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## Unilateral absence of right pulmonary trunk: A rare case presentation in a 40-year-old male with respiratory symptoms

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### Abstract

We present a unique case of a 40-year-old male presenting with chief complaints of breathlessness and intermittent chronic cough. High-resolution computed tomography (HRCT) revealed architectural distortion of the right lung, characterized by irregular reticulations and smooth inter and intra-lobular septal thickening with mild microcytic honeycombing, exhibiting an apicobasal gradient along the sub-pleural border. Additionally, ipsilateral mild pleural thickening was observed. Bilateral lung fields showed multiple fibrotic bands and streaks with tractional bronchiectasis, leading to volume loss manifested by ipsilateral mediastinal shift and contralateral lung hyperinflation. Subcentimetric lymph nodes were noted in the bilateral lower para tracheal and pre-vascular regions. Despite a normal heart size, HRCT revealed isolated unilateral right-sided absence of the pulmonary artery, accompanied by hypervascular ipsilateral collateral vasculature. Atherosclerotic changes and calcified plaque were evident in the aorta, with significant thickening. This case underscores the importance of thorough investigation in patients presenting with respiratory symptoms, as rare anatomical anomalies such as unilateral absence of the pulmonary trunk may have significant clinical implications and require tailored management strategies.

**Keywords:** Unilateral absence of pulmonary artery, high-resolution computed tomography (HRCT), respiratory symptoms

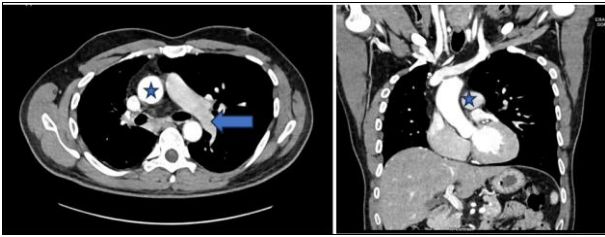
### Introduction

Unilateral absence of the pulmonary artery (UAPA) is an exceedingly rare congenital anomaly, often discovered incidentally or during evaluation for respiratory symptoms. The absence of one pulmonary artery, particularly the right-sided variant, poses unique challenges in diagnosis and management due to its infrequent occurrence and diverse clinical manifestations. Here, we present a case of unilateral absence of the right pulmonary trunk in a 40-year-old male presenting with breathlessness and chronic cough, along with a comprehensive radiological description. This case highlights the importance of recognizing and understanding such rare anatomical variations to guide appropriate clinical management. Congenital pulmonary artery anomalies, including UAPA, are thought to arise from disturbances during embryogenesis, specifically during the complex process of septation and development of the truncus arteriosus. While the exact etiology remains elusive, proposed mechanisms include abnormal development of the sixth aortic arch, persistence of the ductus arteriosus, or failure of the primitive pulmonary plexus to form. The incidence of UAPA is estimated to be approximately 1 in 200, 000 live births, with a slight predilection for the right pulmonary artery absence compared to the left.

Clinical presentation of UAPA varies widely, ranging from asymptomatic individuals to those with severe respiratory compromise, exercise intolerance, or hemoptysis. The absence of symptoms in some cases may delay diagnosis until adulthood, as was observed in our patient. Diagnostic modalities such as transthoracic echocardiography (TTE), magnetic resonance imaging (MRI), or computed tomography angiography (CTA) play crucial roles in confirming the diagnosis and delineating associated abnormalities.

Radiological findings in UAPA typically demonstrate ipsilateral lung hypoplasia or aplasia,

compensatory hyperinflation of the contralateral lung, and hypertrophy of the ipsilateral bronchial and pulmonary arteries. Collateral circulation, often via systemic-to-pulmonary arterial collaterals, is a hallmark feature, contributing to the preservation of pulmonary perfusion to the affected lung.



**Fig 1:** CECT thorax showing isolated unilateral left pulmonary trunk (Blue arrow) with absence of right pulmonary artery. Main pulmonary trunk (Star asterisk)



**Fig 2:** CECT thorax showing multiple hypervascular right-sided collateral vasculature (Blue arrow) and on VRT image vessels arising from abdominal aorta extending and supplying right lung.



**Fig 3:** CECT thorax showing architectural distortion of the right lung with irregular reticulations and septal thickening with mild microcystic honeycombing showing apico-basal gradient – features of interstitial lung disease.

## Discussion

UAPA, a rare condition, is estimated to affect approximately 1 in 200,000 young adults [1]. Typically, it presents alongside cardiovascular anomalies like tetralogy of Fallot or cardiac septal defects, although it can manifest independently as well [2]. In cases of isolated UAPA, the right lung is implicated in roughly two-thirds of instances. Despite common occurrences on the side opposite the aortic arch due to embryologic relations, our patient deviated from this trend [3, 4].

The precise embryologic mechanism behind UAPA remains debated, varying between left- and right-sided occurrences. Nonetheless, it is generally understood that abnormal development of the sixth aortic arch segment leads to a pulmonary artery deriving from the ductal tissue, which regresses at birth, resulting in the proximal interruption of the vessel [4]. Despite this interruption, distal intrapulmonary branches of the affected artery often persist, nourished by collateral vessels stemming from bronchial, intercostal,

internal mammary, subdiaphragmatic, subclavian, or even coronary arteries [5, 6].

In this report, we describe the clinical presentation, radiological findings, and management approach in a rare case of unilateral absence of the right pulmonary trunk in an adult male. By elucidating the clinical course and challenges encountered in this patient, we aim to contribute to the understanding of this uncommon anomaly and its implications for patient care.

## Conclusion

Unilateral absence of the pulmonary artery (UAPA) is a rare congenital anomaly associated with diverse clinical presentations and diagnostic challenges. Through the presented case of a 40-year-old male with UAPA and associated respiratory symptoms, we highlight the importance of recognizing and understanding such rare anatomical variations to guide appropriate clinical management. High-resolution computed tomography (HRCT) plays a crucial role in confirming the diagnosis and delineating associated abnormalities, including compensatory hypertrophy of the contralateral pulmonary artery, systemic-to-pulmonary arterial collaterals, and associated lung parenchymal changes.

Management of UAPA is tailored to the individual patient, considering the severity of symptoms, associated anomalies, and potential complications. While asymptomatic cases may require conservative management and regular monitoring, symptomatic individuals may benefit from surgical interventions to alleviate respiratory symptoms and optimize pulmonary perfusion. Continued research and clinical vigilance are essential to further elucidate the pathophysiology and optimal management strategies for UAPA, ultimately improving patient outcomes and quality of life.

## Conflict of Interest

Not available.

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Not available.

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