JOHN M. SMITH, M.D.*
Lexington, Kentucky

This nine-year-old white girl has suffered from repeated respiratory infections and chronic cough for many years. Physical examination is unremarkable except for rales in the left base. Routine laboratory studies are within normal limits.

*From the Department of Radiology, University of Kentucky Medical Center.

Figure 1
Answer: Agenesis of the Left Main Pulmonary Artery

The posteroanterior chest radiograph (Fig. 1) shows the left hemithorax to be smaller than the right. The heart and mediastinum are displaced toward the left. Bronchography was negative and bronchoscopy revealed no abnormality other than minor chronic inflammatory changes in the left lower bronchus.

Angiograms were obtained after injection of contrast material into the outlet of the right ventricle (RV—Fig. 2A). Figure 2A demonstrates contrast filling of the right pulmonary artery (RPA) and absence of a left pulmonary artery. Figure 2B shows the return flow to the left heart and a right aortic arch (RAA). The source of blood flow to the left lung is not clearly defined. The left atrial appendage is visualized just inferior to the catheter tip in Fig. 2B.

Congenital unilateral absence of a pulmonary artery is a rare condition.1 With a normally positioned heart, absence of the right pulmonary artery occurs more often than absence of the left, and usually there is no associated congenital heart disease. Agenesis of the left pulmonary artery is most uncommon as an isolated anomaly. It is usually present in conjunction with tetralogy of Fallot,2 but atrial and ventricular septal defect, and less often patent ductus arteriosus,3 have also been reported as associated lesions. Some authors report that agenesis of the left pulmonary artery is almost always accompanied by a right aortic arch.4 Small branches from the aorta supply the left lung. Possible embryologic explanations for this difference in frequency of congenital cardiac anomalies associated with absence of the right and left pulmonary artery have been presented by several authors.4

The most striking radiologic manifestations of agenesis of a pulmonary artery are reduction in size of the affected hemithorax and shifting of the heart and mediastinal structures toward the involved side. No definite pulmonary artery shadow is visible on the affected side. Fluoroscopy shows no shift of the heart or mediastinum on inspiration, indicating there is no obstruction to aeration of the affected lung. The definite diagnosis is made by angiography of the pulmonary arteries.

References

For reprints, please write: Dr. Smith, 3419 Brookhaven Drive, Lexington, Kentucky.