

# A rare Congenital Left Lung Hypoplasia with Absent Left Pulmonary Artery and Right-Sided Aortic Arch: a Dilemma in work Fitness Radiograph

## Case Report

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### Abstract

**Purpose:** The purpose of this study is to describe congenital anomaly of unilateral absence of pulmonary artery (UAPA) which lead to a dilemma in health clearance certificate.

**Material and Methods:** Two consequent cases were included in the current study presented to the Communicable Disease Center (CDC) referring from Medical Commission with abnormal chest x-ray for further evaluation. Plain x-ray well studied and followed by CT scan with contrast with post processing were done.

**Pictorial review:** UAPA is considered rare of entity constituting a ratio of 1: 200,000, however it should be included in the differential diagnosis and containment by CT scan and post processing technique.

**Conclusion:** CT scan and post processing technique should be carried out to confirm the suspicious of UAPA which should be included as a differential diagnosis.

**Keywords:** Unilateral absence of pulmonary artery (UAPA); Pulmonary hypertension (PHT); Tetralogy of Fallot (TOF); Computer tomography scan (CT scan); Volume rendering techniques (VRT); Maximum intensity projection (MIP); Multiplanar projections reconstruction (MPR); Tissue transparent projection (TTP)

## Introduction

Congenital anomalies of the respiratory system include wide range of varieties. It might include lung hypoplasia or aplasia and a wide range of associated vascular and cardiovascular anomalies. Unilateral absence of pulmonary artery (UAPA) is extremely rare pathological entity, with its prevalence around 1:200,000 young individual adults [1,3,4]. It has no sex prevalence, with equal male to female ratio. It might be associated with cardiovascular abnormalities such as tetralogy of Fallot (TOF), septal defect arterial or ventricular as associated congenital malformation in 0.34% of the congenital heart disease, yet it can present as isolated entity [17].

Usually the patient with sole UAPA is asymptomatic until adulthood. However associated symptoms may include dyspnea, chest pain, hemoptysis and recurrent attacks of the chest infection which can supervene.

The routine investigation including chest x-ray, echocardiography may suggest the diagnosis of absent pulmonary artery, but conformation is required by CT scan or magnetic resonance angiography (MRA). An early diagnosis of UAPA and appreciate intervention may significantly improve the outcome. It might be a cause of diagnostic dilemma during radiography for work fitness purpose or residence permit.

**Case Report**

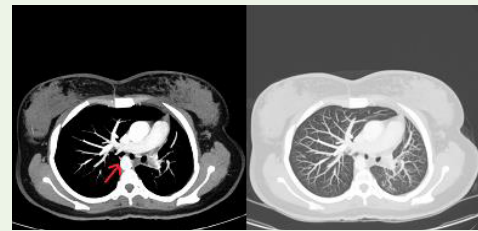
The current study includes two patients, a 27 years old male and a 26 years old female, both came from Indian subcontinent. The patients were referred from Medical Commission Center to the Communicable Disease Center in Doha with abnormal Chest X-ray findings in order obtain health fitness report clearance for working permit, especially to rule out TB. Both patients were clinically and physically healthy and fit with no positive medical or surgical history. The patients had no history of parent’s consanguinity or other siblings’ congenital anomalies, no history of drug intake by their mothers during pregnancy period, or any abnormal incidental radiation exposure, normal vaginal delivery for both of them. Both patients with average body weight and height, normal vital signs, normal oral temperature, peripheral pulse, respiratory rate, blood pressure and oxygen saturation (SPO2). No history of any allergy, night sweating or weight loss. On physical examination there were: normal breath sound, normal heart sounds, no skin rash, no palpable lymphadenopathy, soft lax abdomen, no organomegaly, no neurological deficit. Unremarkable CBC, QUANTIFERON test is negative and TB PCR was negative.

A multidetector CT scan (MDCT) of the chest before and after IV contrast administration of non-ionic contrast media was done, it revealed reduced volume of the left lung/hypoplasia, absence of the left pulmonary artery/agenesis with otherwise normal caliber and opacification of the main pulmonary trunk and the right pulmonary artery and its segmental and its subsegmental branches, the aortic arch was abnormally located on the right side with slightly higher position of the left diaphragmatic copula (Figure 1,2).

Furthermore, there was evidence of arterial collaterals on the left chest wall as well as prominence of the left internal mammary artery. The findings are clearly visualized with the use of volume rendering techniques images (VRT), maximum intensity projection images there (MIP) as well as the reconstruction in multiplanar projections (MPR) (Figures 3,4,5).



**Figure 1:** CT scan to program of the chest AP view shows discrepancy of both lung fields being of reduced volume on the left, compensatory hyperinflation of the right lung, shift of mediastinum into the left side (similar findings was noticed in the other patient).



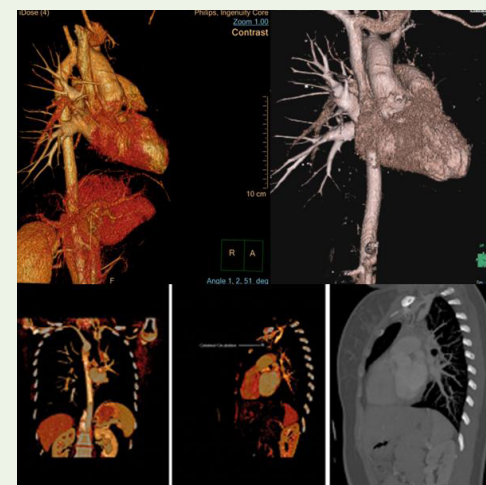
**Figure 2:** Selected mediastinal window and lung window on axial CT scan images of chest post contrast series confirming the above mentioned finding in figure1 in addition to the right-sided aortic arch (ARROW) and absence of left-sided pulmonary artery.



**Figure 3:** Selected mediastinal window and lung window on axial CT scan images of chest post contrast series confirming the above mentioned finding in figure1 in addition to the right-sided aortic arch (ARROW) and absence of left-sided pulmonary artery.



**Figure 4:** TTP (Tissue transparent projection) and VRT coronal orientation shows reduced left lung volume and normal trachea and bronchial tree.



**Figure 5:** VRT of the vessels of shows absence of the left pulmonary artery and right sided aortic arch as well as the prominent collaterals (ARROW).

## Discussion

The unilateral absence of pulmonary artery (UAPA) is a rare vascular malformation discovered and it was published for the first time at the year 1868 [1]. The UAPA can occur in both sexes equally, the mean age of UAPA at presentation is mostly at the age of 14 years [1,2]. In our cases there was slightly delayed diagnosis up to 26 years/27 years. The UAPA statistically founded isolated between 1:200,000 to 1:300,000 [1, 3, 4]. Right-sided UAPA is more predominant and appear about twice than the left side, however left-sided UAPA is found to be less frequent [1,3]. It can be embryologically explained as a result of failure in the connection of the sixth aortic arch with the pulmonary trunk [5,6]. There are multiple factors that may play a role in pulmonary artery (PA) agenesis with pulmonary hypoplasia among the etiology such as chromosomal defect, exposure to intrauterine infection, Vitamin A deficiency and some environmental factors [1,2]. However, UAPA associated with regression of the blood supply to the affected lung causing a result of paucity of vascular markings, lung to appear hypoplastic with reduced volume. Most of the patients presented in the literature were found asymptomatic, which is similar to our cases. Up to 30% of them may stay clear and asymptomatic until adulthood [1,2]. However, some symptoms can be occurring such as exercise intolerance, recurrent cough or recurrent chest pain [7-11]. The pulmonary hypertension (PHT) mainly discovered at the infant age in those cases of left side UAPA, and those patients are mostly surviving until adulthood [7,8]. Life threatening complication such as haemoptysis due to excessive development of collateral can be presented in some cases [7,8,12,13]. Bronchiectases may appear as sequel of recurrent pulmonary infection, they were not present in our cases [8,9]. Diagnosis of UAPA can be suspected from plain CXR with such findings as lung volume reduction, crowding of the ribs, ipsilateral elevation of the diaphragm, contralateral compensatory hyperinflation, absence of the pulmonary artery shadow on the affected side, slight paucity of vascular markings [8,14]. CT scan of the chest with IV contrast and MRI plays an important role in confirming the diagnosis of UAPA, which gives accurate anatomic details, vascular collaterals or any cardiac anomaly. Pulmonary angiography is considered as a gold stranded procedure for accurate diagnosis of UAPA. However, it is an invasive radiological procedure which is mostly advices for those patients who need bronchial arterial embolization as a treatment of haemoptysis. [8,9,14]. Trans thoracic echocardiogram has an important role in most of associated cardiac anomalies and for assessment of pulmonary hypertension (PHT) [12]. While the perfusion-ventilation scan (V/Q) is used to evaluate the pulmonary activity in the normal lung and in the affected one. The difference in activity in bilateral lung fields considered as important part in confirming the diagnosis [12]. The patients who were proved to have UAPA, need further evaluation including routine echocardiography for monitoring of asymptomatic patients to rule out pulmonary hypertension as a complication [14,15]. This could be usually managed with pulmonary hypertension vasodilator therapy [7,14,16]. Patients with congenital heart defects associated with UAPA need palliative surgical intervention as pulmonary artery shunt, transluminal balloon pulmonary valvuloplasty or palliative reconstruction of the right ventricular outflow tract [17,18]. Our two cases were referred to rule out tuberculosis infection, which

was excluded based on x-ray and CT. The chest x-ray and CT chest revealed congenital absence of the left pulmonary artery, left lung hypoplasia and right sided aortic arch. Bronchoscopy done for one patient and result was negative for TB. Medical examination including chest radiography is essential to get significant of medical clearance and accordingly the process of residency can be continued. Any suspicious or any abnormal findings result in a delay of the process and it is considered as a dilemma for work fitness it needs further workup for deceleration.

## Conclusion

Agenesis of the left pulmonary artery is a rare entity and adult patients are often asymptomatic. Imaging plays a major role to confirm diagnosis and detecting the associated findings in heart and lungs. Those patients with chest x-ray usually show unilateral relatively small left hemi thorax. The deferential diagnosis (UAPA) should be considered and kept in mind and subsequently the mystery and clinical diagnostic challenge situation can be solved. On the other hand, early diagnosis help in avoidance of serious complications, like haemoptysis that might supervene and work fitness. Despite of being a rare entity (UAPA), however it should be kept in mind, including in differential diagnosis and confirm by cross-sectional images (CT scan).

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