitis from the third postoperative month onward.

Computed tomography performed one year after the procedure revealed complete atelectasis of the affected right lung, atresia of the intermediate and upper lobe bronchi, a mediastinal shift toward the right, and compensatory hyperinflation of the contralateral (left) lung. Despite occasional mild chest discomfort, the patient's

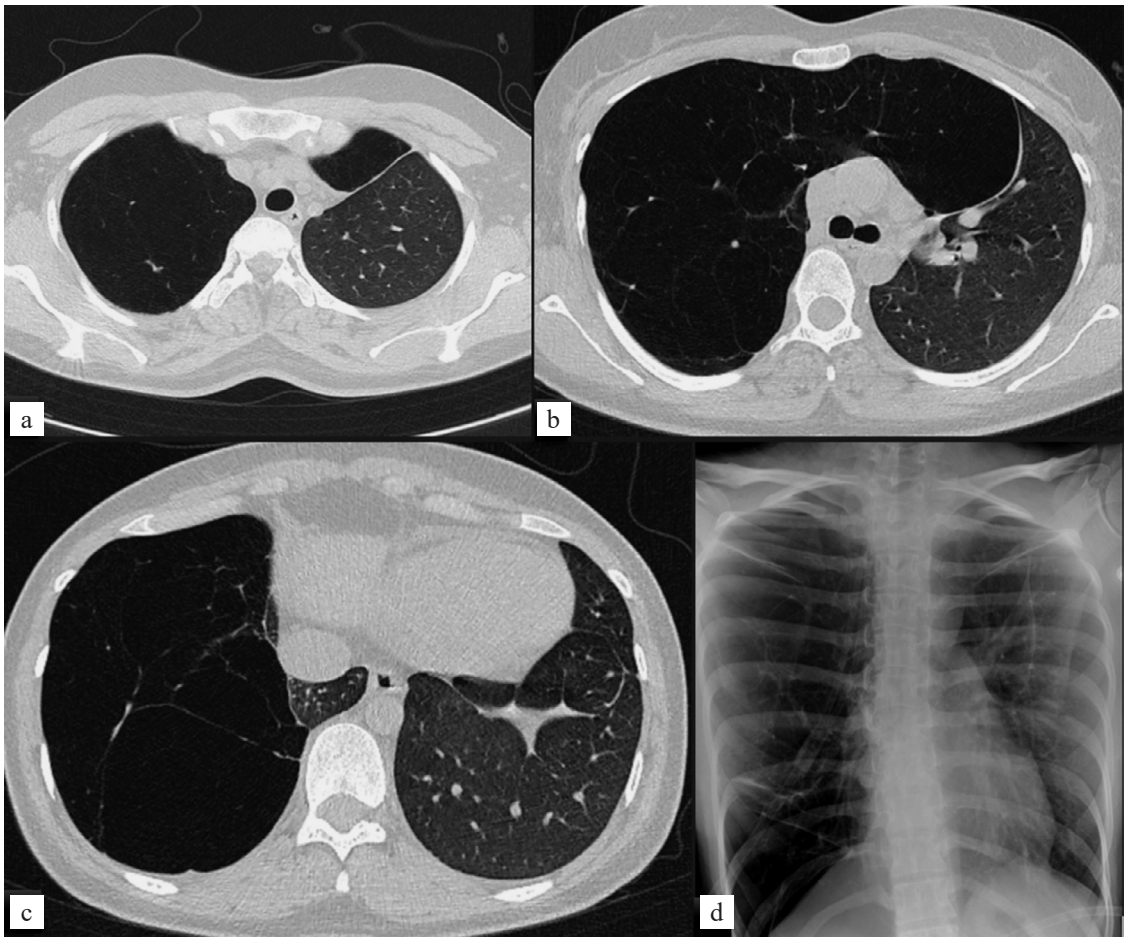

Fig. 2. Radiological findings prior to surgical intervention: a, 3rd thoracic vertebra; b, 5th thoracic vertebra (tracheal bifurcation); c, 7th thoracic vertebra (beginning of the diaphragm); d, frontal chest X-ray

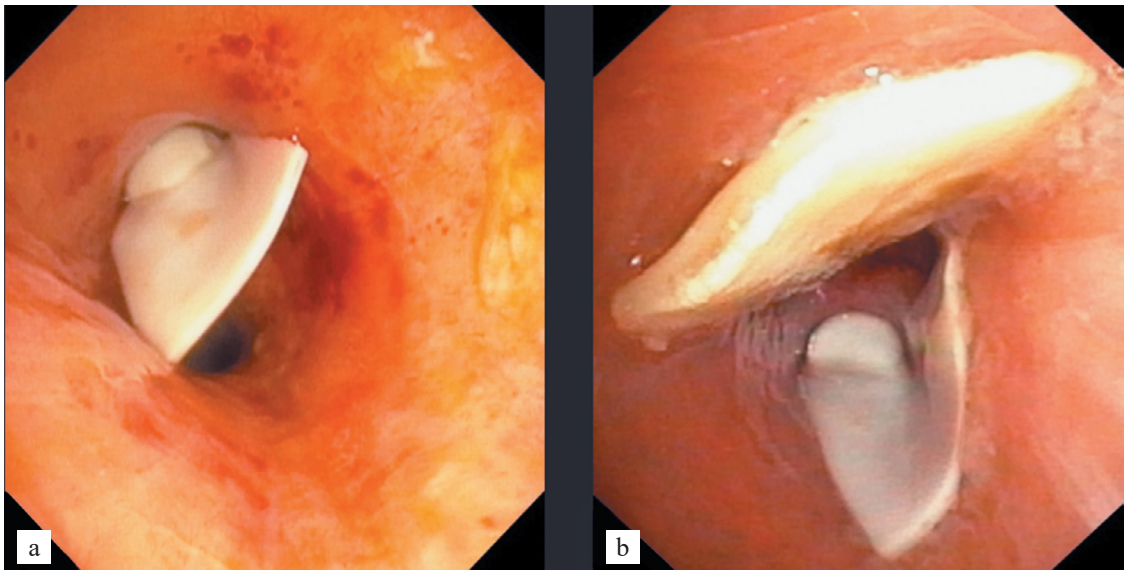

Fig. 3. Endoscopic valve bronchoblockade of the upper lobe of the affected right lung: a, upper lobe; b, intermediate and upper lobe bronchi

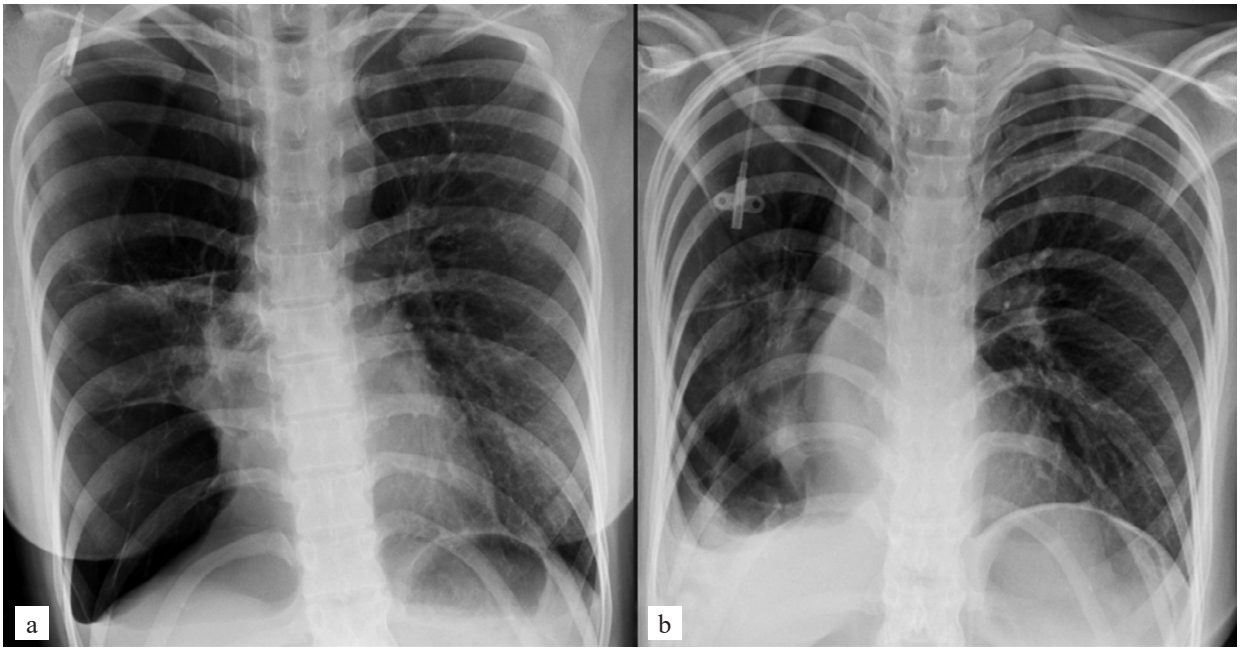

Fig. 4. Control chest X-rays following the insertion of an endobronchial valve into the right upper lobe and intermediate bronchi: a, 3 days post-procedure; b, 2 weeks post-procedure

pulmonary function parameters improved substantially, with FEV_1 2.15 L (56%), FVC 1.87 L (56%), MIP 87%, and $MMEF_{25–75}$ 1.74 L/s (43%). No evidence of heart failure was observed on echocardiography, ECG, or functional tests (Figs. 5 and 6).

CONCLUSION

Throughout the entire inpatient and outpatient observation period, spanning more than three years, no serious infectious or cardiovascular complications were recorded. This fact underscores the safety and effectiveness of the treatment approach applied. Moreover, the patient achieved not only a sustained, but also a significant improvement in respiratory function, which was accompanied by a notable increase in exercise tolerance.

The positive outcomes suggest that endobronchial valve therapy with subsequent argon plasma coagulation may represent a highly effective alternative to radical surgical procedures. This is particularly significant for patients with Swyer–James–MacLeod syndrome and other conditions associated with panlobular emphysema. Thus, implementation of this therapeutic approach has the potential to enhance clinical outcomes and signifi-

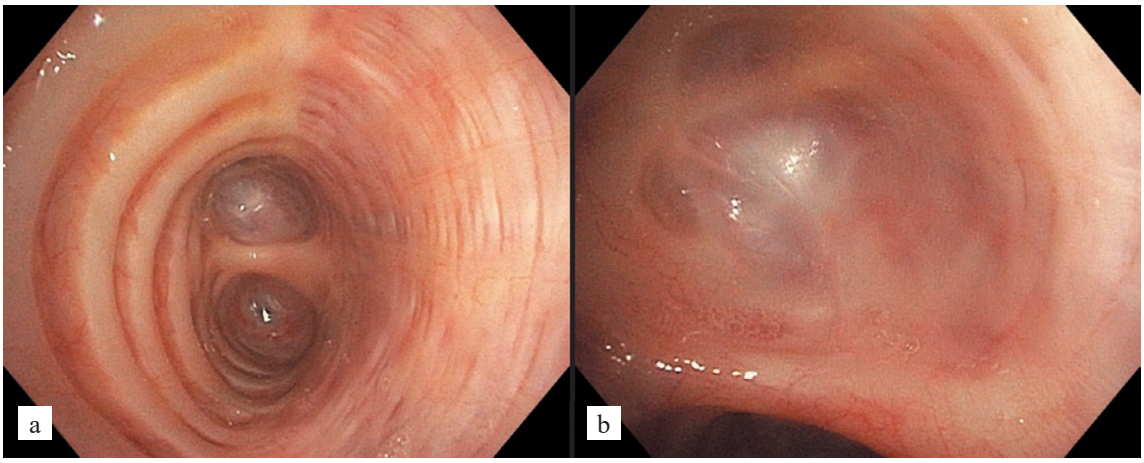

Fig. 5. Endoscopic views of complete atresia of the right upper lobe and intermediate bronchi: a, atresia of the intermediate and upper lobe bronchi on the right; b, atresia of the upper lobe bronchus on the right

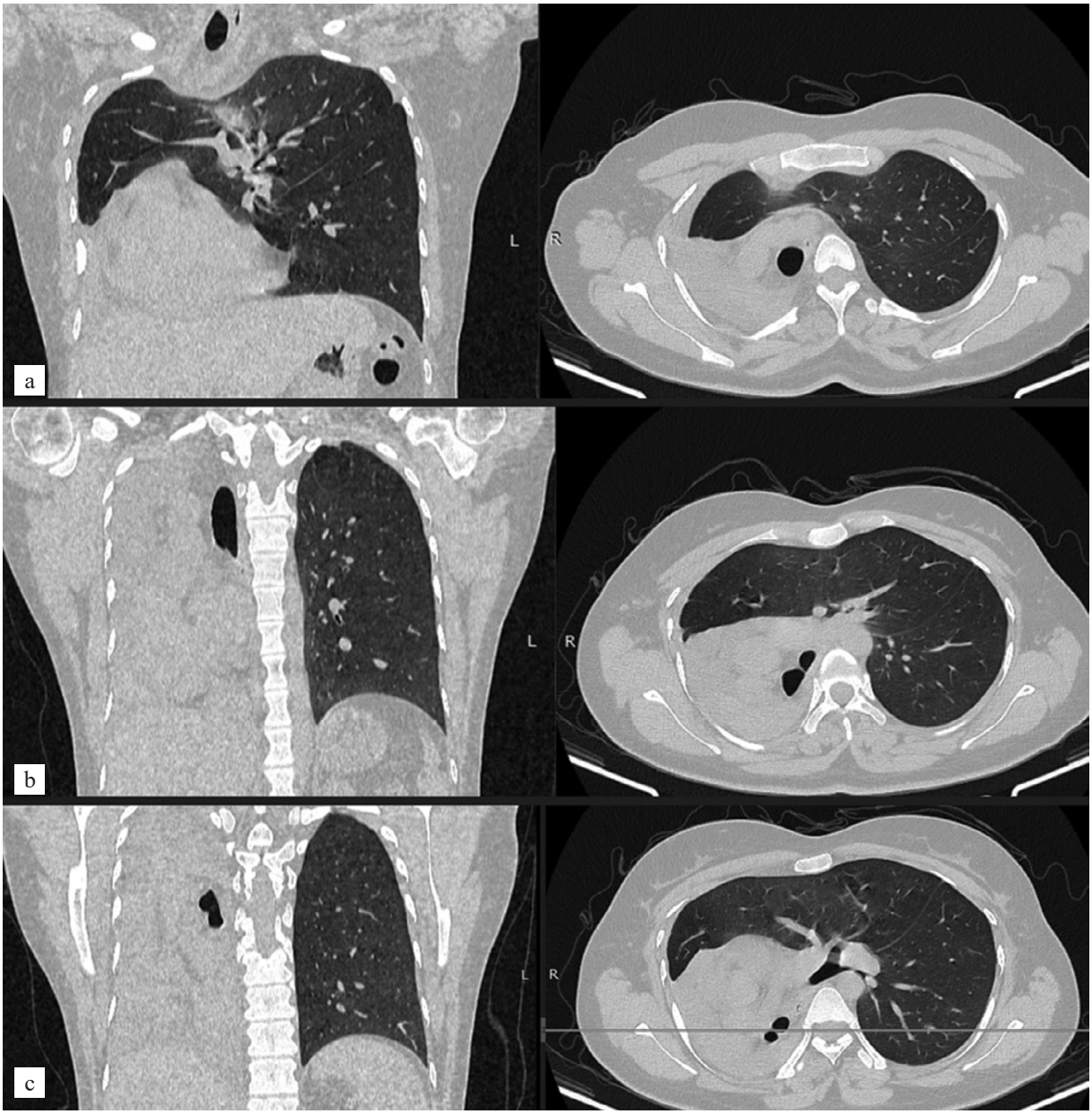

Fig. 6. Computed tomography findings 1 year after endoscopic treatment: a, 3rd thoracic vertebra; b, 5th thoracic vertebra (tracheal bifurcation); c, 7th thoracic vertebra (site of right main bronchus atresia)

cantly improve patients' quality of life, while avoiding the risks inherent in major surgical interventions.

The authors declare no conflict of interest.

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