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# Isolated Right Pulmonary Agenesis in a 2-Month-Old Infant in the Pediatric Ward of Hospital of Mali

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## Abstract

**Introduction:** Pulmonary agenesis is a rare congenital abnormality defined by the absence of parenchyma, bronchi and pulmonary vessels, frequently associated with other malformations. We describe a case of isolated right pulmonary agenesis diagnosed in the pediatric ward of the Mali Hospital. **Clinical case**: He was a month-old baby boy, born at term by cesarean section of a well-followed pregnancy. He was not resuscitated. He developed dyspnea after a cold. An emergency chest X-ray revealed a right opaque lung. He was referred to the hospital for the exploration of this opacity. At the entrance, it weighed 3910 g, size: 54 cm and the temperature was 36.9˚C. He had a polypnea, an intercostal print. The sounds of the heart were diverted to the right. The rest of the clinical examination was without much particularity. The chest CT scan confirmed the right pulmonary agenesis. Abdominal and cardiac ultrasound was normal. A healing includes oxygenotherapy, serum glucose 10% minding 100 ml/ Kg/day and amoxicillin minding 100 mg/ Kg/day at 3 times by oral way. After 24 hours of treatment, we observed an improvement in dyspnea. On D2, he was weaned off oxygen and resumed feeding. **Conclusion:** Pulmonary agenesis is a rare congenital anomaly that is frequently associated with other malformations, thus making its prognosis poor. The search for malformative abnormalities should be systematic in right pulmonary agenesis.

## Keywords

Pulmonary, Isolated Agenesis, Pediatric, Hospital of Mali

## 1. Introduction

The abnormalities in lung development encompass a wide spectrum of pulmonary aberration. The size and growth abnormalities of the pulmonary parenchyma represent a subset of the developmental pulmonary anomalies [1].

Pulmonary agenesis is a rare congenital malformation of the development of the lung defined by the total absence of lung tissue, bronchi and pulmonary vessels [2]. It results from arrest in lung development linked to factors which occurred before the 26th day of embryonic life [3].

Its prevalence is from 24 to 34 per 1,000,000 live births and 1 per 10,000 to 15,000 autopsies. The etiology is unknown, but its pathogenesis may be associated with genetic factors, such as reproduction of the distal part of the arm of chromosomes 2, viral factors or vitamin A deficiencies [4].

Both lungs can be affected (incompatible with life), but agenesis of the right lung has a poor prognosis [4] [5].

In the absence of other malformations, right unilateral pulmonary agenesis is compatible with normal life, but it is often associated with malformations, severe respiratory infections during childhood associated with high mortality rates [4].

We describe a clinical case in a 2-month-old infant diagnosed in the pediatric ward of the Mali Hospital.

## 2. Clinical Case

IS was a 2 month old male infant who was hospitalized for respiratory distress. Her father was a sickle cell form SS and her mother had no significant medical history except an AS sickle cell trait (A = 79.8% and S = 20.2%). There was no notion of consanguinity in marriage. She had 2 pregnancies and the children were alive. Rubella, toxoplasmosis, syphilis, hepatitis B and HIV serologies performed during pregnancy were negative. The three ultrasounds performed during pregnancy returned to normal.

Our infant was born from a pregnancy well followed to term without major incidents. He was born at 40 weeks of gestation by caesarean section for neonatal suffering. The Apgar at birth was 7 in the first hour and 8 in the fifth minute. The birth weight was 2510 g, the height: 48 cm the cranial perimeter: 33 cm.

IS was the second child in a family of 2 children. Her big sister was doing well. Her vaccination was in progress.

The onset of the disease dates back to a week marked by respiratory distress after an episode of serous rhinorrhea in the context of family flu motivating a consultation in a private clinic where an urgent chest X-ray revealed a right opaque lung (**Figure 1**). He was referred to the hospital for the exploration of this opacity.

At admission he weighed 3910 g, size: 54 cm, head circumference and the temperature was 36.9˚C. His general condition was slightly altered, the conjunctivae were well colored.

The chest was symmetrical with a respiratory rate at 41 cycles/min. The oxy-

gen saturation fluctuated between 91% and 96%. There was a slight intercostal pull, bronchial groans in the left hemithorax. The vesicular murmur was decreased in the right hemithorax. The heart sounds were deflected to the right with an intense systolic murmur audible at the aortic focus. The rest of the clinical examination is without much particularity.

In total, it was a 2-month-old infant admitted for respiratory distress after rhinorrhea, the chest x-ray of which revealed heterogeneous opacity of the right hemithorax. We raised the hypothesis of an acute bacterial pneumonia complicating an infection respiratory viral.

The chest CT scan showed a total absence of the right pulmonary parenchyma, bronchus and pulmonary artery with a deviation of the heart in the right hemithorax (**Figure 2(a)** and **Figure 2(b)**). The blood count showed a hypochromic microcytic anemia at 9.9 g/dl. The white blood cells were at 8510/mm$^3$ and the platelets were at 331,000/mm$^3$. The reactive protein C was at 3.1 mg/l. The rest of the biological assessment was not done for technical reasons and the low socio-economic level of the family.

The abdominal and cardiac ultrasound requested in search of associated malformations had returned to normal. The esogastroduodenal transit also returned to normal (**Figure 3**).

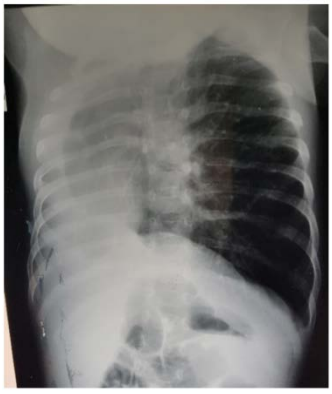

**Figure 1.** Chest x-ray showing opaque and retracted right hemithorax.

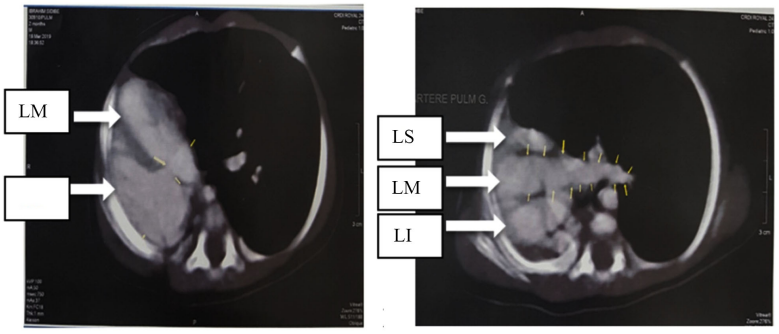


**Figure 2.** CT chest mediastinal window: (a) meadle and inferior lobar agenesis, (b) superior meadle and inferior lobar agenesis.


**Figure 3.** Esogastroduodenal transit in admition.

A treatment combining nasopharyngeal obstruction, bronchial aspiration, oxygen therapy sufficient amount for saturation ≥ 95%, 10% glucose serum infusion: 100 ml/Kg/d over 1 hour and amoxicillin 100 mg/Kg/d in three administrations per os.

After 24 hours of hospitalization, we observed an improvement in respiratory distress and a decrease in oxygen requirements. On the second day of hospitalization, he was completely weaned off the oxygen and released the same day after resuming feeding.

## 3. Discussion

Exceptional bilateral pulmonary agenesis is incompatible with life [1] [6]. Unilateral pulmonary agenesis is also very rare. In 8 years we have recorded 2 cases in the service.

It is usually suspected antenatal before complete absence on the right or left of the lung, bronchus and pulmonary vascularization [6]. It is 2 times more frequent in the male sex. It sits on the left in 70% of cases [3]. The always associated cardiac malposition is an important sign to look for unilateral prenatal pulmonary agenesis [6].

In at least half of the cases, agenesis is associated with other malformations: cardiovascular, gastrointestinal, skeletal, or urogenital [6] [7]. These associated malformations however seem to be much more frequent in right agenesis [6]. Usually symptomatic soon after birth, this malformation can remain asymptomatic until adulthood with a fortuitous diagnosis on the occasion of a chest x-ray [6] [8].

The diagnosis can be suspected on a thoracic asymmetry with reduction of the respiratory amplification of the hemithorax and reduction of the vesicular murmur on this side. Very often the thorax appears symmetrical and the subnormal auscultation due to the distension of the lung from the healthy side to the sick side. The picture is very different in the forms with associated malformations. They can dominate the clinical picture and the prognosis [3].

Pulmonary agenesis radiologically results in an opaque, retracted hemithorax

attracting the mediastinum. The contralateral lung is the site of hypervascularisation with compensatory expansion and anterior mediastinal hernia which can sometimes make clinical diagnosis difficult with a pulmonary auscultation most often normal and a chest X-ray showing pulmonary parenchyma on the side of agenesis [3] [6].

The chest scanner with injection of contrast product confirms the diagnosis. It can be completed by bronchial endoscopy. These examinations make it possible to differentiate agenesis where the root bronchus follows the trachea without keel from the aplasia where there is a keel and a bronchial stump [6].

The prognosis outside of scoliosis, which may require surgical treatment, depends on the associated malformations. A better prognosis seems to be associated with left agenesis. Thus survival after 40 years would be 35% in left agenesis more often isolated and 10% in right forms where the malformation context is more frequent. The existence or not of hypertension on the healthy side is decisive for the prognosis [3].

The installation of an expandable prosthesis seems in the empty hemithorax has been proposed to correct the mediastinal deviation [6].

Its management is based on monitoring and preserving the remaining lung [9].

Our observation was a 2-month-old male infant with right unilateral pulmonary agenesis. It was a broncho-pulmonary malformation less frequent than the left unilateral pulmonary agenesis reported by Tournier *et al.* [3] in the literature and by Malcon *et al.* [10] in an 8 year old boy. The diagnosis of our patient was made postnatal because of the low level of our technical platform. In recent years, advances in obstetric imaging have made prenatal diagnosis possible, so fetal ultrasound and MRI allow early and accurate diagnosis [2]. Pulmonary agenesis was revealed at 2 months of life by intense respiratory distress like the cases reported by Khurram *et al.* [11] in a 2 month old infant and Dinamarco *et al.* [4] in a 5 month old infant. It was an isolated right pulmonary agenesis with no other associated malformations. Excluding right pulmonary agenesis is often associated with abnormalities of the cardiovascular, gastrointestinal, urogenital or musculoskeletal system, thus making its prognosis gloomy [2] [4]. Miyano *et al.* [12] described right pulmonary agenesis associated with esophageal atresia, tracheoesophageal fistula and Nandan *et al.* [13] reported a case associated with dextrocardia.

The radiological diagnosis was made by standard chest X-ray which showed an opaque hemithorax. Confirmation of diagnosis was made by chest CT scan. Treatment included oxygen therapy and antibiotic therapy based on amoxicillin.

## 4. Conclusion

Right pulmonary agenesis is an infrequent bronchopulmonary malformation that is defined by the absence of the pulmonary parenchyma of the spindle and the pulmonary artery. It is frequently associated with heart, gastrointestinal,

urogenital and musculoskeletal malformations, thus making its prognosis poor. The search for these malformation anomalies must be systematic in right pulmonary agenesis.

## Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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