A 40-year-old man with the acquired immunodeficiency syndrome (AIDS) was admitted for evaluation of dyspnea, nonproductive cough, and fever of five days' duration. Six months earlier, he had been treated at another hospital for Pneumocystis carinii pneumonia and was subsequently begun on therapy with 3'-azido-3'-deoxythymidine (AZT). His past medical history was remarkable for the presence of a right-sided aortic arch, which had been noted on chest radiographs for many years. The patient reported that this abnormality was also present in his father. His prior pulmonary history was remarkable for an episode of dyspnea and cyanosis during a trip to Quito, Ecuador, at age 17, which was diagnosed as high-altitude pulmonary edema. At age 28, a similar episode occurred with the development of dyspnea and angina during a trip to Aspen, Colorado. With the exception of these episodes, he was without respiratory complaints. There was no history of asthma, bronchitis, tuberculosis, or cigarette smoking.

On physical examination, he was tachypneic with a respiratory rate of 24 breaths per minute and a temperature of 38°C. His neck veins were not distended. Result of chest examination was normal except for a few scattered rales. Cardiac examination revealed a S4 gallop and an increased pulmonic component of the second heart sound. Arterial blood gas determination revealed acute respiratory alkalosis with moderate hypoxemia that worsened with exercise. Diffusing capacity was 50 percent of predicted. Chest radiographs three months prior to admission (Fig 1) and at the time of admission (Fig 2) are shown. The development of new diffuse, bilateral interstitial infiltrates on the admission chest radiograph was suggestive of Pneumocystis carinii pneumonia (PCP), given his underlying diagnosis of AIDS. Therapy for PCP was instituted and this diagnosis was confirmed by bronchoalveolar lavage. A further study was performed to evaluate his pre-existing radiographic abnormality.
Diagnosis: Proximal interruption of the left pulmonary artery (unilateral absence of the left pulmonary artery)

A pulmonary perfusion scan (Fig 3) revealed absent perfusion to the left lung consistent with the above diagnosis. This uncommon congenital abnormality has also been termed unilateral absence of a pulmonary artery, but proximal interruption is a more accurate description since the distal pulmonary vasculature is usually intact, although decreased in size. The involved lung is hypoplastic and receives its blood supply from the bronchial circulation, as well as via collaterals from intrathoracic and subdiaphragmatic arterial sources. Several radiographic features allow a presumptive diagnosis to be made. These include: (1) a small ipsilateral thorax with narrowed intercostal spaces and an elevated hemi-diaphragm; (2) displacement of the heart and mediastinum to the affected side, without shift during respiratory maneuvers; (3) absence of a pulmonary artery shadow; and (4) reticular densities in the ipsilateral perihilar region representing the hypertrophied bronchial circulation. Intercostal and subdiaphragmatic collaterals may produce rib notching and pleural telangiectasias, manifested as hazy pulmonary densities and pleural thickening. The interrupted pulmonary artery most often occurs on the right side, since it is usually on the side opposite the aortic arch. In our patient with a right-sided aortic arch, it occurred on the left.

Patients with proximal interruption of a pulmonary artery (PIPA) are usually asymptomatic unless pulmonary hypertension or associated cardiovascular abnormalities are present. Atrial and ventricular septal defects, patent ductus arteriosus and tetralogy of Fallot have all been associated with PIPA. Pulmonary hypertension occurs in 88 percent of cases with an associated cardiovascular shunt, and in 19 percent of isolated cases.

Individuals with PIPA have been reported to be susceptible to high altitude pulmonary edema (HAPE). Because life-threatening problems have occurred even at moderate altitudes, these individuals should be advised not to travel higher than 2,000 meters. Although HAPE is a form of noncardiogenic pulmonary edema, its exact mechanism is unclear. It had been thought to represent in part a vascular leak caused by overperfusion of the pulmonary vascular bed. Analysis of HAPE bronchoalveolar lavage fluid, however, has revealed markedly increased concentrations of high molecular weight proteins and a high ratio of edema fluid protein to serum protein suggesting a permeability type of edema. The cause of injury to the endothelial-epithelial barrier is unclear, but may involve the effect of hypoxia, high pulmonary pressure and high linear velocity of blood flow. Since individuals with PIPA have increased blood flow to the one lung with an intact pulmonary artery and in some cases frank pulmonary hypertension, they may be especially predisposed to the development of hypoxia-related pulmonary vascular damage.

In our patient, the diagnosis of PIPA was established by the characteristic chest radiographs and perfusion scan. The possibility that a thrombotic or embolic event occurred early in life and resulted in the same abnormality cannot be excluded.

It is important to recognize the presence of PIPA so that these individuals can be advised of the risk of HAPE. If a patient with PIPA requires bronchoscopic examination, we feel that it would not be advisable to perform a transbronchial biopsy. Since the hypoplastic lung is perfused by collaterals from the systemic circulation and the contralateral lung may have pulmonary hypertension, significant bleeding may follow a biopsy of either lung secondary to the presence of elevated vascular pressures.

REFERENCES