

Adult patient with pulmonary agenesis: focusing on one-lung ventilation during general anesthesia

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Abstract: Congenital pulmonary agenesis is a rare condition with high mortality. Mechanical ventilation in these patients is challenging and there has no such case been reported in the literature. We reported a 61-year-old female with lung agenesis who presented to our hospital with pneumonia and pelvic mass. In the past, she had undergone repairing of atrial septal defect and mitral valve forming surgery at 6-year-old. Thereafter she had remained asymptomatic until this time of hospital admission. The patient underwent operation for the pelvic mass with one-lung ventilation (OLV) under general anesthesia. We highlighted the use of protective ventilation (PV) strategy during OLV.

Keywords: Congenital abnormalities; lung agenesis; one-lung ventilation (OLV); protective ventilation (PV)

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Introduction

Pulmonary agenesis is a rare congenital anomaly with a reported incidence of 1:15,000 autopsies. The pathogenesis of the disease is largely unknown and it can be classified into bilateral and unilateral forms. The bilateral form is incompatible with life. Herein, we reported a case of 61-year-old female presented to our hospital with pelvic mass and then underwent operation under general anesthesia. One-lung ventilation (OLV) and protective ventilation (PV) strategy was adopted during anesthesia. Unilateral pulmonary agenesis was diagnosed by chest computed tomography (CT) before operation.

Case presentation

A 61-year-old female presented to the hospital with cough and fever for one week. She also complained abdominal

distension for 3 months. There was no chest pain, dyspnea or vomiting. Her medical history included repairing of atrial septal defect and mitral valve formation when she was 6-year-old. Past histories of her parents were unremarkable. She had been born at full term, and there had been no complications during pregnancy. On physical examination, she appeared alert, acyanotic, and breathed peacefully. However, the movement of her left chest decreased remarkably with breathing, and there was no breathing sound on this side. A grade II/VI systolic murmur was noted at the left sternal border. The second sound was higher than the first one on the pulmonary valve auscultation area. There was no tenderness or organomegaly on abdominal examination. The neurologic examination was unremarkable.

Chest X-ray performed at admission showed opacity on the bottom left thorax (*Figure 1*), together with deviation of the trachea and mediastinum to the left. Chest computed

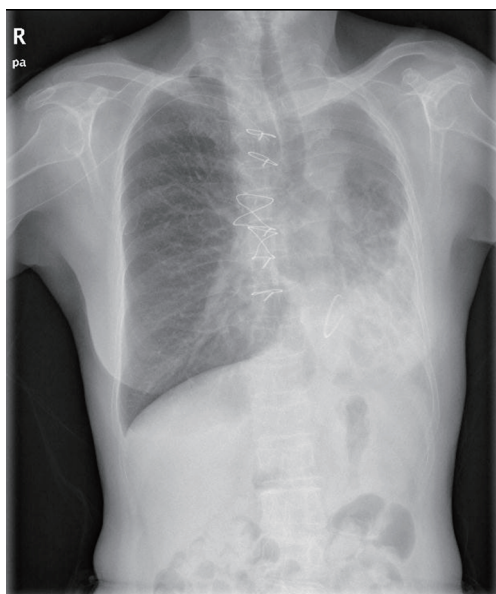


Figure 1 Plain chest X-ray showing left pulmonary agenesis with mediastinal shift and right lung hyperinflation.

tomography (CT) showed collapse of the left lung with respiratory dead spaces and the right lung enlarged for compensation (*Figure 2*). A diagnosis of left lung agenesis was therefore established. CT 3D reconstruction showed dysplasia of the left lung with compensatory hypertrophy of the right lung, and the left pulmonary artery was thin with leftward deviation of the mediastinal structures (*Figures 3,4*). An electrocardiogram revealed sinus rhythm with a ventricular rate of 70 beats per minute, and there was a left-axis deviation. There were no changes in upper abdominal structures. Bronchoscopy revealed a blind-ending left grade three bronchus. Culture of protected specimen brush revealed *Pseudomonas aeruginosa* which was susceptible to ciprofloxacin. Treatment with intravenous fluids and antibiotic IV ciprofloxacin (400 mg q12h) were initiated, and oxygen was given by nasal catheter. Histopathology showed that the right lung was hyperinflated with normal tissues. The left lung showed atrophy and some of the tissue presented bronchiectasis (*Figure 5*).

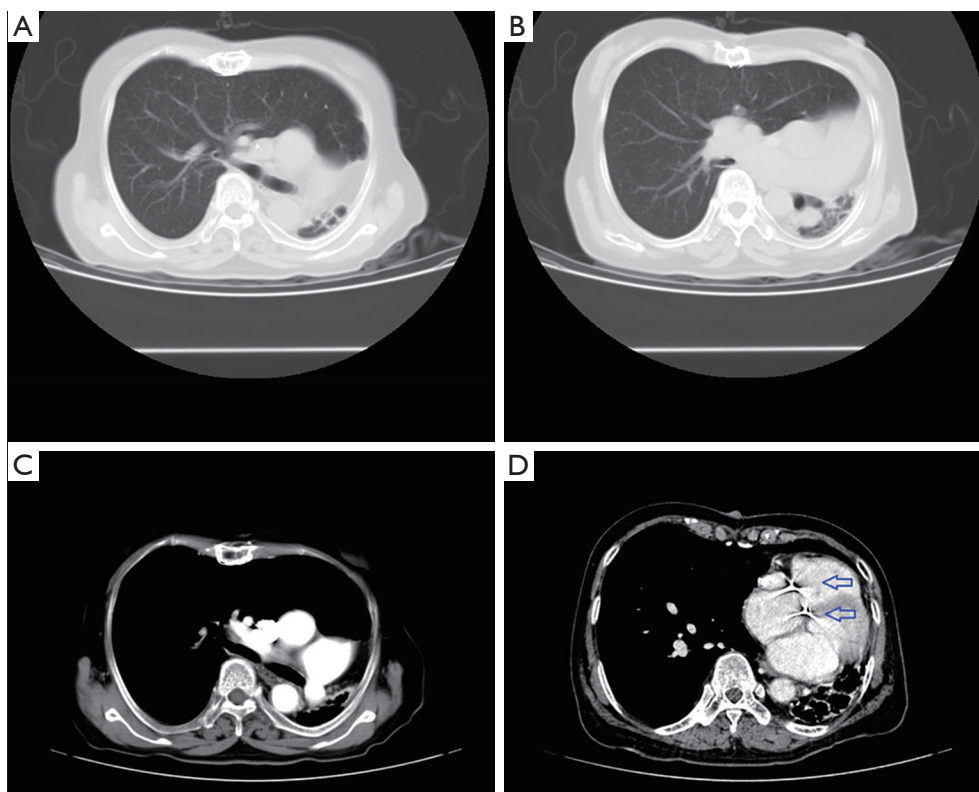


Figure 2 Chest CT scan. (A,B) Axial CT shows hypoplastic left lung with respiratory dead spaces and the right lung showed compensatory enlargement; (C,D) axial CT of mediastinum window showed cardio-mediastinal shift to left side and Carpentier-Edwards ring was visible (blue arrow). CT, computed tomography.

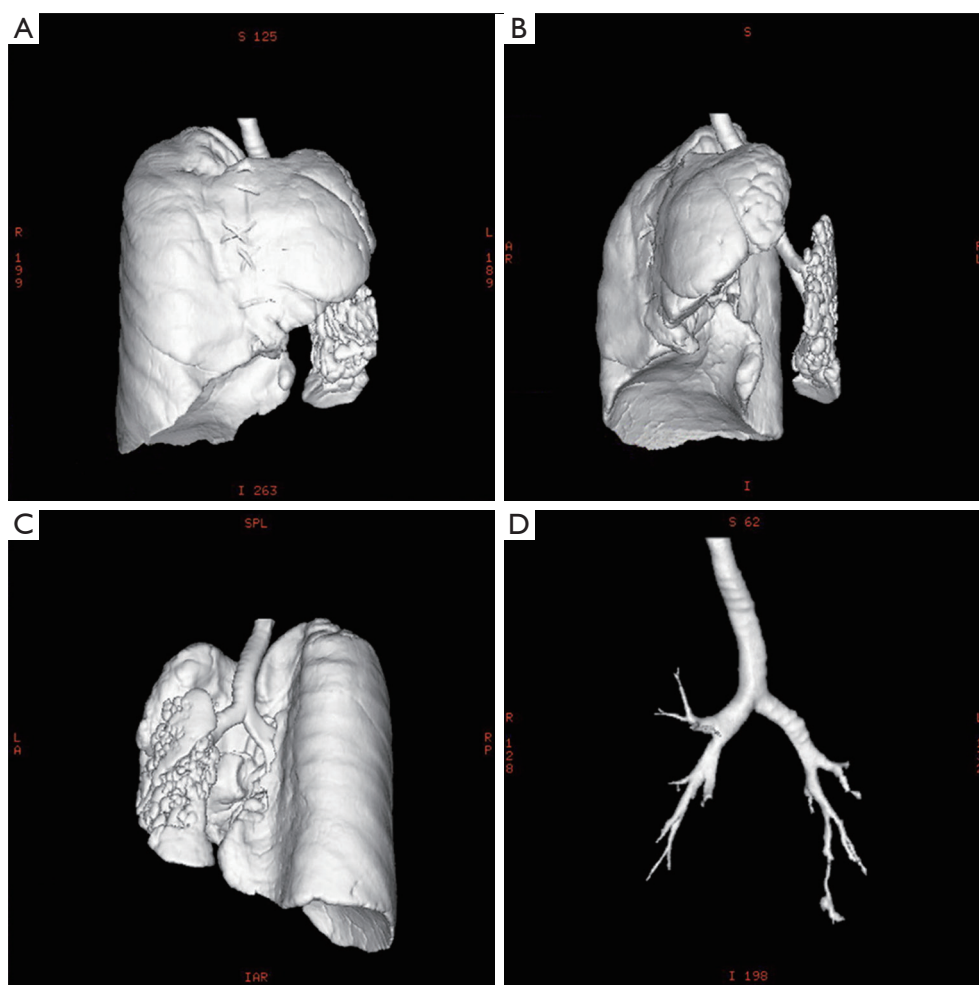


Figure 3 3D reconstruction of lung and bronchial tree. (A,B,C) 3D reconstruction of CT scan showed the collapse of the left Lung and the right lung showed compensatory enlargement; (D) the bronchial tree showed main bronchus shift to the left side and the wall of left bronchi is relatively thin. CT, computed tomography.

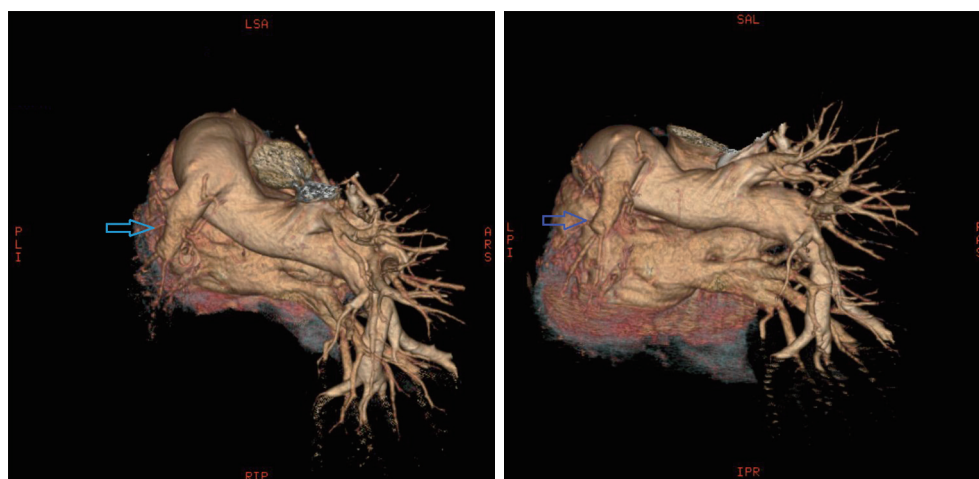


Figure 4 3D reconstruction showed thin left pulmonary artery with leftward deviation of the mediastinal structures (blue arrow).

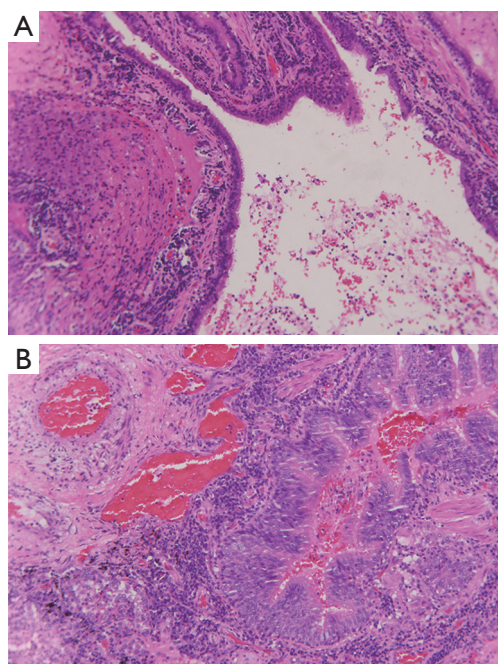


Figure 5 Pulmonary pathology. (A) Histopathology showed left lung atrophy and some of the tissue showed signs of bronchiectasis; (B) the right lung was hyperinflated with air (HE 100×).

Laboratory investigations revealed mild elevation in arterial partial pressure of carbon dioxide ($\text{PaCO}_2 = 53$ mmHg). Doppler echocardiogram showed that the heart was positioned in right thorax. The atrioventricular level shunt and anomalous pulmonary venous return were not seen and pulmonary artery pressure was 58 mmHg. No genetic tests were performed yet.

Because of left lung collapse, the patient had to undergo OLV during the general anesthesia. A systematic review showed that PV could protect against the development of acute lung injury in patients who underwent major surgery. So prophylactic lung-protective mechanical ventilation strategy was adopted. To the best of our knowledge, there has been no case with lung agenesis undergoing OLV and PV being reported.

To prevent hypoxemia during OLV, we improved lung function before operation. The standard five pronged attack was used for pre-operative lung function improvement. It included reducing irritant exposure including smoking, bronchodilators for airway dilatation, mucolytic agent administration, chest physiotherapy to remove secretions and antibiotics to treat suspicious infection. Before

operation the hemoglobin level was raised to 110 g/L by blood transfusion to improve oxygen supply. Double-lumen tube (DLT) was used for intubation, which was the standard method of lung isolation. Difficulty in selecting proper size is one of the disadvantages of DLT. With respect to the trachea 3D reconstruction, 35-Fr right-sided DLT was selected during anesthesia. Flexible fiberoptic bronchoscope was used to confirm the position of DLT. Once DLT was positioned at place, the adequacy of lung isolation was confirmed by ultrasonography. In the right lung, there was a to-and-fro movement at the pleural line that corresponds to tidal movement of the lung (lung sliding sign). The left lung showed the absence of lung sliding. Tidal volume was set to 6 mL/kg, corresponding to roughly 200 mL tidal volume for OLV. A positive end expiratory pressure (PEEP) of 5 cmH_2O was given to the patient. The dynamic lung compliance was 120 mL/ cmH_2O and the airway resistance was 12 cmH_2O . The pelvic lump was confirmed to be cervical carcinoma by histopathology. There were no postoperative pulmonary complications after surgery.

Discussion

Pulmonary agenesis, first reported by Brunner in 1963 is a rare congenital anomaly (1). Vitamin A deficiency, folic acid deficiency and salicylates were shown in experiments as the causes of pulmonary agenesis (2). The bilateral form is incompatible with life. In the absence of other malformations, unilateral pulmonary agenesis is compatible with normal life. The trachea continues directly into the main bronchus of the normally developed lung, and respiratory distress usually occurs due to retention of bronchial secretions and inflammations. Severe respiratory infection during infancy is common in these children and may lead to pneumonia and death before 5 years of age (3). Thus, patients usually present with cough, tachypnea, wheezing, respiratory distress, cyanosis or with chronic unproductive cough because of inflammations or bacterial colonization (4).

Its pathology has been categorized as per the classification by Schneider & Schwalbe: (I) agenesis-complete absence of a lung and bronchi, with no blood supply to the affected side; (II) aplasia-presence of a rudimentary bronchus with complete absence of parenchyma; and (III) hypoplasia-lobe agenesis and hypoplastic lung. Symptoms can manifest at birth as respiratory distress syndrome. Otherwise, patients may remain asymptomatic until adulthood when the defect is identified during routine examination for other purposes.

Unilateral right lung agenesis is frequently associated with other congenital abnormalities like cardiovascular anomalies, and its prognosis is worse than the left side agenesis. This difference can be explained by a greater mediastinal shift for the right lung agenesis, leading to tracheal compression. Left lung agenesis is more common, and it typically results in compensatory growth of the right lung and ensuing herniation into the contralateral hemithorax (5).

OLV is frequently used in both adult and pediatric patients due to increasing use of thoracoscopy and video-assisted thoracoscopic surgery. However, the disadvantage of OLV is its association with more manipulation and damage of airway, leading to significant physiological derangements such as ventilation perfusion mismatching and early development of hypoxia (6,7). Development of hypoxaemia (arterial oxygen saturation <90%) caused by OLV can be explained by three interacting factors: (I) poor oxygenation and compromised ventilation, resulting in reduced oxygen stores; (II) dissociation of oxygen from haemoglobin; and (III) ventilation-perfusion mismatch (8). Although lung isolation has long been used in clinical practice, it remains controversial due to lack of specific and clear guidelines with respect to the choice of tube size, method of isolation and insertion, confirmation of correct placement, optimum FiO₂ before and during OLV and the limits of acceptable degree of desaturation (9,10).

Mechanical ventilation is an essential supportive therapy to maintain gas exchange during general anesthesia, but inadequate ventilator settings can aggravate and even initiate lung damage in patients with healthy lungs at the onset of ventilation (11). Recent evidence showed that lung-protective mechanical ventilation using low tidal volume (6–8 mL/kg of predicted body weight), moderate PEEP (6–8 cmH₂O), and recruitment maneuver is associated with improved postoperative outcome in patients undergoing major surgery (12). However, there are no recommendations on optimal ventilator settings in patients without lung injury during general anesthesia (13). Although lung-PV may be beneficial for the lung, it may impair the cardiovascular system, reduce venous return and cardiac output and require the use of fluids and vasopressors. Thus, the risks and benefits of lung-PV need to be balanced in each individual patient.

Conclusions

In aggregate, we reported the first case of OLV and PV strategy in patient with pulmonary agenesis during general

anesthesia. The patient benefitted from PV without any postoperative pulmonary complications.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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