

## CASE REPORT

# Isolated Unilateral Pulmonary Artery Agenesis: Report of Two Cases

Muniza Bai<sup>1</sup>, Govindaraj Vishnukanth<sup>2</sup>, Dharm Prakash Dwivedi<sup>3</sup>, Adimoolam Ganga Ravindra<sup>4</sup>

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## ABSTRACT

Unilateral agenesis of pulmonary artery (UAPA) is a rare congenital anomaly that occurs due to malformation of the sixth aortic arch and about 70% of the patients have associated cardiovascular anomalies. However, UAPA without any associated cardiovascular anomalies is termed isolated UAPA. Cases of isolated UAPA may remain asymptomatic and survive into adulthood. Majority of the adults are asymptomatic and can present with recurrent respiratory tract infections. Presence of pulmonary hypertension carries a poor prognosis. There is no specific treatment for UAPA. We report two patients of UAPA with adult presentation.

**Keywords:** Computed tomography pulmonary angiogram, Lung, Unilateral agenesis of pulmonary artery.

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## ABBREVIATIONS USED IN THIS ARTICLE

UAPA = Unilateral agenesis of pulmonary artery; CTPA = Computed tomography pulmonary angiography; CT = Computed tomography

## INTRODUCTION

Unilateral agenesis of pulmonary artery is a rare congenital anomaly that occurs in the embryonic period due to malformation of the sixth aortic arch.<sup>1,2</sup> It is frequently associated with cardiovascular anomalies<sup>2</sup> and UAPA without any associated cardiovascular anomalies is termed as isolated UAPA. Although UAPA presents during the patient's first year of life, isolated UAPA may remain undiagnosed until adulthood and then it is frequently misdiagnosed.<sup>3</sup> Herein, we describe a case series of two patients who were diagnosed in adulthood, as having had isolated UAPA. The resultant observations emphasize that a high index of clinical suspicion is necessary to correctly diagnose UAPA and clinicians should consider the possibility of UAPA in their differential diagnosis of patients presenting with recurrent respiratory tract infections or hemoptysis.

## CASE REPORTS

### Case 1

A 76-year-old woman presented with complaints of exertional dyspnea and recurrent respiratory tract infections since childhood. She had no prior diagnosis of asthma or tuberculosis. She had no comorbid illness. She was taking bronchodilators and antibiotics during increase in symptoms. On presentation to our department, she was stable and maintained normal saturation. Clinical examination was unremarkable. Lung auscultation revealed bilateral normal vesicular breath sounds. Chest radiography showed an absent right hilum with features of volume loss in the right side with tracheal and mediastinal shift to the right. The left lung appeared hyperinflated (Fig. 1). Computed tomography pulmonary angiography (CTPA) showed absent right pulmonary artery and right lung hypoplasia. The main pulmonary artery was seen continuing as the left pulmonary artery (Fig. 2). Cardiac

<sup>1-4</sup>Department of Pulmonary Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India

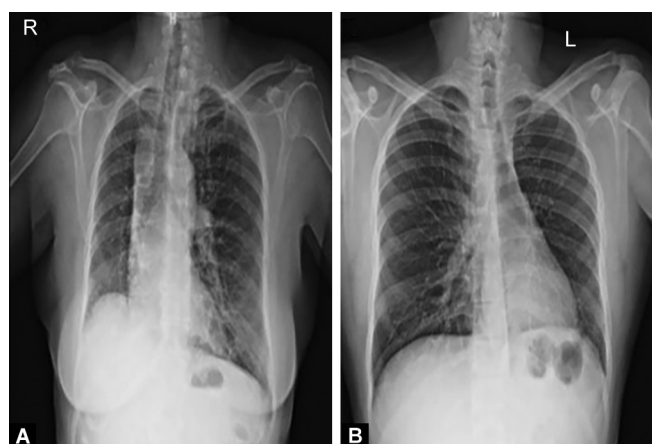
**Corresponding Author:** Govindaraj Vishnukanth, Department of Pulmonary Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India, Phone: 0413-2296202, e-mail: vishnu1429@gmail.com

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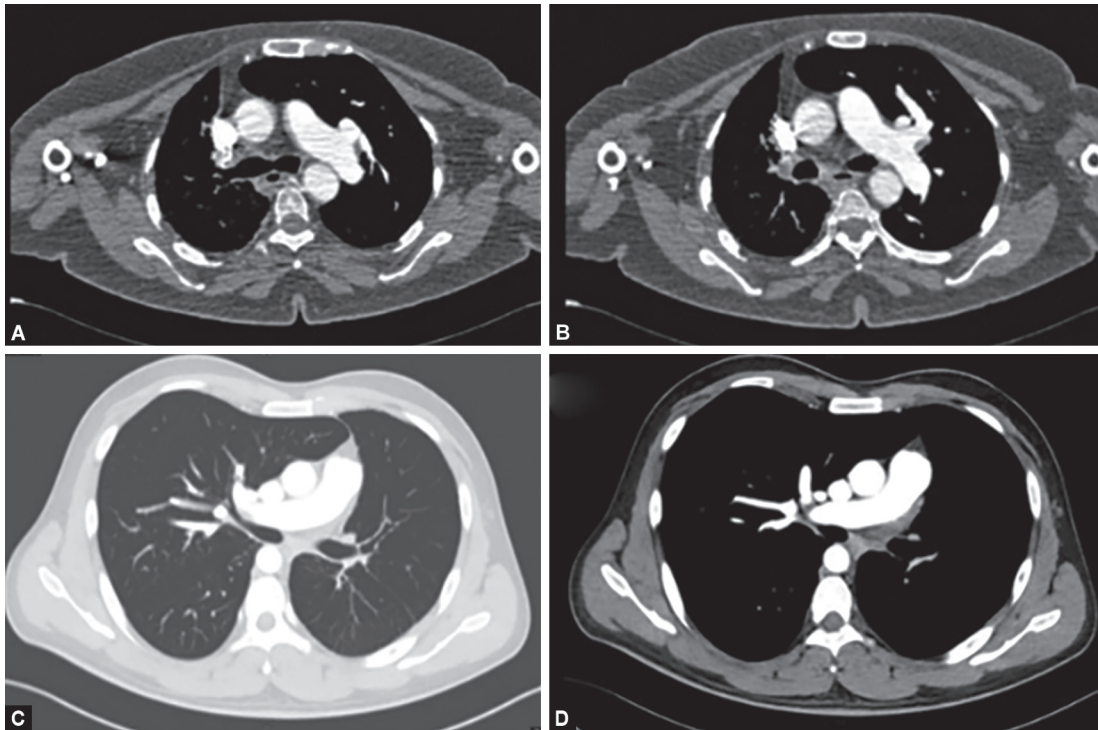
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ultrasonography showed no cardiac anomalies and no signs of pulmonary hypertension. She was explained about her condition and was managed symptomatically.



**Figs 1A and B:** Chest radiograph (postero–anterior view) (A) Case 1 showing an absent right hilum, tracheal, and mediastinal shift to the right and compensatory left lung hyperinflation. (B) Case 2 showing features of left lung volume loss and compensatory right lung hyperinflation



**Figs 2A to D:** (A and B) CTPA of case 1 showing absent right PA with main PA continuing as Left PA; (C and D) CTPA of case 2 showing absent left pulmonary artery with few collaterals from aorta and bronchial arteries

### Case 2

A 25-year-old male was admitted for emergency laparotomy for adhesiolysis and appendectomy for intestinal obstruction and appendicular perforation. Post-extubation, he developed breathlessness for a day without any other respiratory complaints and had no significant past medical history. Clinical examination was unremarkable. Electrocardiogram showed sinus tachycardia. Chest radiography showed tracheal and mediastinal shift to the left with left hemidiaphragm elevation and right lung hyperinflation (Fig. 1B). Computed tomography pulmonary angiography showed right-sided aortic arch with mirror image branching, absent left pulmonary artery with few collaterals from aorta and bronchial arteries (Figs 2C and D). Ventilation–perfusion scan revealed severely reduced ventilation and perfusion of the entire left lung and a normal right lung (Fig. 3). No signs of pulmonary hypertension and cardiovascular malformations were noted in cardiac ultrasonography.

Both patients were conservatively managed, counselled regarding their clinical condition, and advised regular follow-up in pulmonology clinic.

### DISCUSSION

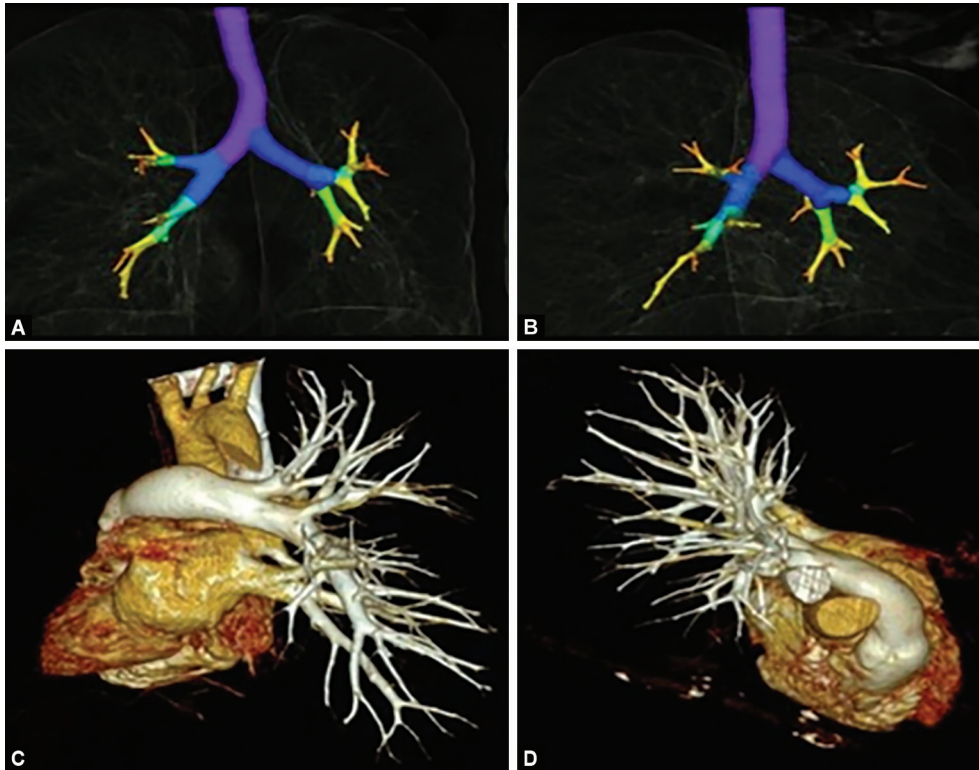
Unilateral agenesis of pulmonary artery is a congenital anomaly that occurs as a result of failed migration and rotation of the sixth aortic arch during embryogenesis.<sup>1</sup> It is a rare condition with its prevalence in the general population ranging from 1 in 200,000 to 1 in 300,000.<sup>2</sup> Approximately 70% of the patients with UAPA have associated cardiovascular congenital abnormalities, such as truncus arteriosus, atrial septal defect, tetralogy of Fallot.<sup>2</sup> Cases of isolated UAPA may remain asymptomatic and survive into adulthood.<sup>3</sup> The patients who present late come with a few symptoms, an abnormal chest radiograph, and usually a variety of erroneous diagnoses.

Unilateral agenesis of pulmonary artery is found to commonly occur on the right side.<sup>3</sup> Left-sided UAPA is usually associated with cardiovascular malformations and rarely presents as isolated UAPA. It is found to affect both sexes equally. It can manifest itself anytime from infancy to adulthood, the median age of presentation being 14 years.<sup>4</sup> Majority of the adults are asymptomatic. When symptomatic, they may present with exertional dyspnea (20–40%), hemoptysis (20%), recurrent respiratory tract infections, and chest pain.<sup>3</sup> Pulmonary hypertension is observed in 25% of patients and it is associated with poor prognosis.

A lung with decreased or absent perfusion has reduced defense mechanisms. Patients can present with recurrent respiratory tract infections which could be due to impaired mucociliary clearance and defective delivery of the appropriate inflammatory cells to deal with inhaled microorganisms. Reduced mucociliary clearance and repeated infections can lead to bronchiectasis. Hemoptysis in UAPA is caused by excessive collateral circulation.<sup>5,6</sup>

The affected lung may become hypoplastic due to reduced blood supply, although the distal intra-pulmonary branches of the anomalous pulmonary artery may receive collateral blood supply from other arteries, such as bronchial, internal mammary, subdiaphragmatic, subclavian, intercostal, and even the coronary arterial networks.<sup>4</sup> Other reasons for the hypoplastic lung include the following: (i) Alveoli not involved in the exchange of gases due to poor pulmonary circulation atrophy; (ii) alveoli with an insufficient blood supply eventually collapse due to its inability to produce adequate amounts of surfactant; and (iii) poor pulmonary circulation may accelerate alveolar involution due to aging. However, none of these possibilities is currently supported by clear evidence and need further studies.

Bahler classified congenital isolated absence of the right pulmonary artery into three groups. Group I is an isolated absence



**Figs 3A to D:** (A and B) Virtual reconstruction images of bronchial tree showing normal bronchial anatomy and left lung volume loss; (C and D) Virtual reconstruction images of vascular structures showing absent left pulmonary artery

of the right pulmonary artery complicated by a left-to-right shunt. These patients tend to develop cardiac failure or respiratory symptoms during infancy and thereafter. Group II is complicated by pulmonary hypertension and most patients die in infancy. Group III is not complicated by pulmonary hypertension and most patients survive to adulthood. But they may suffer from repeated episodes of hemoptysis or respiratory infection. Some may have no symptoms and would be incidentally detected during medical examination.<sup>7</sup>

An abnormal chest radiograph can prompt a diagnostic suspicion in adulthood. The characteristic findings on the plain chest radiograph include ipsilateral cardiac and mediastinal displacement, ipsilateral volume loss features, ipsilateral absent or reduced pulmonary vascular markings, contralateral compensatory hyperinflation and herniation of lung across the midline. The differentials include lobar atelectasis, post-lobectomy status, chronic pulmonary thromboembolism, Swyer–James syndrome, pulmonary artery hypoplasia/agenesis, and pulmonary hypoplasia/agenesis.<sup>8</sup>

Although plain chest radiograph serves as a very useful tool, the diagnosis of UAPA can be confirmed by chest computed tomography (CT), magnetic resonance imaging (MRI), or nuclear ventilation-perfusion imaging. Chest CT can reveal parenchymal findings like bronchiectasis, mosaic attenuation pattern in both lungs, possibly caused by an increased perfusion of the unaffected lung, by the development of pulmonary hypertension or by a compensatory over-inflation of the unaffected lung. It can also provide information on pulmonary hypertension, collateral circulation and congenital cardiovascular anomalies.<sup>9</sup> The diagnosis of UAPA can be confirmed by ventilation-perfusion imaging. Although the usual description of the nuclear ventilation-perfusion scan in UAPA is no perfusion

with intact ventilation, diminished ventilation in UAPA has also been reported confirming our observation.<sup>10</sup>

Not all patients with UAPA have pulmonary hypertension at the time of presentation. Yearly echocardiography can serve as a useful tool to pick up pulmonary hypertension during follow-up. Bronchoscopy may not help in the diagnosis of UAPA.<sup>3</sup>

The overall mortality is 7%.<sup>4</sup> Common causes of death include respiratory failure, massive hemoptysis, right heart failure, and high-altitude pulmonary oedema. While there is no specific treatment for UAPA, pharmacological, surgical and behavioral therapies are indicated.<sup>4</sup> Vasodilator therapy is recommended for UAPA associated with pulmonary hypertension. Lobectomy or pneumonectomy is recommended in patients with massive hemoptysis or recurrent respiratory tract infections.<sup>11</sup> For patients not fit for definite operative management, selective embolization of bronchial or non-bronchial systemic arteries can be an alternative option.<sup>6</sup> Revascularization of the peripheral branches of the affected pulmonary artery to the pulmonary hilum has been attempted successfully, with better results noted in the pediatric population.<sup>11</sup>

In conclusion, given the possibility of undiagnosed cases of UAPA continuing into adulthood, the presence of pulmonary artery agenesis should be considered in differential diagnosis while treating patients with recurrent respiratory tract infections or hemoptysis. A high index of suspicion and judicious use of a battery of multiple imaging techniques can lead to the correct diagnosis.

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