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ISOLATED AGENESIS OF LEFT PULMONARY ARTERY, A CASE REPORT: MID-TERM FOLLOW-UP AND REVIEW OF THE LITERATURE

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BACKGROUND

Unilateral absence of a pulmonary artery (UAPA/UHPA) is a rare congenital anomaly, which occasionally occurs as an isolated finding, but more often is associated with other congenital cardiac heart diseases, such as Tetralogy of Fallot, atrial septal defect, aortic coarctation, truncus arteriosus, and pulmonary atresia.

CASE PRESENTATION

We present the case of a female patient **with isolated agenesis of the left pulmonary artery and associated hypoplasia of the homolateral lung**, who was referred to our Unit at the age of 4 months for a heart murmur.

Perinatal history: She had a full-term normal delivery with a normal APGAR score. Conception/delivery for the mother was eventless. Birth weight 2,780 kg (AGA). Her medical and familiar history was negative. There was no history of cardio-respiratory symptoms.

Clinical presentation: On admission, her weight and height were 4,65 kg (PCT < 3°) and 58 cm (PCT 3°), respectively. A cardiac examination revealed a regular rate and rhythm and a heart systolic murmur.

The **ECG** was normal. The **echocardiography** showed a right aortic arch and a mild dilatation of the right pulmonary artery; the left pulmonary artery was not detected (**figure 1** and **figure 2**). Then, a **chest X-ray** was performed, which highlighted a left-shifted mediastinum, associated with a hypoplastic left lung and a normal right lung. A **Computer Tomography Angiography** was fulfilled (**figure 3**) and revealed unilateral agenesis of the left pulmonary artery without hypertrophy of bronchial and intercostal arteries; right aortic arch, hypertrophy of the azygos and hemiazygos veins with agenesis of the hepatic part of the inferior vena cava and suspected azygos continuation; hypoplasia of the left lung with ground glass area and air trapping; left-shifted mediastinum.

Follow-up: As the patient was asymptomatic and hemodynamically stable, we decided to begin a multidisciplinary follow-up, composed of pediatric cardiologists and pulmonologists. She had good health, except only admission to the Children's Hospital of Parma at the age of 4 years for mild pneumonia. She had no growth delay or other abnormal health problems on medical examination during the follow-up. She regularly attends nursery school. **Figure 4** and **figure 5** show CT at age of 6 years.

DISCUSSION

Agenesis of the pulmonary artery not associated with other congenital cardiac malformations is an uncommon condition. Due to embryological relationships, UAPA commonly occurs on the right, while Unilateral agenesis of the left pulmonary artery is a rarer finding. Interestingly, as in our case, a possible additional finding in this malformation is the presence of a right aortic arch.

UAPA can run asymptomatically until adulthood, causing a diagnostic delay. However, the onset of recurrent respiratory symptoms already in childhood lead to the execution of thorough examinations.

There is no specific treatment for UAPA, although surgical, pharmacological, and behavioral management are available. The long-term survival of patients with this disease is negatively influenced by conditions like pulmonary hypertension (PHT) and pulmonary hemorrhage. A yearly evaluation with echocardiography helps to identify any early signs of PHT or heart failure. Vasodilator therapy is recommended for patients who develop PHT.

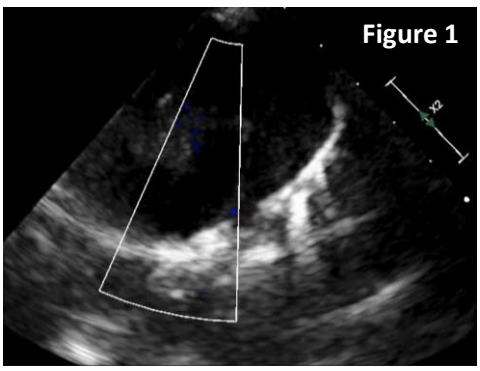
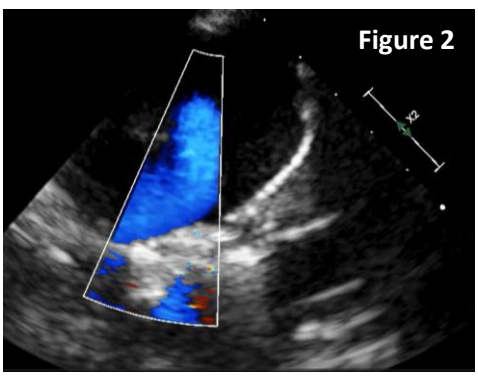


Figure 1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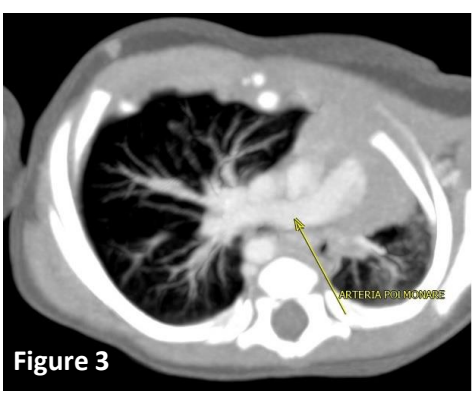

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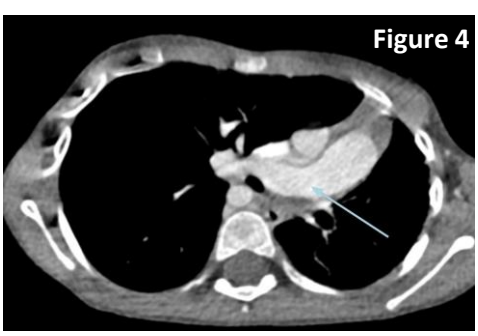
Figure 4

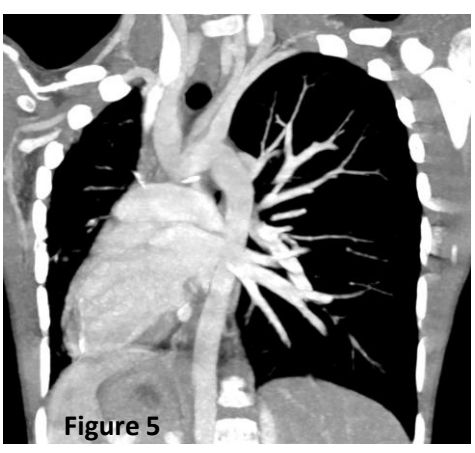
Figure 5