

# Unilateral Absence of a Pulmonary Artery (UAPA): Case Report and Literature Review

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One of the more infrequent congenital diseases with a wide spectrum of symptoms and more complications of the right side of pulmonary artery (63% according to Ten Harkel) is unilateral pulmonary artery agenesis (UPAA). The clinical presentation is variable, and many patients can be asymptomatic for many years and even throughout their lives. Although the disease more frequently appears in childhood with moderate to severe respiratory symptoms, some patients continue living with the disease up to adulthood. The abnormality is sometimes concurrent with cardiac anomalies. Patients with UAPA may be asymptomatic or may present with recurrent pulmonary infections. In a report, a 25-year old woman who had inductional termination of pregnancy at 35 week last menstrual period (LMP) in a rural hospital due to preeclampsia reported as the UAPA patient.

**keywords:** case report; respiratory symptoms; unilateral pulmonary artery agenesis

Unilateral absence of a pulmonary artery (UAPA) is a rare congenital anomaly most often accompanied by cardiovascular disorder [1-4] or may occur as an isolated finding [5]. Although patients with isolated type

may remain asymptomatic until adulthood [6], others may have severe pulmonary hypertension. According to a retrospective cohort study, the analysis of comparison between isolated UAPA and UAPA with Patent ductus arteriosus (PDA) revealed higher prevalence of pulmonary hypertension, higher mortality rate and even earlier diagnosis [7].

The most common symptoms include frequent pulmonary infection (37%), dyspnea or limited exercise tolerance (40%), hemoptysis (20%) and pulmonary hypertension (44%) [8-9]. The hypothesis of recurrent infectious pathogenesis is unknown, but it is assumed that dysfunctional mucociliary clearance and decreased inflammatory cells activity resulting from alveolar hypocapnia have an important role. Excessive systemic collateral circulation causes hemoptysis, which may involve bronchial, intercostal, subclavian, or sub diaphragmatic arteries [10-11].

There is no right versus left or male versus female predominance [6], but right pulmonary agenesis has a poor prognosis and a high risk of coexistence with other congenital anomalies [12]. In this regards, the literatures declaimed that patients with left-sided agenesis, which is more common, have a longer life expectancy than those with right-sided agenesis. For unknown reasons, pulmonary agenesis of the right side is associated with a higher frequency of other congenital anomalies than the left side [13]. A majority of patients with unilateral agenesis die soon after birth or in early childhood, in some extreme cases, but patients with no associated anomalies and minimal or no symptoms may survive into adulthood. In the manuscript, the researchers discussed the case of UAPA in a 25-years-old woman which was diagnosed after early termination of pregnancy by induction.

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## Case report

Patient was a 25-year old woman who had inductional termination of pregnancy at 35 week LMP in a rural hospital due to preeclampsia. Six hours after her delivery, she feels progressive dyspnea and respiratory distress, thus she was intubated and admitted to the intensive care unit (ICU) and mechanical ventilation was begun. Because of the left sided white lung on the first CXR (Figure 1), a chest tube was inserted but there was no drainage of fluid or air and then the patient was transferred to our hospital for more sophisticated diagnosis and treatments.

In the past medical history, patient reported dyspnea, easy fatigue and exercise intolerance which made her refer to a cardiologist and a large PDA with bidirectional flow and high PAP (65/35) was found. Then she was operated by banding and closure of PDA and discharged and advised to follow ten years later for second stage of surgery. Moreover, in the past medical history was one full term pregnancy with living baby, and mild respiratory distress after delivery which was resolved by diuretics.

At the time of admission, meanwhile stability of patient condition in our ICU, the patient was sedated and was put under mechanical ventilation upon Synchronized Intermittent mandatory ventilation (SIMV) mode and high PEEP (15cm H<sub>2</sub>O) for correcting intractable hypoxemia.

Echocardiography showed normal function of left ventricle and right ventricle, ejection fraction of 60% and PAP=65 cm H<sub>2</sub>O. Chest spiral CT-scan was done which revealed disseminated alveolar infiltration in left lung, air bronchogram in lingual lobe and atelectatic changes in lower lobe of right lung (Figure 1). Ultra sonography of lower limbs, abdomen and pelvis showed no deep venous thrombosis.

However, with aforementioned condition in spite of patient condition she was scheduled for CT-angiography, however, the limitation in transferring patient, postponed the test and treatments continued. By improvement of general condition, weaning was begun again and patient was extubated two days later.

Then, patient was sent for CT-angiography which demonstrated incidental finding of right pulmonary artery hypoplasia (Figure 2).

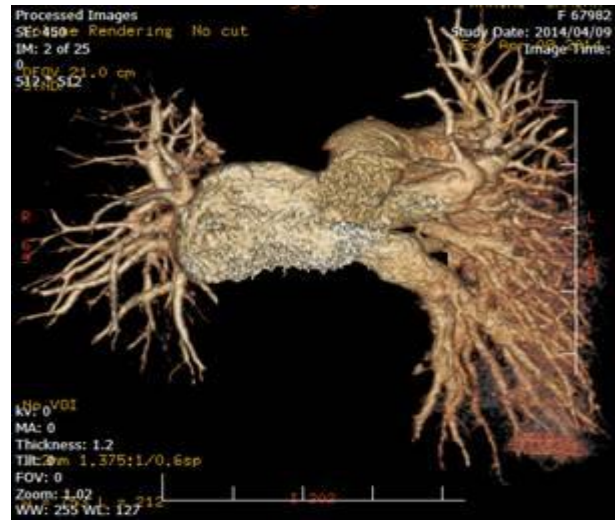
In angiography, it was confirmed in addition to small collaterals to right lung from brachial arteries (Figure 3).

At last, in spite of medical advices, the patient refused to continue more work up and treatments and was discharged. Three months after discharge, she was in good condition with limitation in supra ordinary activity, but without dyspnea at usual activity or any requirement for supplemental oxygen.

**Figure 1- Non-enhanced thoracic CT scan reveals ground glass opacity and consolidation in left lower lobe and lingula. Chest tube is present in left hemithorax.**



**Figure 2- Coronal reconstruction (a) and volume rendered (b) images of pulmonary CT angiography depicts prominent left pulmonary artery and its branches and non-visualization of right pulmonary artery and its branches. Consolidation is noted in left mid-lower lung field.**



**Figure 3- Thoracic angiographic views with the injection (a) in the origin of main pulmonary artery depicts non-filling of right main pulmonary artery and (b) injection in the origin of right internal mammary artery shows enlarged main artery and intercostal branches making collateral network in right pulmonary hilum which supplies right lung.**



## Discussion

Extensive review of the published literatures in pub med showed that up to now (22 January 2016), 181 papers with the title UAPA were published. UAPA is a rare developmental defect which is characterized by a complete absence of the lung and its bronchus [14]. Actual prevalence of UAPA is difficult to establish because many patients with UAPA can remain asymptomatic for a long period. The majority of patients are identified incidentally during routine medical evaluations performed for different reasons [10]; however, some studies reported that prevalence of the rare congenital aberration is 34 per million live births [13] or its incidence varies between 1/10,000 to 12,000 births [14] that was first observed and described by De Pozze during the autopsy of an adult woman that had a failure of development of the primitive lung bud [13].

Pulmonary agenesis may be diagnosed in isolation, but is

frequently associated with other congenital abnormalities, particularly tracheal-esophageal fistula and VACTEL (vertebral, anal, cardiac, trachea-esophageal, and limb) syndrome [15-17] or in other studies it is elucidated that associative congenital abnormalities have been found in more than 50% of the patients, which mainly involve the cardiovascular, gastro-intestinal, musculoskeletal and urogenital systems [12,18], chest wall, diaphragm, ipsilateral face and abdominal wall, vertebral anomalies are also seen [19].

The anomaly initially classified into 3 variants through type one to three including "agenesis (complete absence of pulmonary parenchyma and bronchus and absence of the pulmonary artery on the affected side)", "aplasia (complete absence of pulmonary parenchyma but with a rudimentary bronchus), and "hypoplasia (variable amounts of pulmonary parenchyma, bronchi, and supporting vessels are present)" [20]. Although the exact etiology of this condition is

unknown, as per experimental evidence, genetic factors (maybe an anomaly in the short arm (p arm) of chromosome 2 [12]), viral agents, dietary deficiency of vitamin A during pregnancy, folic acid deficiency and salicylates are alleged to be responsible [14,21].

The diagnosis of agenesis of the lung is usually suspected from the chest radiograph, which commonly reveals a homogeneous opacity occupying most of the affected hemithorax and displacement of the mediastinal contents into the empty hemithorax [13]. Radiographically, agenesis of a lung may appear as pneumonectomy or a total collapse of the lung. The differential diagnosis may also include diaphragmatic hernia and eventration, pneumonitis, pleural effusion, hypoplasia, obstructive lung diseases and mainly, lung cancer [14]. If an abnormal shadow is not observed in the lung field and lung volume reduction is observed on one side in CXR, the possibility of UAPA should be taken into account for hemoptysis. In such cases, collateral circulation should be evaluated by angiography to obtain useful information for treatment [5]. CECT findings show an opaque hemithorax with a shift of the mediastinum to affected side and herniation of the healthy lung to the affected side. Although not necessary, bronchoscopy, if available, may be performed for visualization of the rudimentary or absent bronchus. Regarding management, asymptomatic cases need no intervention, but prevention of infection in the solitary lung has paramount importance. Chest infections of the solitary lung can be life-threatening and should be treated promptly and aggressively with antibiotics, bronchodilators, and physiotherapy. Asymptomatic cases and patients with minimal symptoms have good prognosis.

Unilateral agenesis of the pulmonary artery is often diagnosed initially as another disease, such as pneumonia, because the condition is rarely reported; moreover, the most common clinical symptoms in patients with pulmonary artery atresia are recurrent pulmonary infection, mild dyspnea and exercise intolerance; however, in this case, acute pulmonary edema in pregnancy is a dangerous and life threatening event [1]. Its etiology can be divided in pulmonary edema without hypertension and with hypertension [2]. In the first group, (acute pulmonary edema without hypertension), the most common associated factors and causes are tocolysis, sepsis, pre existing cardiac disease; pregnancy associated cardiac disease (cardiomyopathy, ischemic heart disease), amniotic fluid embolism, aspiration and iatrogenic excess intravenous fluid administration. These women may be normotensive or hypotensive. Preventing strategies include early recognition of high risk patients and careful fluid management. The second group, hypertensive disease of pregnancy, nearly affects 15% of pregnant women [3]. Hypertension is the main manifestation of preeclampsia. Acute pulmonary edema is the leading cause of death in preeclampsia [4-5]. The underlying mechanism of pulmonary edema in preeclampsia was not clearly defined but a range of cardiovascular abnormalities, such as elevated systemic vascular resistance, and reduction in plasma colloid pressure and altered endothelial permeability was assumed [6-7].

At the other hand, cardiogenic pulmonary edema usually represents a bilateral "butterfly" or "bat's wing" appearance with symmetrical homogeneous density, but there are cases which pulmonary edema is unilateral. Unilateral pulmonary edema may be a confusing entity, causing delay in diagnosis

and treatment. Unilateral cardiogenic pulmonary edema is a rare clinical disorder with diagnostic dilemma and is correlated with an independent increased risk of mortality which must be recognized as soon as possible to avoid delay in treatment [1,22-24]. Various factors, including neurogenic alterations in capillary size and permeability, local variations in pulmonary venous pressure, anomalous vascular distribution, local emphysematous changes, and pulmonary inflammatory diseases, have been postulated as causes of unilateral pulmonary edema.

Most cases of unilateral pulmonary edema reported in the literature occur on the upper right side and are caused by severe mitral regurgitation [2]. It is a common finding in those who have a unilateral perfusion defect, in those whose perfusion defect of one lung was distorted and in those with cardiac failure who lie in one side for a long time [8-9].

Chest radiography and even echocardiography are conductive diagnostic test to find UPAA, however to confirm the diagnosis usually computed tomography and MR angiography are required.

Our case had a unique condition, because she was a case of preeclampsia with a history of congenital cardiac anomaly. In this case, presence of pulmonary hypertension and unilateral absence of pulmonary artery, in addition to hypertension and pregnancy induced changes in hemodynamic of patient, resulted in the development of unilateral pulmonary edema. Although most of the findings were detectable on the chest X-ray of our patient, but as these findings are not specific, possible diagnosis such as pneumonia, pulmonary tuberculosis and pulmonary thromboembolism were considered. Misdiagnosing of unilateral white lung as pleural effusion and insertion of chest tube was the result of failure in notifying past medical history and also the rapidity of formation of white lung, which in it; pleural effusion is slower to form than pleural edema.

Hence to achieve the final diagnosis, CT angiography was done and revealed a diagnosis which often not counts as a differential diagnosis.

## Conclusion

The unilateral pulmonary agenesis in an adult patient with radiographic findings such as asymmetric aeration of the lungs should be kept in mind, even in absence of major respiratory symptoms, as in our case. Moreover, the past cardiovascular history of diagnosed patient should be considered. As in the present case, the consideration of PDA history could prevent probable pulmonary edema and acute heart failure in regard to pregnancy induced hypertension, which resulted in decreasing level of aspiration and then pneumonia.

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