Case Report: Agenesis of Right Pulmonary Artery

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A young woman with absence of the right pulmonary artery was presented. The diagnosis was suspected on a chest x-ray and confirmed with a lung scan.

The development of the human arterial system occurs during embryonic life. Early in the process of formation of the arterial system, six aortic arches from which the pulmonary, common, external, internal carotid, as well as other arteries arising at different times, are present. The branches of the pulmonary artery are derived from the proximal portions of the sixth aortic arch. The distal portion of the right arch disappears while the distal portion of the left forms the ductus arteriosus. Disappearance of the proximal portion of either the right or the left sixth arch results in absence of a main branch of the pulmonary artery.

The vascular system of the lungs does not depend on the sixth aortic arch. Early in its development, the pulmonary vascular system is connected to ventral branches of the dorsal aorta. These communications disappear at a later stage and the lungs are then supplied by developing branches of the right and left pulmonary artery (*1*).

Case Report

The patient is a 23-year-old black woman in good health until March 1974 when she was admitted with fever, chills, and cough of three-days duration. Her medical and family histories were unremarkable. Her blood pressure was 90/50 mm Hg; pulse—95/min and regular; temperature—103° F. Examination of the chest showed some tubular breathing on the left side, but no rales. The heart rhythm was regular with a short apical systolic murmur. The first and second sounds were of normal quality and there was no gallop. The abdomen was soft. A chest x-ray demonstrated an infiltrate in the left lower lobe. The trachea, mediastinum, and heart were moderately shifted to the right. The right hemithorax was smaller than the left. Her EKG and laboratory studies were within normal limits. It was concluded that the patient had a viral pneumonia. After treatment, she rapidly improved and was discharged.

Until May 1977, when she was seen in the medical clinic because of sore throat and dry cough, the patient had been asymptomatic. The physical examination was unremarkable. A repeated chest x-ray (Fig. 1) revealed the heart and mediastinum somewhat shifted towards the right. The left hemithorax appeared larger than the right. The right lung vessels appeared small; however, the possibility that they were bronchial shadows rather than pulmonary vessels was raised. A perfusion-ventilation lung scan was performed. Immediately after injection of 4 mCi of Tc-99m aggregated albumin, serial views were obtained. They showed visualization of the heart, left pulmonary artery, and left lung. The right pulmonary artery was not seen and there was no flow to the right lung (Fig. 2). The static images showed the perfusion of the left

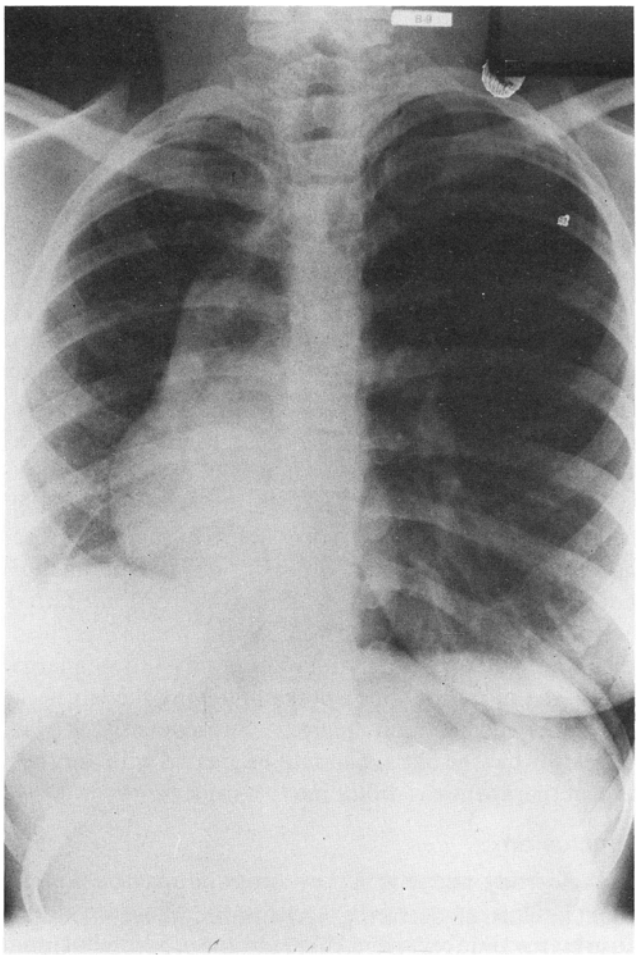

FIG. 1. P.A. roentgenogram of the chest demonstrates shift of the heart and mediastinum to the right. The right hemithorax is smaller than the left; the right hemidiaphragm is raised. Vascular shadows are less prominent than those on the left.

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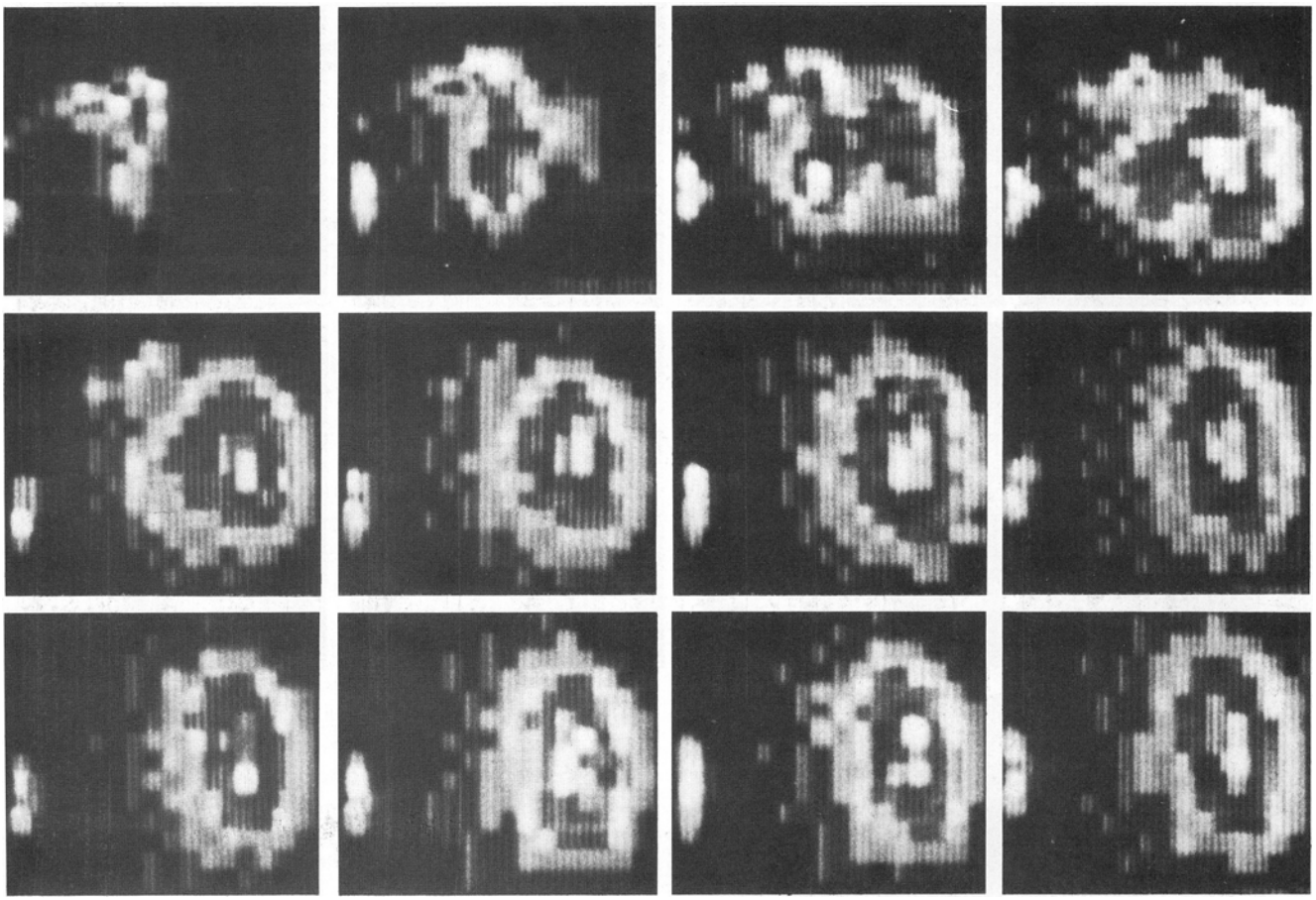

FIG. 2. Serial dynamic anterior views at 5-sec intervals following the administration of 4 mCi of Tc-99m aggregated albumin. Note the visualization of superior vena cava, right atrium, right ventricle, left pulmonary artery, and left lung. There is no visualization of the right pulmonary artery and right lung; the spot on the left side corresponds to the injection site.

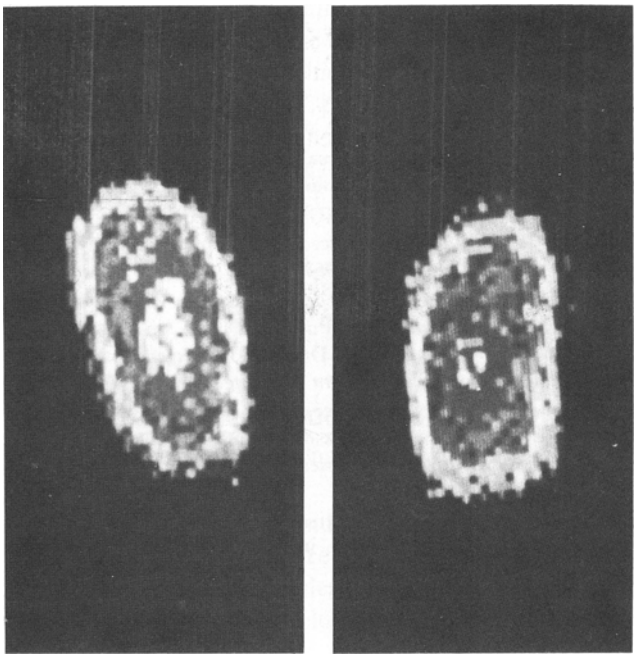

FIG. 3. Static images of the perfusion lung scan—anterior (left) and posterior (right) views. Note uniform perfusion of the left lung; the right lung is not seen.

lung (Fig. 3). A ventilation scan revealed aeration of both lungs, but the right lung was smaller than the left (Fig. 4). Because of the lung scan findings, no angiography was performed. It was concluded that the patient has an absence of the right pulmonary artery.

Discussion

Agenesis of one of the branches of the pulmonary artery is an uncommon anomaly. Most individuals with this anomaly die in infancy, but it is not necessarily fatal in early life.

The right pulmonary artery is absent more often than the left. Absence of the left pulmonary artery is associated in many cases with other congenital malformations. The left lung usually receives its blood supply through one or more expanded bronchial artery or additional accessory arteries (*2*).

Absence of the right pulmonary artery is usually not associated with other malformations. The affected lung is generally supplied through an artery that arises from the ascending aorta. Because the lung is receiving systemic and abnormal blood supply, it often becomes hypoplastic and shows bronchiectasis or cystic changes (*2*).

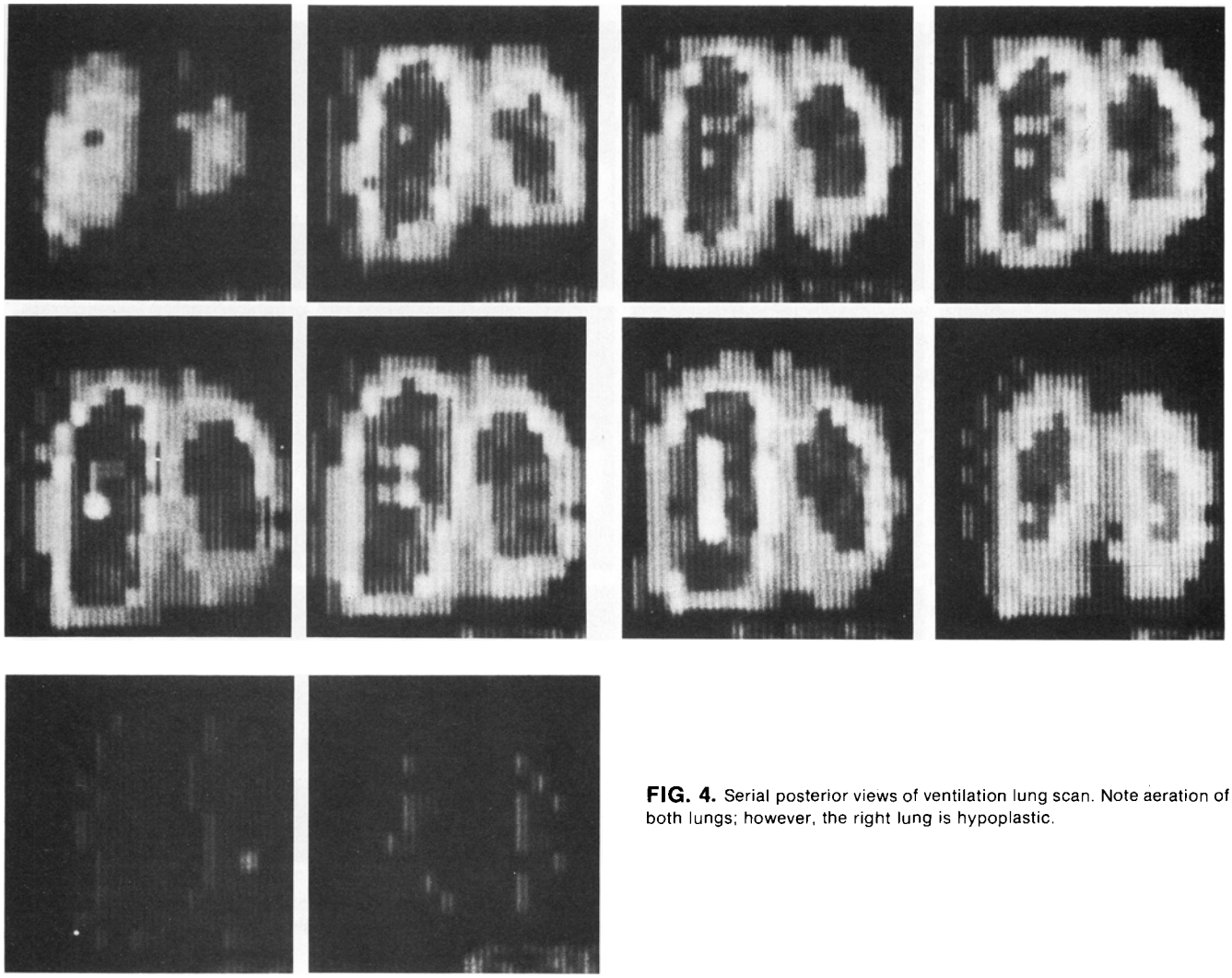

FIG. 4. Serial posterior views of ventilation lung scan. Note aeration of both lungs; however, the right lung is hypoplastic.

Pool et al. (*3*) collected 98 cases demonstrating the absence of one of the branches of the pulmonary artery in 1962. Most of the cases with absence of the left pulmonary artery had Fallot's tetralogy.

The chest x-ray findings are characteristic enough to make this diagnosis or strongly suspect it in most cases. The involved hemithorax is smaller than the normal side. The mediastinal structures are shifted to the affected side; the normal shadows of the pulmonary arterial branches in the hilum and in the lung are absent (*4*).

It has been postulated that establishing defintive diagnosis of absence of a main branch of the pulmonary artery depends on angiocardiography (*4–7*). Our case, however, suggests that the use of a noninvasive technique such as the dynamic lung scan permits a definitive diagnosis—without using a more risky diagnostic procedure such as the angiogram.

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