A 20-year-old man complained of occasional breathlessness on exertion and cough productive of scanty mucoid expectoration. No other notable facts could be obtained from the clinical history except for one episode of bronchopneumonia during infancy. The first in the family of three children, he was born to nonconsanguineous parents. The delivery had been uncomplicated, and the family history was negative for congenital malformations.

Physical findings in this well-built individual included a leftward shift of the trachea and the cardiac impulse. Breath sounds were audible over the left hemithorax, though diminished in intensity in the lower zones. Fine inspiratory crackles with occasional rhonchi were heard at the left base. Other systems did not reveal any abnormalities.

A posteroanterior chest roentgenogram (Fig 1) demonstrated marked displacement of the trachea and the heart to the left. The heart borders were indistinct, and the anterior mediastinal septum was deviated leftward with herniation of the contralateral lung to the left. Aeration was evident in the left upper zone. Radiographs of the spine, ECG, and intravenous urographic findings were normal. The vital capacity, FEV₁, and peak expiratory flow rate showed a mild decrease relative to the predicted values. A diagnostic procedure was performed.

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Diagnosis: Agenesis of the left lung, with anterior rotation of the right anterior segment to the left side

The patient underwent fiberoptic bronchoscopy, which demonstrated absence of the left main bronchial orifice. The suspicion of agenesis of the left lung was confirmed by bronchography (Fig 2) and pulmonary angiography (Fig 3).

Agenesis of the lung is a rare congenital anomaly representing failure of development of the lung bud. Unilateral agenesis of the lung has been reported to occur in one in 10,000 persons.1 Three degrees of developmental deficiencies of the lung have been reported—agenesis, aplasia, and hypoplasia.2 Both sexes are equally affected. There is no predilection for either side,3 although Weir4 reports that it occurs twice as often on the left side. Agenesis of the lung is often associated with major extrapulmonary developmental abnormalities, particularly those involving the heart, kidneys, and diaphragm. Although the exact etiopathogenesis is not clear, a genetic basis, vitamin A deprivation, and unrecognized abnormalities in CNS control of fetal respiratory movements have been suggested.1

Plain chest radiographic findings of unilateral agenesis include an opaque, grossly contracted hemithorax with ipsilateral mediastinal shift. The contralateral lung, which often undergoes extreme degrees of overinflation, may occupy a portion of the affected side, along with the deviated anterior mediastinal septum. Indeed, the presence of this configuration caused some confusion in the plain film interpretation in our case. The differential diagnosis in such a situation includes atelectasis of the entire lung, far advanced fibrothorax, and severe bronchiectasis with collapse.1 In distinguishing between congenital and acquired forms of minithorax, Daves and Walsh5 observed that although the first five ribs show crowding in both situations, symmetrical lower anterior ribs favor a congenital anomaly, while crowding and downward displacement of the lower anterior ribs on the affected side indicate an acquired etiology.

Bronchography demonstrates absence of the bronchus on the affected side. A definitive diagnosis is possible with pulmonary angiography, which may also aid in identifying other associated cardiac anomalies.6

The clinical symptoms and signs are governed by the associated congenital malformations. Except for one episode of bronchopneumonia during infancy, our patient did not have significant respiratory complaints. Isolated lung agenesis may be a chance finding on radiographs obtained for some other purpose, and affected patients may often be asymptomatic. However, the clinical course may be punctuated by recurrent lower respiratory tract infections.

The prognosis in cases of agenesis depends on the functional integrity of the remaining lung as well as the presence of associated anomalies. We believe that in the absence of such anomalies our patient will have a normal life span.

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Roentgenogram of the Month (Araja, Seetharaman)