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Unilateral Pulmonary Agenesis-Presentation of Two New Cases*

Olcay Oran, M.D. / Melda Çağlar, M.D.*** / Gülsev Kale, M.D.**** / Güler Kanra, M.D.****

Until today, descriptions of about 200 cases of this condition have been published.[1] Schneider first classified the condition according to the extend of the defect in 1912.[1]

Group I - Complete absence of the lung and bronchus

Group II - Beginning of the bronchus is blind pit or pouch

Group III - Bronchus ending in a fleshy structure

In 1955 Boyden proposed a new more practical classification based on the degree of developmental arrest.[2]

Group I - Complete absence of one or both lungs (agenesis)

Group II - Suppression of all but rudimentary bronchus (aplasia)

Group III - Abortive growth (hypoplasia).[1]

In the pathogenesis of pulmonary agenesis embryonal developmental errors are strongly considered, but genetic, teratogenic and mechanical factors cannot be excluded either.

Koymen reported the first case of pulmonary agenesis from Turkey in 1955,[3] however, the investigations of the case were not complete, first fully investigated case was reported by Dr. Aksugur in 1961,[4] since then 11 more cases have been reported in Turkey.[5]

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* From Children's Medical Center, Division of Neonatology, Hacettepe University, Ankara.

** Associate Professor of Pediatrics.

*** Professor of Pediatrics, Pathologist.

**** Associate in Pediatrics.

Case Report

Case 1: (H. Ç. H. 857641). A 10-day old male infant was brought to Hacettepe Children's Hospital because of dyspnea and cyanosis. The baby was born after a normal course of labor, the mother, fertigravida, was 24 years old. At birth the baby was cyanotic and cyanosis increased gradually. The infant's siblings were healthy.

Physical examination: Temperature 35.5 C, pulse 120/min, respiration 48/min, height 57 cm, weight 3.8 kg. Femoral arterial pulsations were palpable bilaterally. The patient appeared deeply cyanotic, dyspneic, intercostal, subcostal, subrasternal retractions were seen, on auscultation of hemithoraces, bilateral crepitant rales were heard, heart sounds were stronger on the right side.

On chest X-ray there was no visible shadow for right main bronchus and right bronchovascular. (Figure 1) ECG showed clockwise rotation, no dextrocardia and no ventricular hypertrophy were observed.

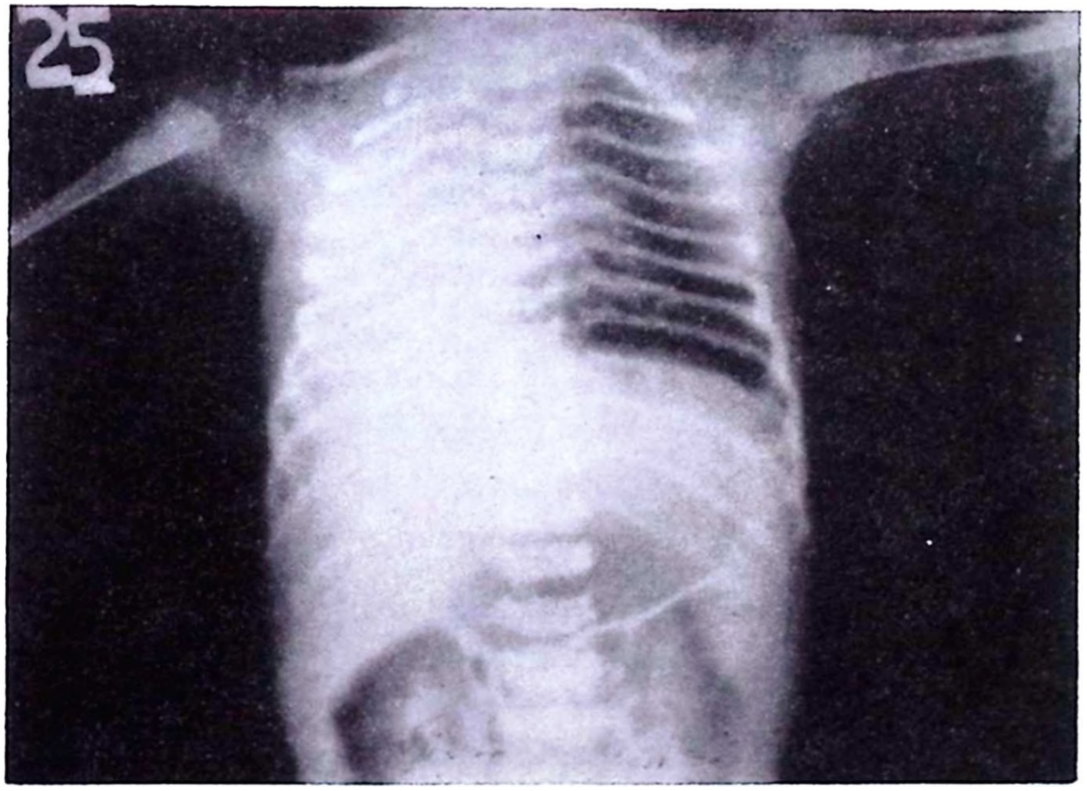

Figure 1
Case 1 Chest X-ray

The infant was placed into the incubator and given oxygen and 5 % Dextrose-1500 cc/m^2 I. V. and crystalline penicillin intramuscularly 100.000 u/kg in 2 doses. The Baby's condition deteriorated and he arrested, 1 1/2 hours after the admission resuscitation was unsuccesful.

At autopsy absence of right pulmonary artery and lung tissue were noticed, there was stenosis at the level of bifurcation and the left lung was monolobular. (Figure 2)

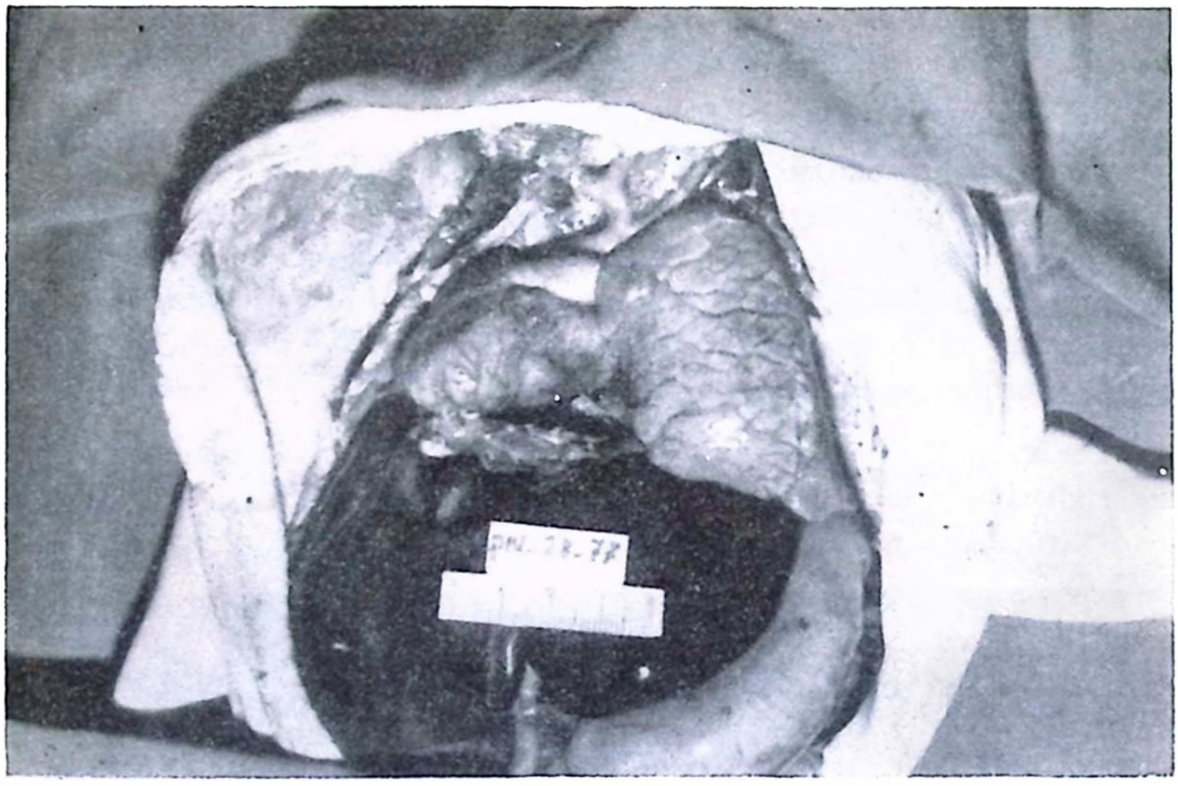

Figure 2
Case 1 Autopsy findings

Case 2: (H. C. H. 893552) A 19 day old male infant was admitted to Hacettepe Children's Hospital because of fever, abdominal distension and roaring respirations.

The baby was born at term pregnancy with vaginal delivery. Mother was 34 years old and this was the 5th pregnancy. The parents were cousins. One sibling had died at the age of 5 years during operation for congenital heart disease. The other 3 siblings were healthy.

Physical examination: Temperature 37.5 C, pulse 160/min, respiration 40/min, height 37 cm, weight 3.2 kg. Femoral pulsations were palpable bilaterally. The baby was in apparent respiratory distress, there was intercostal and suprasternal retractions. The respiratory sounds were reduced on the right hemithorax and bilateral ronflan rales were noted. Heart sounds were normal, no murmur was audible.

Chest X-ray showed no bronchopulmonary or vascular markings on the right hemithorax. (Figure 3) ECG showed no abnormalities.

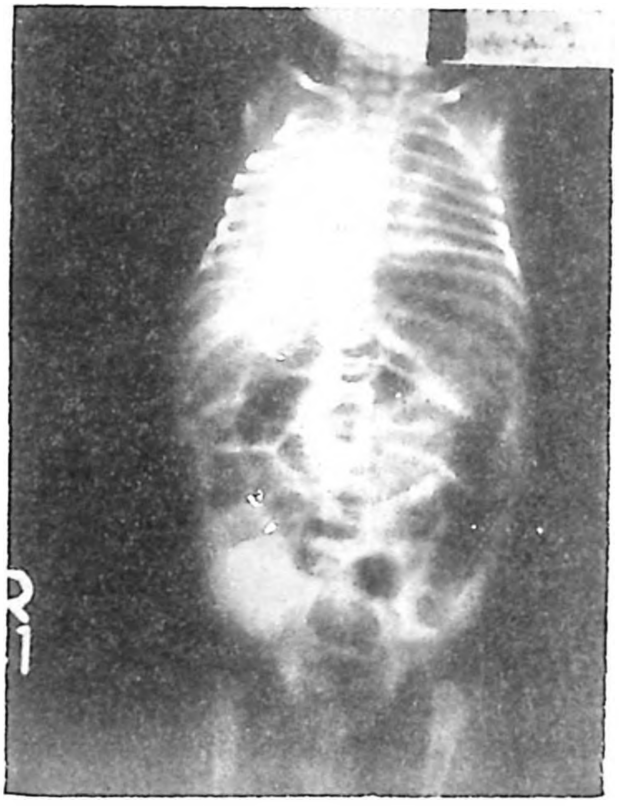

Figure 3
Case 2 Chest X-ray

Crystalline penicilline and kanamycin sulfate were given intramuscularly and in 4 days the condition of the baby improved and he was discharged from the hospital in good condition with advice to return for control after 1 month, however the baby expired meanwhile at home, making further investigations impossible.

Discussion

Up to 1950. out of 55 reported cases the diagnosis during life had been made on only 16 occasions,[1] since then, the percentage of diagnosis during life has risen considerably. Absence of a lung has been demonstrated at necropsy on sevaral adults who died from causes other than pulmonary failure, however, it remains true that more than half of the patients die during the first year of the life.

Agenesis of the lung is generally unilateral and both hemithoraces and sexes are equally affected. In the review of Maltz and Nadas, out of 164 cases, 148 had unilateral agenesis. Seventy-four had agenesis of the right lung, 72 of the left and in two the agenetic side is unknown. Of 148 instances of unilateral agenesis 57 were males, 74 were females and in 17 the sex was not defined.[6]

The difficulty of the diagnosis depends on the side of the absent lung, the apparent dextracardia induced by the absence of the right lung being a striking clinical finding. When the right lung is absent, the heart is rotated in a clockwise direction and the apex beat is felt in the 3rd or 4th space in the right mid axillary line.[1-7] One of our cases showed clockwise rotation, but no other abnormalities on ECG. There may be obvious flattening of the chest on the affected side with impaired movement on inspiration, the asymmetry of the chest becomes more noticeable in the adult patients than in infants. Although it is reasonable to think that respiratory sounds should be totally absent on the agenetic side, because of the herniation of the other lung diminished respiratory sounds may be audible on the agenetic side. This is especially true when the right lung is agenetic, like in our cases.

For the diagnosis of agenesis of the lung, bronchoscophy, bronchography, and angiocardiography are necessary.

Cardiovascular, gastrointestinal, urogenital, spinal, and ipsilateral facial abnormalities may be associated with lung agenesis.

Recurrent pulmonary infections and the side of the agenesis affect prognosis. The other important prognostic factors are the associated abnormalities. Agenesis of right lung generally has poorer prognosis, the cause may be the associated cardiac malformations.

Summary

Two cases of right lung agenesis are presented. Literature concerning this condition is briefly reviewed. We want to put emphasis on this abnormality especially in the newborn period.

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