Agenesis of the Lung

Case Report With Five Year Follow-Up

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Agenesis of the lung is a congenital absence or hypoplasia of one or both lungs. This is not a common condition. As of January 1, 1954 there were less than 150 cases reported in the medical literature.\(^1\) Most of these were discovered at autopsy.

In 1950 Ingram\(^2\) reported that only 18 cases had been diagnosed during life up to that time.

In 1953 Oyamada, Gasul and Holinger\(^3\) reviewed the literature to date listing 73 cases of agenesis, 38 cases of hypoplasia, 4 cases of newborn infants with congenital absence of both lungs and 21 cases with agenesis or congenital hypoplasia of the lung but with the description being incomplete or the diagnosis not being definitely established.

Schneider\(^4\) has described three types of pulmonary agenesis depending on the development of the bronchus and lung tissue: (1) One lung is absent without any development of the lung or bronchus, (2) there is complete absence of lung tissue but there is a poorly developed bronchus on the involved side and (3) the bronchus is small, poorly developed and connected to a mass of rudimentary lung tissue.

The etiology of this faulty lung development is unknown. The condition is often associated with other anomalies of the tracheobronchial tree and even other parts of the body. Cases coming to autopsy have uniformly shown emphysema of the remaining lung, malformation of the cartilagenous skeleton of the trachea such as extra cartilagenous rings or narrowing of trachea and absence or malformation of pulmonary vessels.\(^5\) Hypertrophy of the right heart has not been noted. There is little difference in incidence between the right and left side. About 14 per cent of the cases are stillborn or die within the first few weeks of life.\(^6\)

Symptoms: The patient may be completely asymptomatic. He may have mild respiratory distress on exertion. A few cases have dyspnea, cyanosis, recurring choking spells, wheezing, stridor and hemoptysis on occasions. The amount of respiratory distress is dependent upon the condition of the remaining lung and the associated congenital anomalies. Respiratory infections are common. The infant is susceptible to attacks of bronchopneumonia and later the child is susceptible to frequent colds and dyspnea on exertion.

Diagnosis: The condition can usually be distinguished from atelectasis of the newborn by absence of respiratory distress. The chest is usually symmetrical in the infant but as the child grows older there may be slight asymmetry. There is mediastinal shift to the involved side. The
remaining lung may extend past the midline. Bronchoscopy reveals the absence or deformity of bronchi to the effected side. Bronchograms confirm this deformity. Angiocardiograms reveal absence or rudimentary blood supply to the involved side. Prognosis: The prognosis in any case is influenced more by the associated anomalies than by the agenesis of one lung. Many die in infancy but some live a normal life span. Twenty-three persons (27 per cent) of the series reviewed by Hochberg and Naclerio were known to have lived to be between 15 and 72 years of age.

The involvement of the only existing lung by severe infection, foreign body or trauma may prove fatal at any time. Due to the fact that there may be unknown associated congenital anomalies these individuals are not always good surgical risks. This should be borne in mind when submitting them to elective surgery.

Case Report

E. I. K. white, girl, age 6½ months, was first admitted to the Mississippi Baptist Hospital on May 25, 1951. The mother stated that she was an 8 month baby, weighing 5 lbs. 11 oz. at birth. She was fed from birth by formula and ate well. She was never cyanotic and only slight dyspnea was noted on exertion at any time. This was not marked. She did not seem unusual except in failure to gain weight as rapidly as the average child. About three weeks before admission, she developed what was thought to be a cold. Dr. Swink Hicks of Natchez, Mississippi, was consulted and referred her for further examination.

This revealed an alert, apparently healthy, well nourished white girl, weighing 12 lbs. The chest was symmetrical but there was impaired resonance to flatness over the right hemithorax. There was no cardiac dullness on the left. Heart sounds were best heard on the right. Breath sounds were absent over the right hemithorax. X-ray films made before admission with barium swallow revealed no evidence of diaphragmatic

FIGURE 2: Photograph of patient, age 5 years, with agenesis of right lung. Note slight asymmetry of the chest with prominence of right supraclavicular and infraclavicular fossae. The right shoulder is lower than left.
hernia. The stomach and intestine were in normal position. Roentgenogram of chest, Fig. 1, taken May 26, 1951, revealed homogenous density in right hemithorax with shift of mediastinal structures to that side. The left lung was clear. The chest cage appeared symmetrical. The intercostal spaces were found to be equal on the two sides.

A blood examination revealed a red blood cell count of 13 grams, white blood cell count 10,100 with 63 per cent lymphocytes, 16 per cent segments, 15 per cent monocytes and 6 per cent eosinophils.

On May 26, a 3½ mm. bronchoscope was passed. The trachea was deviated to the right and it appeared to be of normal size. The carina was much wider than normal and was a little red. The right main stem bronchus took off at more of an angle than usual and ended abruptly at about 1 cm. beyond the bifurcation of the trachea. At the end it appeared to be covered with normal mucous membrane. There was no evidence of intraluminal tumor or foreign body. The blind bronchial stump was probed gently with a suction tip. An unsuccessful attempt was made to obtain a biopsy. She did well after this examination did not develop evidence of laryngeal obstructions and was discharged about 36 hours later.

A diagnosis of agenesis of the right lung was made. Bronchography and angiocardiography did not appear justified at the time.

During the last five years her general health has remained good except for frequent colds. She has not gained weight as rapidly as the average child. When last examined in February, 1956, she weighed 32½ lbs. and her height was 45 inches. There was

FIGURE 3: X-ray film of chest November 18, 1955. Mediastinal structures in right hemithorax, some narrowing of intercostal spaces on right and right shoulder dropped.
slight asymmetry of her chest, Fig. 2. The right shoulder is carried at a lower level than the left.

X-ray film of chest on November 18, 1955, Fig. 3, revealed normal appearing left lung with mediastinal structures shifted to the right. There is slight narrowing of the intercostal spaces on the right.

She is extremely active and does not experience dyspnea even on extreme exertion. We have not been able to detect other congenital anomalies. Apparently her prognosis is good.

REFERENCES


