

AGENESIS OF THE LUNG WITH PERSISTENT DUCTUS ARTERIOSUS

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This single case is recorded on account of the rarity of the association between agenesis of the lung and persistence of the ductus arteriosus, especially with survival into childhood.

CASE REPORT

A girl, aged 7 years, was referred to the Cardiosurgical Unit on January 17, 1956, complaining of listlessness and shortness of breath when playing with her young friends. She had been otherwise healthy apart from tonsillitis, for which tonsillectomy had been performed at the age of 3 years.

On examination, she was a frail, undernourished (height 47 in. (119.4 cm.), weight 42 lb. (19 kg.)), but active child. Although there was slight flattening of the left side of the chest, the deformity was not so

great as to suggest the absence of a lung. There was equal movement on both sides of the chest and no deviation of the trachea. The apex beat was not palpable. The cardiac dullness was absent in front, where the air entry was good. At the back, the breath sounds were generally diminished. Auscultation of the heart revealed a continuous murmur, loudest in the second left intercostal space in front, and at the same level at the back.

Fluoroscopy revealed an apparently normal lung field on the right, but on the left there were lung markings in the upper zone with the cardiac silhouette below (Fig. 1a).

Bronchography (Fig. 1b) showed the trachea to continue directly into the right lung, the bifurcation being represented merely by a small blind diverticulum to the left. The middle lobe bronchus coursed to the

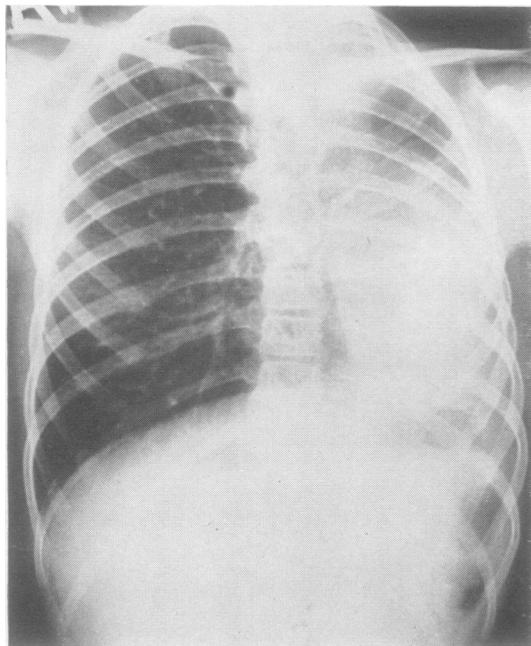


FIG. 1a

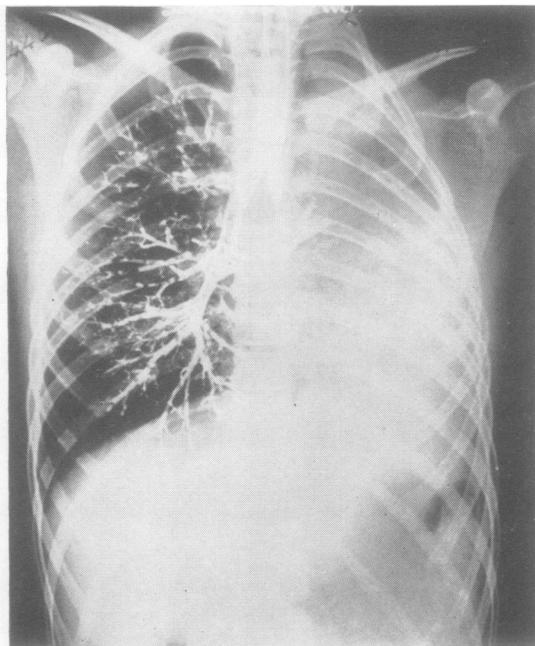


FIG. 1b

FIG. 1.—(a) Plain chest x-ray. The translucency in the left upper chest has been shown to be a mediastinal hernia containing the middle lobe. (b) Bronchogram.

left side, demonstrating an anterior mediastinal hernia. At bronchoscopy the trachea was normal, the distorted carina marking the origin of a left main bronchus which was about half the diameter of the right and lined by healthy mucosa. This appeared to terminate 1 in. from its origin when probed by a thin polythene tube, an impression confirmed by injection of iodized oil to outline the part not visible through the bronchoscope.

The lung function tests showed a vital capacity of 1,050 ml. and a maximum breathing capacity of 24 litres per minute, the respiratory wave form being normal.

At operation on February 8, 1956, it was found that the left pleural cavity was absent. The pericardium presented immediately under the chest wall, separated only by fatty tissue and by the thymus gland overlying the arch of the aorta (Fig. 2). There was no left lung, its place being taken by the right lung and pleura, which ballooned across the midline in front, filling the upper part of the chest. A coarse thrill was palpated under the aortic arch and localized the ductus arteriosus, which was exposed after division of a large, unusually placed superior intercostal vein running downwards. On opening the pericardium a large persistent ductus, 0.75 cm. in diameter, was seen to link the main pulmonary artery and the aorta. There was no left pulmonary artery, and, indeed, no left lung could be found.

On histological section, the tissue overlying the hilar region was found to be normal thymus gland. The ductus, which permitted a large flow into the single pulmonary artery, was isolated inside the pericardium, divided between clamps, the ends sutured, and a flap of pericardium interposed. When seen four months

later, the patient had been transformed into a vigorous child of small stature but normal nutrition.

DISCUSSION

Agenesis of the lung is rare, and no record has been found of a case with a large persistent ductus arteriosus surviving beyond infancy and submitted to operation. The high mortality (33%) within the first year of life in the collected series has usually been due to the diverse associated abnormalities, many of a gross nature, present in 50% of cases (Smart, 1946; Wexels, 1951). A few cases complicated by persistent ductus arteriosus and terminating in death during infancy have been reported. One survival to adult life has been recorded by Lukas, Dotter, and Steinberg (1953). This patient, a 22-year-old man with lifelong dyspnoea, was shown by cardiac catheterization to have a persistent ductus complicated by pulmonary hypertension (113/70 mm. mercury) with reversal of flow, cyanosis of the extremities, and clubbing of the toes.

Apart from shortness of breath on exertion, there were no respiratory symptoms in the case reported here. Wheezing, uncomplicated by bronchopulmonary disease, has been described in association with agenesis of a lung and considered to be due, in some cases, in compression of the bronchus between an abnormal pulmonary artery, passing behind it, and the aorta (Maier and Gould, 1953). As in the present case, when the diagnosis is in doubt, agenesis of the lung can usually be differentiated from atelectasis and fibrosis beyond a partial stricture by bronchoscopic examination and the injection of iodized oil through a small polythene tube. The normal pulmonary function tests suggest that the solitary lung had hypertrophied so as to occupy the space of both. Provided serious respiratory disease is avoided, the prognosis should be better than in most cases after pneumonectomy now that the persistent ductus has been divided.

SUMMARY

Division of a patent ductus arteriosus in a child 7 years old with agenesis of the left lung is briefly recorded.

I wish to thank the members of the Cardiosurgical Unit for permission to publish this case, Dr. J. Hinds and Mr. Litherland for the outline drawing and the photographs respectively, and Miss Tibbles, of the Central Medical Library.

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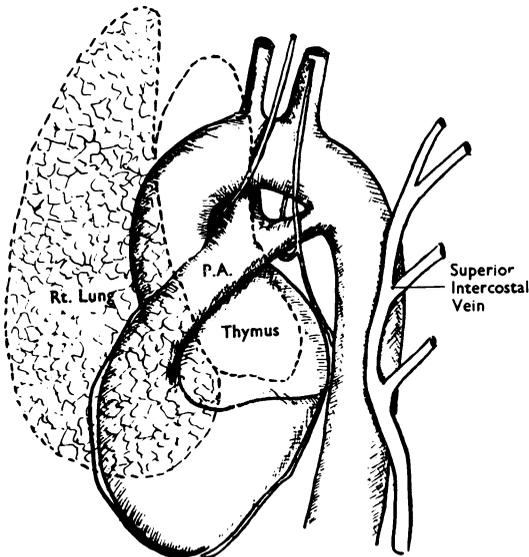


FIG. 2.—Outline drawing of the anatomy which presented at thoracotomy, there being no left lung or pleural cavity.