

# Agnesia of Right Pulmonary Artery Associated with High Output Coronary Fistula for Superior Vena Cava and Intrapulmonary Artery Branches: Case Report

Aline Valério de Lima, Stella Souza Carneiro, José Augusto Murad, José Ayrtton Arruda, José Guilherme Caselli, Edson Egashira

Hospital UNIMED Vitória,<sup>1</sup> Vitória, ES – Brazil; Hospital Universitário Cassiano Antonio Moraes,<sup>2</sup> Vitória, ES – Brazil

## Introduction

Unilateral Pulmonary Artery Agnesia (UPAA) is a rare congenital anomaly that occurs due to malformation of the sixth aortic arch of the affected side during embryogenesis, and may occur in isolation or in combination with other cardiovascular anomalies.<sup>1,2</sup> Diagnosis usually occurs in adolescence, but individuals may be asymptomatic and receive late diagnosis. We report a case of symptomatic patient with right pulmonary artery agnesia accompanied by high-output fistula of the Circumflex Artery (CXA) to the Superior Vena Cava (SVC).

## Case report

Female patient, 60 years old, hypertensive, with pulmonary fibrosis, bronchiectasis and anxiety disorder. She was admitted to the emergency room reporting typical precordial pain, hemodynamically stable, with no ischemic disorders on electrocardiogram and negative myocardial necrosis markers. Chest X-ray showed only right lung volume reduction (Figure 1). She reported previous episodes of a similar condition and undergone coronary angiography 2 years before in another hospital. A previous test suggested CXA high-output coronary fistula to intrapulmonary artery branches and SVC (Figure 2).

Computed tomography angiography was performed to investigate the coronary artery and lungs, revealing CXA of great anatomical importance, originating two marginal arteries, without obstructive lesions, with anomaly characterized by coronary fistula originating in its proximal third and retroaortic path flowing into the SVC, and right lung with volume reduction and left buffalo chest (Figure 3). Absence of right pulmonary artery was found, and pulmonary irrigation originated in the collaterals from the aorta and its branches. Myocardial scintigraphy did not demonstrate ischemia and echocardiogram was also normal. The patient remained stable while in hospital and clinical treatment was chosen.

## Keywords

Pulmonary Artery Agnesia; Fistula; Ischemia.

**Mailing Address:** Aline Valério de Lima •  
Rua Carijós, 625, apto. 114, Jardim da Penha. CEP 29060-700, Vitória,  
ES – Brazil  
E-mail: alinevalerio1@hotmail.com

**DOI:** 10.5935/2318-8219.20190026

## Discussion

The first reported case of UPAA was published in 1868 by Frantzel. Since then, about 420 cases have been reported.<sup>1,2</sup> Its prevalence is around 1:200,000 individuals, with no gender differences. UPAA occurs due to malformation during embryogenesis, with persistence of the pulmonary artery connection to the sixth distal aortic arch, where the ductus arteriosus connects to the primitive dorsal aorta.<sup>1,3</sup> Some studies point out a relationship between absence of pulmonary artery with absence of the ipsilateral ductus arteriosus, obtained in radiological, surgical or post-mortem documentation.<sup>3</sup> Transient systemic-pulmonary collateral arteries may arise over two long periods, even during initial embryonic development, and remain if there is obstruction in the pulmonary outflow tract.<sup>3</sup> When this obstruction occurs at a very late stage of fetal development or after birth, the bronchial arteries may turn into systemic-pulmonary collateral arteries. These collaterals, in turn, arise mainly from the bronchial arteries but have also been documented as arising from other arteries, such as the coronary arteries.<sup>3</sup> It has been shown that in congenital heart diseases some aortopulmonary collateral arteries have a marked histological similarity to the ductus arteriosus.<sup>3</sup>

Patients with UPAA have a normal pulmonary trunk and unilateral absence of one branch of the pulmonary artery,<sup>4,5</sup> with the right side being affected in two thirds of the cases.<sup>1,2</sup> Intrapulmonary vasculature and the distal portion of the trunk can develop normally and receive vascularization of bronchial vessels,<sup>4,5</sup> resulting in small and hypovascular lung on the affected side. In about 4% of the cases, communication between coronary and bronchial arteries is present.<sup>6-8</sup>

The disease presents clinically in several ways, but the most common manifestations are contralateral pulmonary hypertension, present in about 25% of the cases, determining long-term survival, and hemoptysis.<sup>2,9</sup> UPAA can still remain asymptomatic in about of 30% of patients.<sup>1,2</sup> The most common causes of death include right heart failure, respiratory failure, massive hemoptysis hemorrhage and pulmonary edema. The gold standard for diagnosis is digital subtraction angiography, but because it is an invasive test, it is reserved for cases of hemoptysis embolization or coronary artery bypass grafting, and diagnosis is established through other imaging tests such as computed tomography angiography of the lung, which may reveal, in addition to vascular disorders, parenchymal findings, such as bronchiectasis and mosaic attenuation pattern, possibly caused by the increased perfusion of the affected lung, besides the development of pulmonary hypertension.<sup>2,10</sup> The presence of associated malformations can also be well demonstrated by tomography.<sup>2</sup>

Treatment includes surgical, pharmacological and behavioral

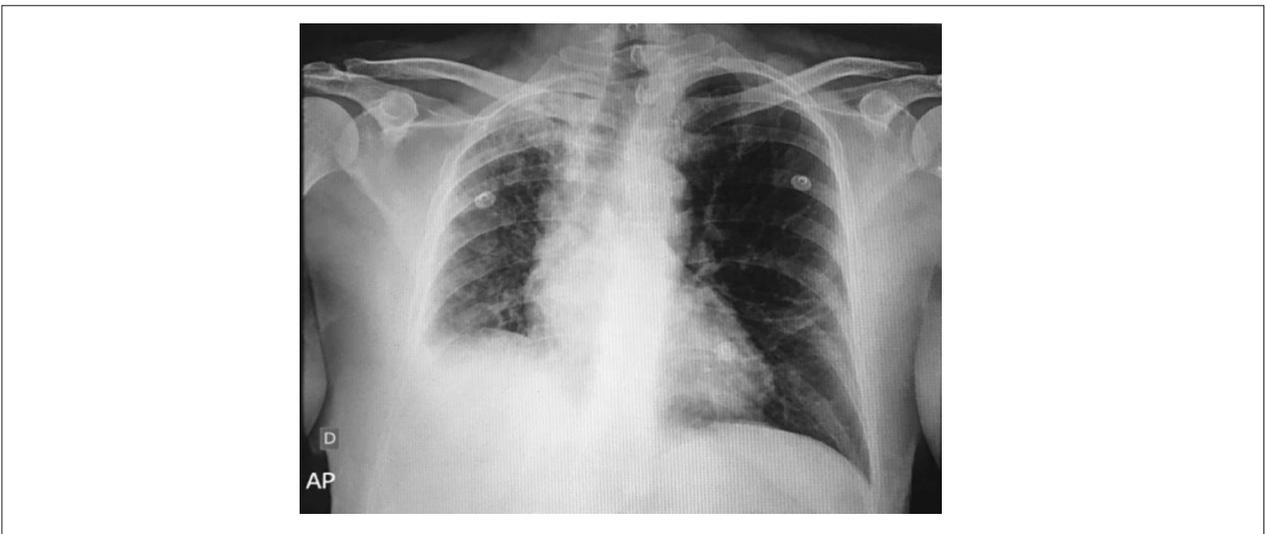


Figure 1 – Chest X-ray showing right lung volume reduction.

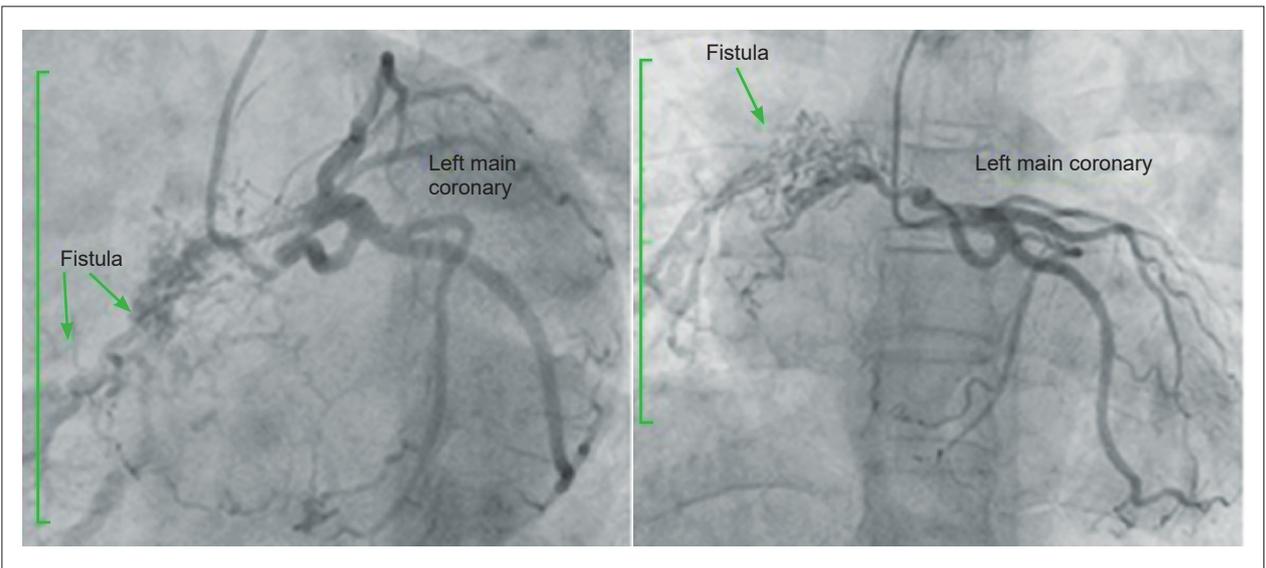


Figure 2 – Coronary angiography suggesting high-output fistula to the intrapulmonary artery branches.

management, with pneumonectomy and/or coronary artery bypass grafting being considered in cases of hemoptysis, pulmonary infections and pulmonary hypertension, and pharmacological measures are recommended only for refractory patients or those that cannot be operated.<sup>2</sup> Asymptomatic patients should undergo serial echocardiography to monitor the development of pulmonary hypertension, which represents a sign of worse prognosis.

### Conclusion

unilateral pulmonary artery agenesis is a rare congenital anomaly that can go asymptomatic for many years, leading to late diagnosis in many cases, despite the various imaging options that may contribute to the investigation.

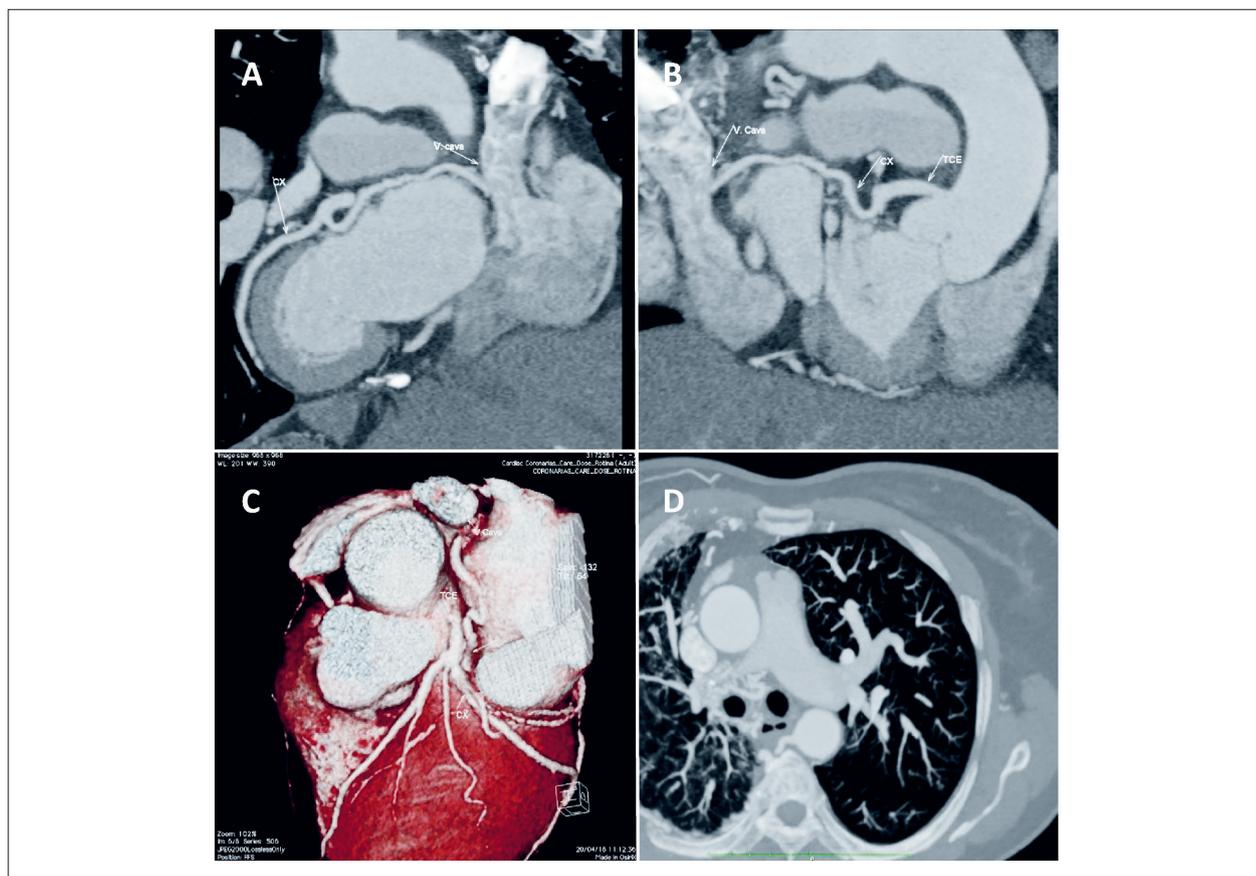
Because it is a rare disease, clinicians should keep in mind the possibility of unilateral pulmonary artery agenesis undiagnosed in adulthood.

### Authors' contributions

Data acquisition: Lima AV, Carneiro SS, Arruda JA, Caselli JG, Egashira E; Data analysis and interpretation: Arruda JA, Caselli JG; Manuscript writing: LimaAV; Critical revision of the manuscript for important intellectual content: LimaAV, Carneiro SS, Murad JA, Arruda JA.

### Potential conflict of interest

The authors declare that there is no relevant conflict of interest.



**Figure 3** – Coronary and pulmonary artery computed tomography angiography. (A and B) Multiplanar reconstruction showing the path of the circumflex fistula - superior vena cava. (C) Three-dimensional reconstruction of the coronary arteries. (D) Axial view at the pulmonary trunk bifurcation level, showing the unilateral pulmonary artery on the right.

## References

1. Saladi L, Roy S, Diaz-Fuentes G. Unilateral pulmonary artery agenesis: An unusual cause of unilateral ARDS. *Respir Med Case Rep.* 2018; 23:148-151
2. Steiropoulos P, Archontogeorgis K, Tzouveleakis A, Ntoliou P, Chatzistefanou A, Bouros D. Unilateral pulmonary artery agenesis: a case series. *Hippokratia.* 2013;17(1):73-6.
3. Adán Lanceta V, Jiménez Olmos A, Martín de Vicente C, García Íñiguez JP. Agenesis aislada de la arteria pulmonar derecha. *An Pediatr (Barc).* 2017 ;86(1):45-46.
4. Nana-Sinkam P, Bost TW, Sippel JM. Unilateral pulmonary edema in a 29-year-old man visiting high altitude. *Chest.* 2002;122(6):2230-3.
5. Vergauwen S, Bracke P, De Schepper A. Unilateral absence of a pulmonary artery. *J Belge Radiol.* 1998;81(5):254.
6. Bockeria LA, Makhachev OA, Khiriev TKh, Abramyan MA. Congenital isolated unilateral absence of pulmonary artery and variants of collateral blood supply of the ipsilateral lung. *Interact Cardiovasc Thorac Surg.* 2011;12(3):509-10.
7. Heper G, Korkmaz ME. High-pressure pulmonary artery aneurysm and unilateral pulmonary artery agenesis in an adult. *Tex Heart Inst J.* 2007;34(4):425-30.
8. De Dominicis F, Leborgne L, Raymond A, Berna P. Right pulmonary artery agenesis and coronary-to-bronchial artery aneurysm. *Interact Cardiovasc Thorac Surg.* 2011;12(3):507-9.
9. Atik E, Tanamati C, Kajita L, Barbero-Marcial M. Agenesis isolada da artéria pulmonar direita ou esquerda: avaliação da evolução natural e a longo prazo, após intervenção corretiva. *Arq Bras Cardiol.* 2006;87(4):423-8.
10. Sakai S, Murayama S, Soeda H, Furuya A, Ono M, Ro T, Akamine T, et al. Unilateral proximal interruption of the pulmonary artery in adults: CT findings in eight patients. *J Comput Assist Tomogr.* 2002;26(5):777-83.