Postpneumonectomy-Like Syndrome in an Infant With Right Lung Agenesis and Left Main Bronchus Hypoplasia

Simone Furia, MD, Paolo Biban, MD,
Monica Benedetti, MD, Alberto Terzi, MD,
Massimo Soffiati, MD, and Francesco Calabrò, MD

Divisions of Thoracic Surgery and Pediatrics, Major City Hospital, Verona, and Thoracic Surgery, S. Croce Hospital, Cuneo, Italy

We report a 1-year-old child born with agenesis of the right lung who sustained an episode of acute respiratory failure related to a postpneumonectomy-like syndrome, with severe mediastinal shift and subsequent stretching and stenosis of the left main bronchus. The insertion of an expandable prosthesis in the right empty pleural space markedly improved the patient's clinical condition.

(Ann Thorac Surg 2009;87:e43–5)

Pulmonary agenesis is defined as total absence of the lung that is associated with the complete absence of the bronchial tree and vascular supply [1] and is observed in one of 10,000 to 15,000 autopsies. The clinical presentation of pulmonary agenesis is variable; symptoms may appear soon after delivery, or the anomaly can have a totally asymptomatic course and may be diagnosed during a routine examination [2, 3]. The true incidence of pulmonary agenesis is unknown, because 50% of cases are stillborn and more than 20% die at birth or during their first few months. Szarnicki and colleagues [2] reported that 30% of infants with agenesis die in their first year of life, and 50% die within the first 5 years.

Pulmonary agenesis can occur between the fourth and fifth week of gestation in the embryonic phase, before the pseudoglandular period, when primitive lung is forming as a diverticulum protruding from the foregut [1]. The etiology [4] may be related to a duplication of the distal part of the upper arm of chromosome 2 (46,XX, 2p+).

We report right pulmonary agenesis in an infant who sustained a severe episode of respiratory failure due to a mediastinal shift stretching the left main bronchus, similarly to what happens in patients with postpneumonectomy syndrome [2, 3, 5].

A full-term newborn (birth weight, 3110 g) born after vaginal delivery showed respiratory distress and stridor immediately after birth. A chest roentgenogram revealed a homogeneous opacity of the right hemithorax. He was diagnosed with polymalformative syndrome, including right lung agenesis, tracheal stenosis, labium palatoschisis, and atrial and ventricular septal defects.

During his first year, the patient underwent an operation for labiopalatoschisis. Complete spontaneous closure of heart defects was observed. Episodes of mild respiratory distress and stridor were initially treated by steroids, with a good response.

Because of a severe episode of respiratory failure with marked hypercapnia not responsive to therapy, the patient was admitted to the pediatric intensive care unit. Arterial blood gas analysis revealed a pH of 7.09, partial arterial pressure of oxygen was 117 mm Hg, partial arterial pressure of carbon dioxide was 81.5 mm Hg, and base excess was +1.9 mEq/L.

Mechanical ventilation was performed for 14 days. Bronchoscopic evaluation identified an obstruction narrowing the distal part of the trachea, with a diameter at this level of less than 2.5 mm. Airway stenosis was confirmed by computed tomography (CT) scan. The CT scan also showed a severe mediastinal displacement, with a complete shift of the heart to the right side of the chest and an atelectasis of the pulmonary lower lobe. The main stem bronchus was compressed between the left pulmonary artery and the aorta (Fig 1). To reduce the bronchial compression and maintain a proper diameter of the airways, a surgical procedure was planned to rotate the mediastinum to the correct position.

The patient underwent a right posterolateral thoracotomy for the insertion of a tissue expander (Laboratoires Eurosilicone, Apt, France) into the empty pleural space. The tissue expander consisted of a reservoir with a maximum volume of 250 mL and a port for refilling. A real pleural cavity was found, and the hemicylinder-shaped expander was placed in the empty hemithorax and was filled from a subcutaneous port under ultrasonographic control. A volume of 140 mL of saline solution was added, but the attempt to increase the volume up to 160 mL failed because of initial right atrium compression. On postoperative day 4, the tissue expander was filled to 200 mL, without any hemodynamic modification.

A CT scan showed the mediastinum was in the correct position and a marked reduction of the tracheal stenosis (Fig 2). However, postoperative bronchoscopy documented a tightening of the main stem bronchus due to congenital hypoplasia, which was hidden by the upstream tracheal compression. A tracheostomy was performed, and the patient was gradually weaned from mechanical ventilation.

At the 2-year follow-up, the child was in stable condition

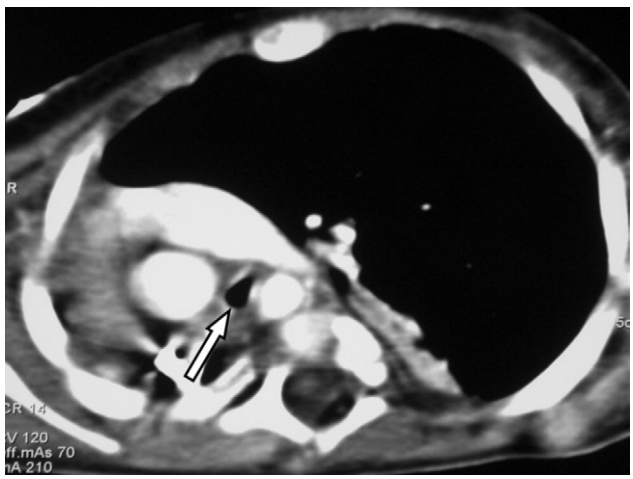

Fig 1. The hyperinflation of the left lung causes the main stem bronchus compression between left pulmonary artery and aorta (arrow).

Accepted for publication Feb 9, 2009.

Address correspondence to Dr Biban, U.O. Pediatria, Piazzale Stefani 1, Verona, 37126, Italy; e-mail: paolo.biban@azosp.vr.it.

Published by Elsevier Inc

0003-4975/09/$36.00
doi:10.1016/j.athoracsur.2009.02.023

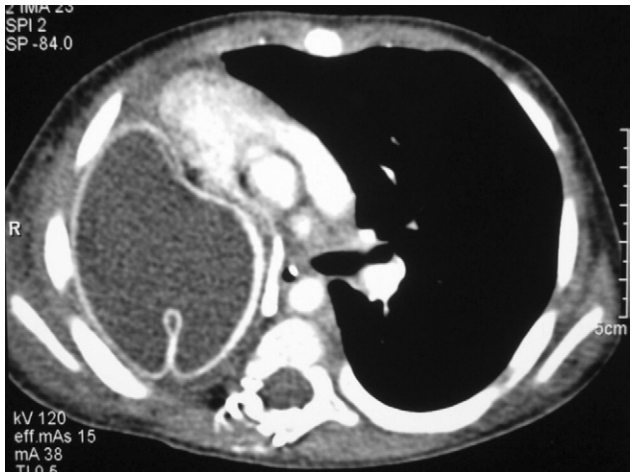

Fig 2. The insertion of the tissue expander stabilized the mediastinum and reduced the tracheal stenosis.

and breathing spontaneously through the tracheostomy. A chest roentgenogram at about 32 months after the operation showed the tissue expander had stabilized the mediastinum (Fig 3). The child is presently 4 years old, and his growth and neurologic development are within the normal limits for his age. Evaluations are currently underway to test the feasibility of decannulation.

Comment

Pediatric patients with unilateral pulmonary agenesis have an increased risk of morbidity and death. Right-sided lung agenesis has a higher mortality rate than left agenesis; this probably depends on the rotation of heart and mediastinum, with a greater displacement of the mediastinal vessels and airway and the higher incidence observed of associated cardiac and vascular anomalies [6].

Most patients with pulmonary agenesis present respiratory distress after birth, even though symptoms may be less severe and specific, including mild cyanosis, tachypnea, dyspnea, and stridor. Persistence of homogeneous opacity of hemithorax and ipsilateral shift of mediastinum on chest roentgenogram should suggest the diagnosis. Either CT scan or magnetic resonance imaging must be performed to define the correct anatomic connections between the mediastinal structures and to identify any significant vascular compression of major airways [6]. Nevertheless, a full bronchoscopic examination is required to exclude possible underdevelopment of the airway and to define any compression or stenosis of the tracheobronchial tree.

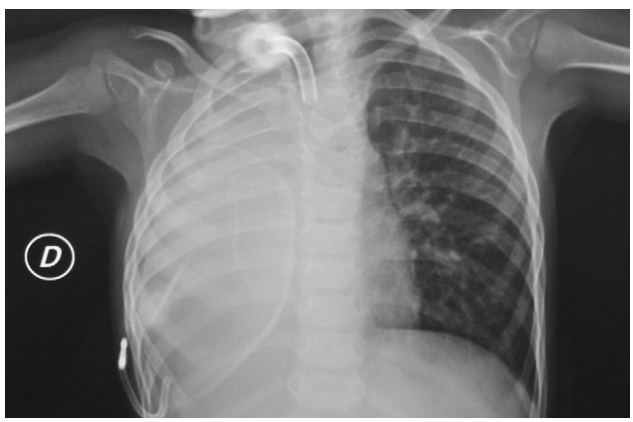

Fig 3. At 32 months after the operation, the tissue expander is correctly placed in the pleural cavity.

Postpneumonectomy syndrome is a severe complication of pulmonary agenesis that may become life-threatening. Surgical treatment includes the placement of a tissue expander, which has been shown to obtain a good outcome mainly in infants [5, 7, 8]. Indeed, the insertion of a tissue expander can limit left lung overdistension that may cause an excessive mediastinal shift; at the same time, the variable volume of the prothesis may be useful for adapting the size of the expander during growth. During the surgical insertion, it is crucial to monitor the patient's hemodynamic status, possibly by cardiac ultrasonographic scan, particularly while the prothesis is being filled, to avoid the compression of the right heart, which represents the main risk of the procedure [5, 7, 8].

The management of pulmonary agenesia includes the thorough investigation of the presence of other anatomic anomalies associated with the main defect: pulmonary agenesis can be just one aspect of a polymalformative syndrome that may involve the cardiovascular, gastrointestinal, and musculoskeletal systems [1].

In fact, in our patient, after the surgical procedure, we unexpectedly found a hypoplasia of the single left main bronchus that appeared as a tightening of the bronchial lumen. Notably, this anomaly had been hidden by the compression and was only revealed after the operation, presumably being very difficult to detect in the preoperative phase.

In summary, postpneumonectomy syndrome is a rare but life-threatening complication of pulmonary agenesis. Management requires a multidisciplinary and coordinated approach that includes a stabilization phase in the intensive care unit and a tailored surgical treatment by means of insertion of a tissue expander. Associated airway anomalies must be carefully scrutinized before and after the operation.

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