



# Pulmonary Agenesis Associated To A Persistent Left Superior Vena Cava In Adult Patient: Case Report

1Orgi Anas, 2Belaouini Elmahdi, 3Et-tahir Youness, 4Merzem Aicha, 5El Benna Naima

1Dr, 2Dr, 3Dr, 4Pr, 5Pr

120 August 1953 Radiology Department, CHU Ibn Rochd,

220 August 1953 Radiology Department, CHU Ibn Rochd,

320 August 1953 Radiology Department, CHU Ibn Rochd,

420 August 1953 Radiology Department, CHU Ibn Rochd,

520 August 1953 Radiology Department, CHU Ibn Rochd

## Keywords:

Pulmonary agenesis, persistent left superior vena cava, azygos venous system anomaly

## Summary:

Introduction: Pulmonary agenesis is a rare congenital anomaly which may be isolated or present as part of a poly-malformative syndrome.

Case report: A 33-year-old female presented with chronic dyspnoea since childhood. Clinical examination revealed auscultatory silence and dullness on percussion of the left hemithorax. A chest X-ray showed an opaque left lung. Chest CT revealed left pulmonary agenesis associated with persistence of the left superior vena cava, draining the homolateral jugulo-subclavian venous confluence, as well as the accessory hemi-azygos and the hemi-azygos veins, and terminating inferiorly in the right atrium. Hypoplasia of the azygos vein was noted.

Discussion: Pulmonary agenesis had an incidence of 1.2 per 100,000 births in the literature It is sometimes associated with anomalies of the azygos venous system. However, association with the persistence of a left inferior vena cava is much rarer.

**Manuscript:****Introduction:**

Pulmonary agenesis is a rare malformative pathology due to the absence of development of the pulmonary bud. It can occur in isolation or be associated with poly-malformative syndromes.

The aim of our work is to report an exceptional case associating two rare pathological entities, namely pulmonary agenesis and persistence of the left superior vena cava.

**Clinical observation:**

We report the case of a 33-year-old female presenting with chronic dyspnea. The medical history revealed a history of recurrent dyspneic episodes during childhood that were never investigated before. Oxygen saturation at rest was normal, BMI was 24 kg/m<sup>2</sup>, and decreased breath sounds and dullness to percussion were noted over the left hemithorax upon auscultation. A chest X-ray showed opacification of the left lung.

Subsequently, a contrast-enhanced thoracic CT scan was performed, including acquisitions without and with automatic injection of contrast medium as a bolus. The scan revealed absence of visualization of the left lung and compensatory hypertrophy of the right lung. A rudimentary blind left mainstem bronchus was observed, without anomalies or distortion of the trachea or bronchial system of the right lung. Additionally, there was agenesis of the left pulmonary artery.

Persistence of the left superior vena cava was found originating from the jugulo-subclavian venous confluence and draining into the right atrium. It received the accessory hemi-azygos vein and the hemi-azygos vein. The latter was dilated with hypoplasia of the azygos vein. The left superior intercostal vein had a normal diameter (4mm), and the left brachiocephalic vein was patent.

**Discussion:**

Pulmonary agenesis is a rare but well-known condition. Epidemiological literature is scarce, and we do not have data on its prevalence in our country. However, based on Thomas M. et al.'s work on the NorCas database, an incidence of 1.2 per 100,000 births was noted. Bilateral form is incompatible with life; hence, only unilateral cases are encountered in medical imaging.

It can be classified into three types: the first type corresponds to agenesis, defined by the absence of all parenchymal, bronchial, and vascular components; the second type corresponds to aplasia, characterized by rudimentary bronchi with total absence of pulmonary parenchyma; and at the other end of the spectrum, the third type corresponds to pulmonary hypoplasia. In our case, it is an aplasia given the presence of a rudimentary mainstem bronchus.

Azygos venous system anomalies are sometimes associated with cases of pulmonary agenesis. The peculiarity of our case lies in the persistence of the left superior vena cava, which is a rare anomaly affecting up to 3% of the population. It is rarely associated with pulmonary agenesis. In such cases, the inferior drainage usually occurs into the coronary sinus or into the right atrium, especially if a cardiac malformation is associated. The drainage of the accessory hemi-azygos vein and azygos vein into the persistent left superior vena cava is also a rare possibility.

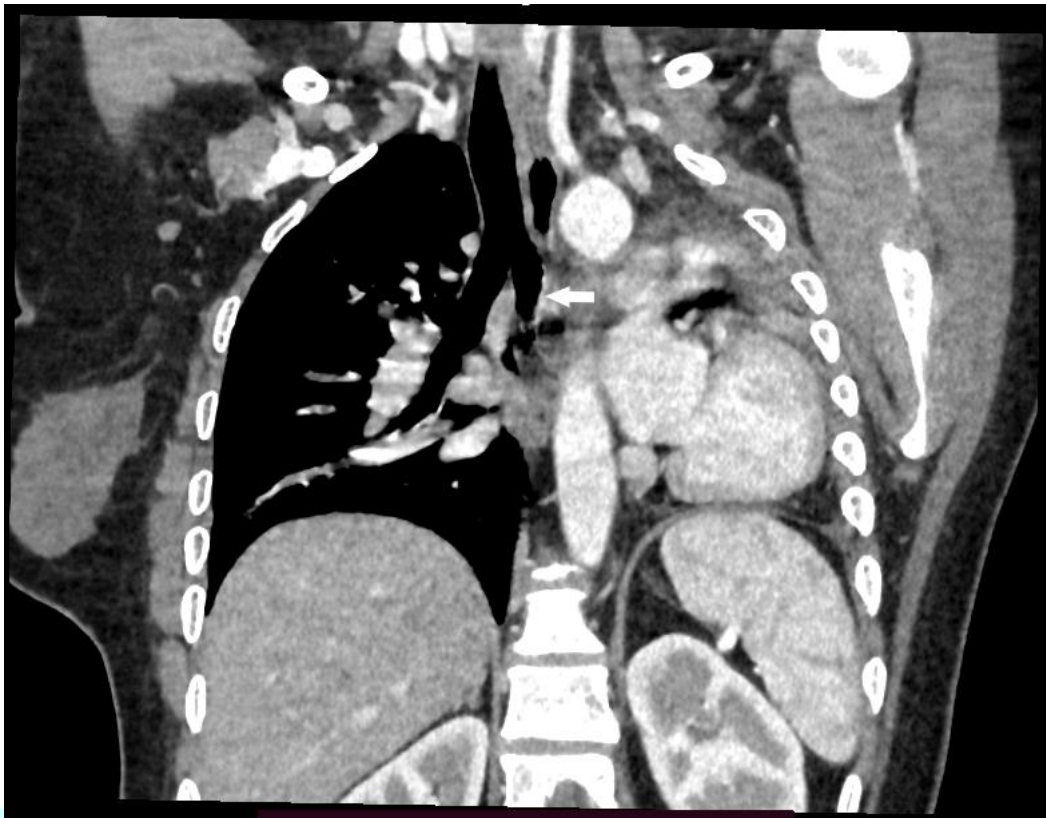


Figure 1



Figure 2

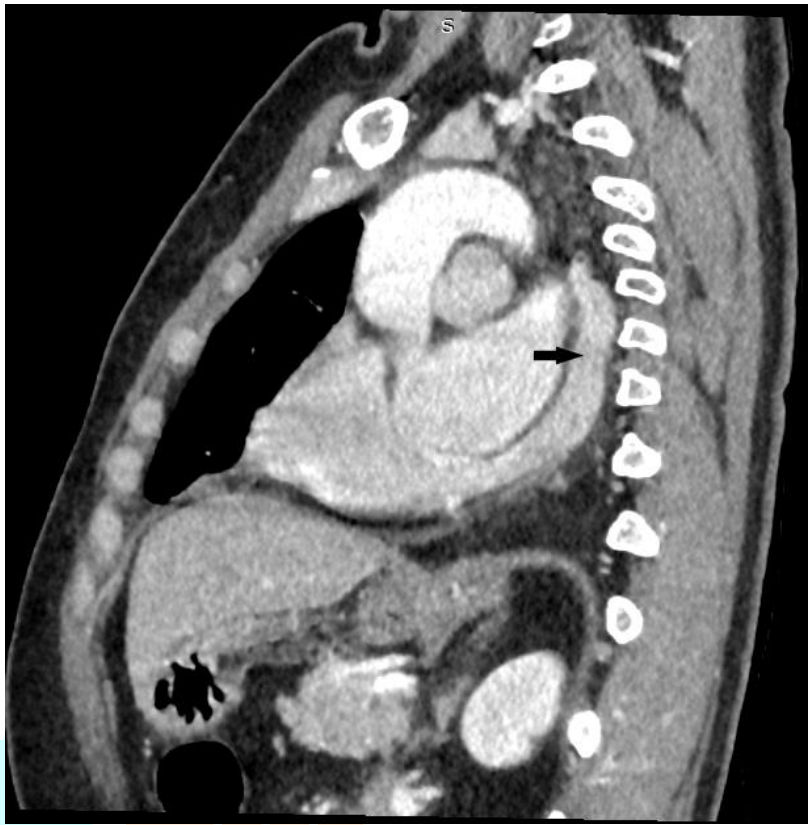


Figure 3



Figure 4

**Figure captions:**

**Figure 1:** Contrast-enhanced thoracic CT scan in arterial phase in coronal section, showing absence of visualization of the left lung with a rudimentary mainstem bronchus (arrow).

**Figure 2:** Contrast-enhanced thoracic CT scan in arterial phase in coronal section, showing the presence of a left superior vena cava (black arrow) originating from the ipsilateral brachiocephalic venous confluence (white arrow).

**Figure 3:** Contrast-enhanced thoracic CT scan in arterial phase in coronal section, showing termination of the left superior vena cava (black arrow) in the right atrium.

**Figure 4:** Contrast-enhanced thoracic CT scan in arterial phase in coronal section, showing drainage of the accessory hemi-azygos vein (white arrow) and hemi-azygos vein (black arrow) into the persistent left superior vena cava (white star).

