

# Compression of Left Main Coronary Artery in Patients with Pulmonary Artery Aneurysm and Pulmonary Hypertension

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## Abstract

**Background:** Pulmonary artery aneurysm (PAA) is an unusual finding and its association with left main coronary (LMCA) compression is even more infrequent. Cardiac CT evaluates of presence and size of PAA and the degree of LMCA compression. The aim of this study is to describe two cases of adults with compression of LMCA with PAA associated with PDA and pulmonary hypertension. **Case presentation:** The first case is a 27-year-old man with PAA (78 mm diameter) and LMCA compression of 70% between the aortic sinus and the PAA. He presented angina as a manifestation of the LMCA compression. During follow-up the patient died. The second case is a 28-year-old man with PAA (110 mm diameter) that compresses LMCA in 55%, he rejected surgical treatment, but he is in close follow-up with medical treatment. **Conclusion:** Cardiac computed tomography played an important role both in the diagnosis and identification of high-risk PAA patients.

## Keywords

Left Main Coronary Artery Compression, Pulmonary Artery Aneurysm, Patent Ductus Arteriosus, Pulmonary Hypertension

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## 1. Introduction

Pulmonary artery aneurysm (PAA), is defined as a pulmonary artery (PA) di-

ameter over 40 mm, or dilatation of at least 1.5 times the normal dimension [1]. PAA is an unusual finding with a prevalence of 1 in 14,000 individuals. However, infrequent but fatal complications such as dissection may occur [2].

The patent ductus arteriosus (PDA), ventricular and atrial septal defects are the congenital anomalies more often associated with PAA [3]. When PAA is associated with PDA, the left to right shunt strikes the wall of the pulmonary artery (PA), causing a progressive weakening of the PA wall [3].

Previous studies have shown that compression of the left main coronary artery (LMCA) combined with pulmonary hypertension was strongly associated with sudden death, but this does not occur with the isolated dilatation of the pulmonary artery [1] [2].

The echocardiogram is the non-invasive imaging method of choice in the diagnosis of congenital heart disease, but this technique is limited in the assessment of extracardiac structures [4].

Cardiac computed tomography (Cardiac CT) plays an important role in the diagnosis and identification of high-risk PAA [1]. The LMCA compression > 50%, the angle < 30° between LMCA with the left sinus of Valsalva and the main PA/aorta diameter ratio > 2, significantly increase the probability of myocardial ischemia [5].

The aim of this study is to describe two cases of adults with compression of LMCA in patients with PAA associated with PDA and pulmonary hypertension.

## 2. Patient 1

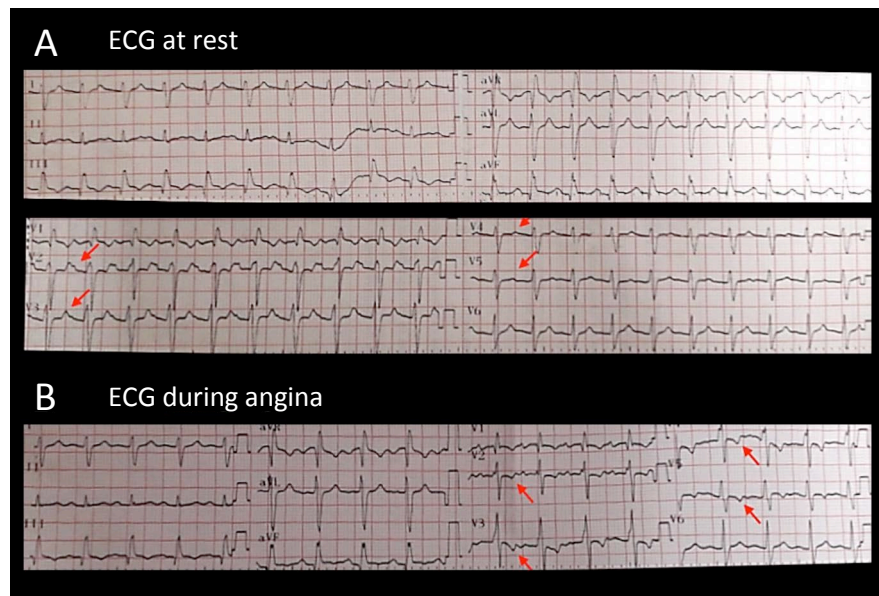
Twenty-seven years-old man with progressive dyspnea, angina and palpitations from 1 year ago.

On physical examination increased intensity of the pulmonary component of the second sound, and tricuspid regurgitation murmur II/VI was heard.

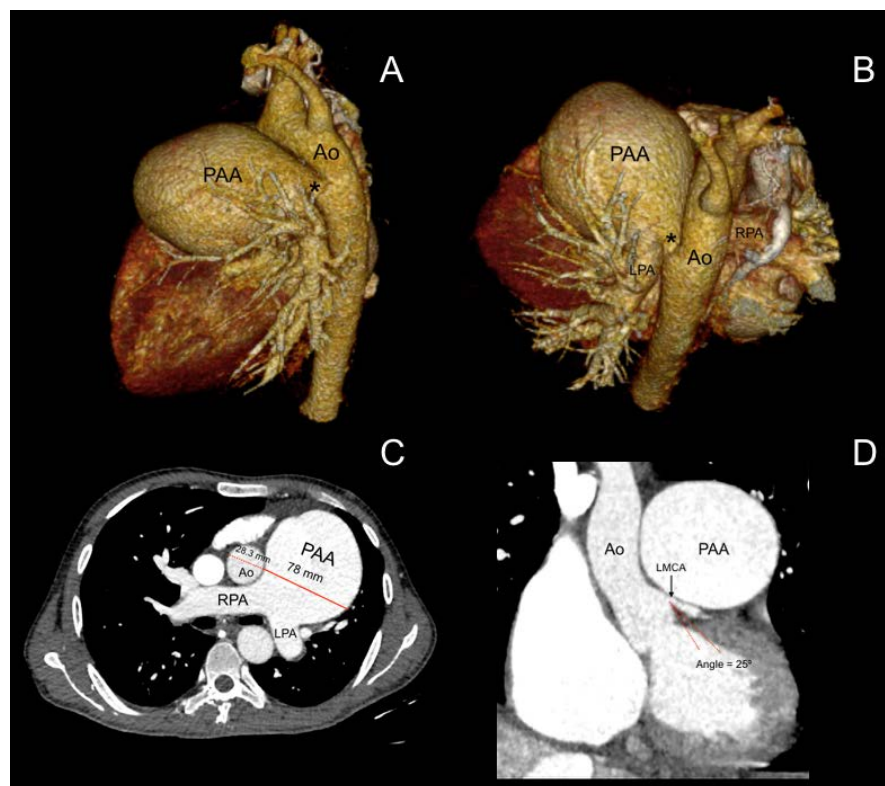
The electrocardiogram (ECG) showed T-wave inversion during angina compared to resting ECG, **Figure 1**. Transthoracic echocardiogram demonstrated PDA with bidirectional shunt (Eisenmenger syndrome), PAA, diastolic basal diameter of right ventricle of 36.3 mm, right atrium area of 31 cm<sup>2</sup>, TAPSE of 22 mm, severe tricuspid and pulmonary regurgitation, and severe pulmonary hypertension with systolic pulmonary artery pressure of 97 mmHg (**Table 1**).

Cardiac CT confirmed the presence of PDA type C of Krichenko with length of 12 mm, aortic side 11 mm, pulmonary side 8.5 mm, PAA with 78 mm diameter, right pulmonary artery with 23 mm and left pulmonary artery with 20 mm diameters, and dilatation of right cavities. Also, Cardiac CT showed LMCA compression of 70% between the aortic sinus and the PAA, and the demonstrated a downward angulation between the LMCA and the left sinus of Valsalva was of 25°. A main PA/aorta diameter ratio was of 2.75 (**Figure 2, Table 1**). The patient was treated with furosemide and enalapril.

During follow-up the patient died. The cause of death was cardiogenic shock probably due to severe pulmonary hypertension.



**Figure 1.** (A) Surface electrocardiogram (ECG) of 12 leads at rest; (B) ECG during angina, with T-wave inversion in precordial leads (red arrows).



**Figure 2.** (A) (B) CT Volume rendering reconstruction showing patent ductus arteriosus (\*). (C) Multiplanar reformatted (MPR) Cardiac CT showing PAA. Main pulmonary artery/aorta ratio was 2.75. (D) MPR PAA that compresses left main coronary artery. The angle between the left main coronary artery and left sinus of Valsalva is of 25° (normal subjects is of 90°). Ao: Aorta. LAD: Left anterior descending artery. LMCA: Left main coronary artery. PAA: Pulmonary artery aneurysm. RPA: Right pulmonary artery. LPA: Left pulmonary artery.

**Table 1.** Baseline characteristics, measurements of PAA, systolic pulmonary arterial pressure, degree of left main coronary artery compression and its angulation relative to the left sinus of Valsalva.

	Patient 1	Patient 2
Age (years)	27	28
Gender	Male	Male
Main symptom	Dyspnea, angina	Dyspnea
PA diameter (mm)	78	110
Pulmonary trunk/aortic diameter ratio	2.75	3.33
Degree of LMCA compression	70%	55%
Angle between the LMCA and left sinus of Valsalva	25°	15°
sPAP (mmHg)	97	82
Pulmonary regurgitation	Severe	Moderate
Outcome	Fatal	Alive

Abbreviations: LMCA: Left main coronary artery; sPAP: systolic pulmonary arterial pressure; PA: Pulmonary artery.

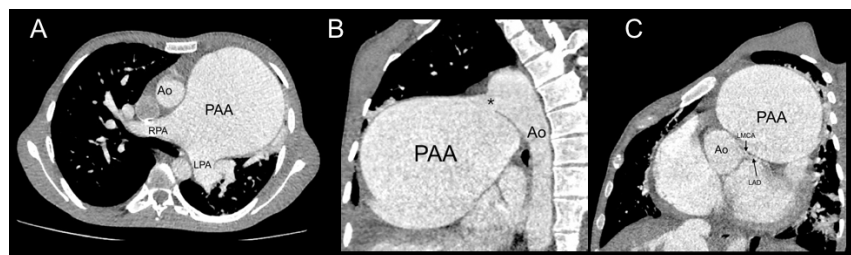
### 3. Patient 2

Twenty-eight years-old man with progressive dyspnea from two years ago.

On physical examination increased intensity of the pulmonary component of the second sound, and systolic tricuspid murmur II/VI was heard.

The ECG in sinus rhythm, without signs of ischemia at rest or physical exertion. Echocardiography demonstrated PDA with shunt left-right, PAA, right atrium area of 35.2 cm<sup>2</sup>, left atrium area of 52 cm<sup>2</sup>, TAPSE of 18 mm, severe pulmonary regurgitation, moderate tricuspid regurgitation and severe pulmonary hypertension with systolic pulmonary artery pressure of 82 mmHg.

Cardiac CT confirmed PDA type C of Krichenko with length of 10 mm, aortic side 20 mm and pulmonary side 22 mm, and PAA 110 mm diameter that compresses LMCA. The right pulmonary artery measured 25.2 mm and the left pulmonary artery 24.2 mm. The LMCA compression was of 55% between the aortic sinus and the PAA, the downward angulation of the LMCA with the left sinus of Valsalva was 15°, and the main PA/aorta diameter ratio 3.33 (**Figure 3, Table 1**).



**Figure 3.** Multiplanar reformatted (MPR) Cardiac CT images. (A) PAA of 110 mm and its bifurcation on right pulmonary artery of 25.2 mm and left pulmonary artery of 24.2 mm. (B) PDA (\*). (C) Compression of the left main coronary artery by PAA. Ao: Aorta. LAD: Left anterior descending artery. LMCA: Left main coronary artery. PAA: Pulmonary artery aneurysm. RPA: Right pulmonary artery. LPA: Left pulmonary artery.

The patient rejected surgical treatment, but he is in close follow-up with medical treatment.

#### 4. Discussion

PAA is infrequent, so there are not many studies or clear guidelines on its stratification of risk and management.

Echocardiography plays a very important role in the diagnosis of congenital heart disease, but this technique is limited in the assessment of extracardiac structures.

Cardiac CT evaluates of presence and size of PAA, detection of complications as compression of coronary arteries, airway and nerve recurrent laryngeal.

Also, the cardiac CT allows to evaluate the degree of LMCA compression > 50% and its angle with the left sinus of Valsalva, and the main PA/aorta diameter ratio. These data are important because they increase the risk of myocardial ischemia [5]. Only the first patient presented angina as a manifestation of the LMCA compression.

The severe pulmonary hypertension is an important factor in the prognosis of these patients, since it has been observed that unexpected deaths and dissection are more frequent in the context of congenital heart disease [2].

Previous studies have shown that LMCA compression and sudden death were strongly associated with pulmonary hypertension [1] [2].

The incidence of LMCA compression due to PA enlargement and pulmonary hypertension according to some series of cases ranges from 5% to 44% [6].

Patients with Eisenmenger syndrome have an increased risk of rupture and dissection, laminated thrombus can also develop in the dilated central pulmonary arteries and thrombosis *in situ* in the distal pulmonary arteries [5].

The isolated PAA is not associated with adverse events [1], the patient 2 had a PAA diameter greater than patient 1 (110 mm versus 78 mm), but the latter had a worse evolution because the compression of the LMCA was greater (55% vs. 70%) and the ECG showed inversion of T wave during angina. Gallego P, *et al.* [1] reported a similar case of LMCA compression due to PAA (48 mm), in a patient with Eisenmenger syndrome who had a sudden death attributed to PA dissection.

The early diagnosis and identification of high-risk PAA patients, is of great importance, because allows better selection of patients for surgical treatment.

Both cases presented high-risk factors for PAA such as an extrinsic compression of the LMCA > 50%, angle < 30° between LMCA with left sinus of Valsalva and pulmonary hypertension.

#### 5. Conclusion

The prognosis of LMCA compression is worse when PAA is associated with PDA and pulmonary hypertension. Cardio CT plays an important role in the diagnosis and identification of PAA high-risk patients.

## Highlights

Cardiac CT is a useful tool to establish the diagnosis of extrinsic LMCA compression and allows the simultaneous evaluation of PAA diameter, the luminal diameter of the LMCA and the angle of LMCA that emerges from the aortic root.

## Author Contribution

DIKT made the concept/design, data, interpretation of the CT images and draft the article. ZIRU worked in the data collection of the patients and made the draft of manuscript. MJS interpreted the CT images and made a critical revision of article. NEZ made a critical revision of article, selected the images and approved the article.

## Conflicts of Interest

No conflict of interest between the authors.

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## Appendix

**Patient Identification Number: 1**

**CONSENT FORM**

**Title of Study: Compression of left main coronary artery in patients with pulmonary artery aneurysm and pulmonary hypertension**

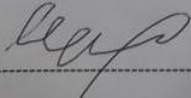
**Name of Researchers: Diana Isabel Katekaru-Tokeshi, Zoila Ivonne Rodriguez-Urteaga, Moises Jimenez-Santos, Nilda Espinola-Zavaleta.**

1. I confirm that I have understand the information about the study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily.

2. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected.

3. I understand that relevant sections of my medical notes and data collected during the study may be looked at by members of the research team, where it is relevant to my taking part in this research. I give permission for these researchers to have access to my records.

4. I agree to take part in the study.

  
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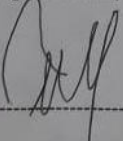
Patient Identification Number: 2

CONSENT FORM

Title of Study: Compression of left main coronary artery in patients with pulmonary artery aneurysm and pulmonary hypertension

Name of Researchers: Diana Isabel Katekaru-Tokeshi, Zoila Ivonne Rodriguez-Urteaga, Moises Jimenez-Santos, Nilda Espinola-Zavaleta.

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