

# Association of aortic coarctation and malignant right coronary artery anomaly: an unusual cause of angina pectoris

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**Abstract:** A 53-year-old man with exercise-induced ischemia was referred for investigation. Coronary angiography revealed a right coronary artery originating from the trunk of the left coronary artery, and an aortic coarctation was observed on aortography. A CT angiogram confirmed these findings. Resection of the aortic coarctation and reimplantation of the ostial portion of right coronary artery into its native site was performed, and resulted in a satisfactory outcome. The association of an anomalous right coronary artery with aortic coarctation has rarely been described and represents a critical situation where early diagnosis and prompt intervention are essential.

**Keywords:** coronary anomalies, single ostium, myocardial ischemia, angina pectoris

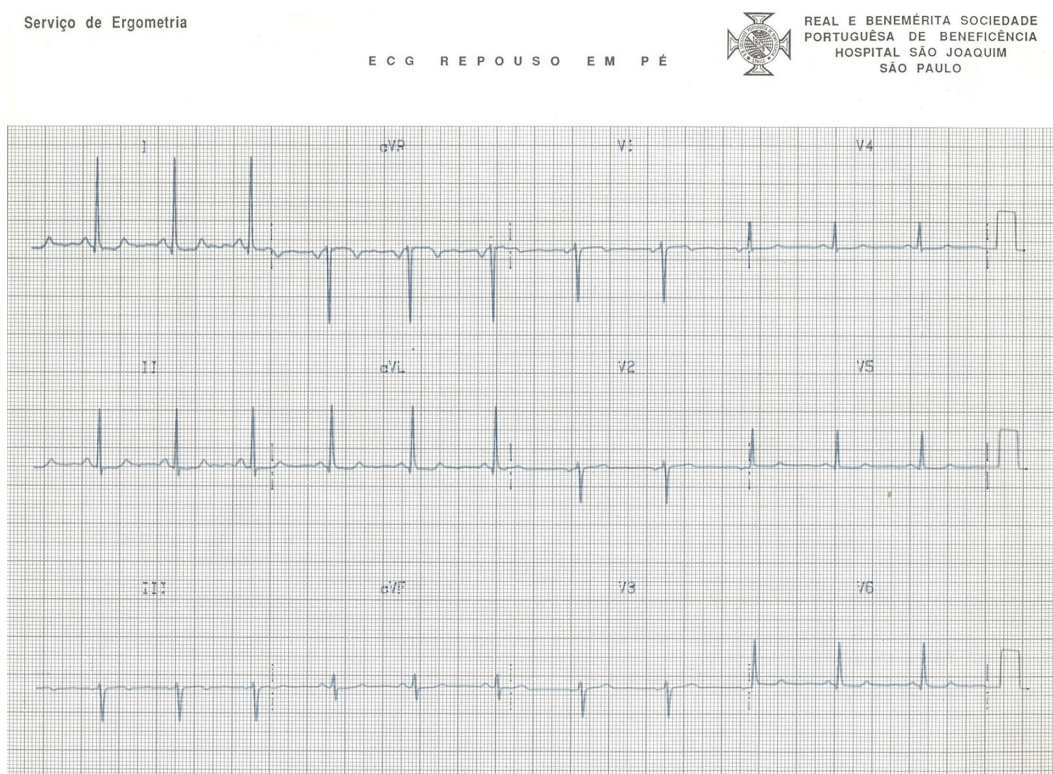
## Introduction

A single coronary ostium is an entity uncommonly observed, accounting for only about 24% of coronary anomalies.<sup>1</sup> Its prevalence on the general population is around 1.3%, and represents quite a rare finding in cineangiographic exams,<sup>1</sup> with incidence rates ranging from 0.0024% to 0.066%. The vast majority of cases are discovered accidentally on routine investigation or on risk stratification tests requested for patients with suspected coronary artery disease.<sup>2</sup> These variations may be associated with other cardiac defects, such as valvulopathies and congenital heart diseases (tetralogy of Fallot, aortic coarctation, and transposition of the great vessels).<sup>3</sup> A single coronary artery is amongst the rarest anomalies (8.8%), and deserves special attention because any kind of alteration that predisposes to a reduced blood flow in this vessel can ultimately result in more extensive myocardial damage.<sup>4</sup> Definitive diagnosis can be confirmed by angiography and computed tomography (CT) angiography, the latter being responsible for the identification of the anatomic coronary course related to the great vessels of the heart. The interarterial course, defined as the passage of the coronary artery between the aorta and the pulmonary artery, is associated with sudden death during or after exertion, mainly in young patients.<sup>5</sup> Here we report the rare association of a right coronary artery anomaly, exercise-induced ischemia, and aortic coarctation, in a scenario where early diagnosis was made and the appropriate surgical procedure resulted in a successful outcome.

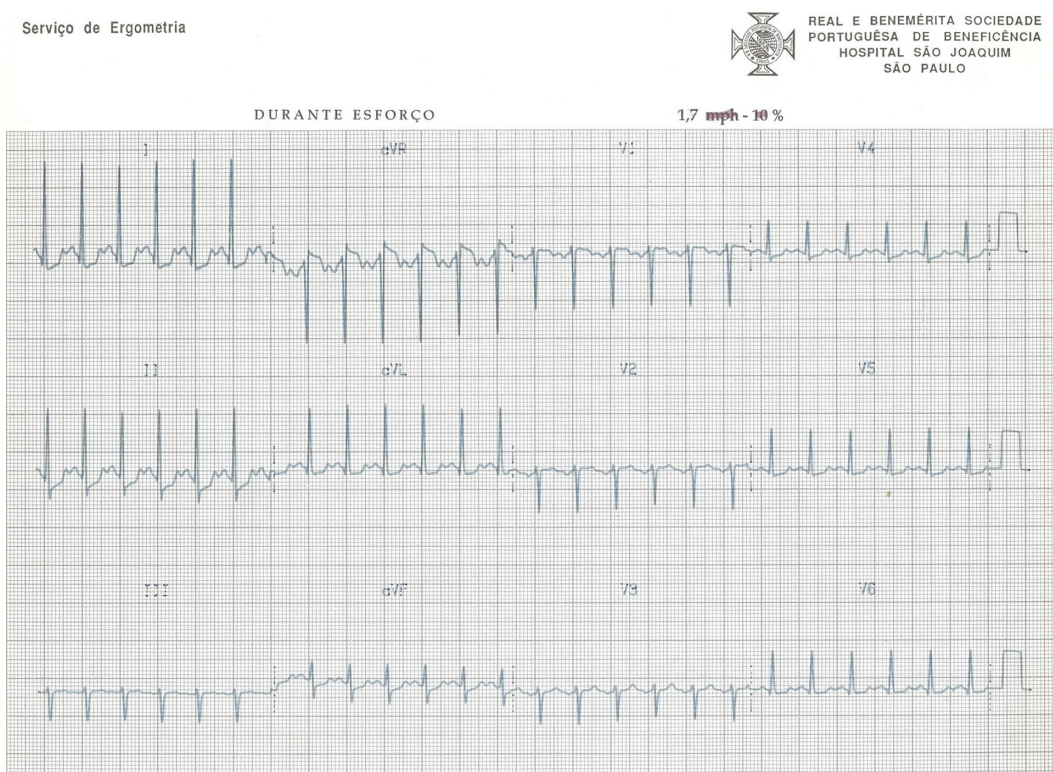
## Case report

A 53-year-old man with a history of hypertension, dyslipidemia, and stable angina CCS 2 underwent an exercise tolerance test which was positive for stress-induced ischemia (Figure 1A and 1B). Based on this result, the patient was referred to our

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**Figure 1A** Resting electrocardiogram showing no abnormalities.

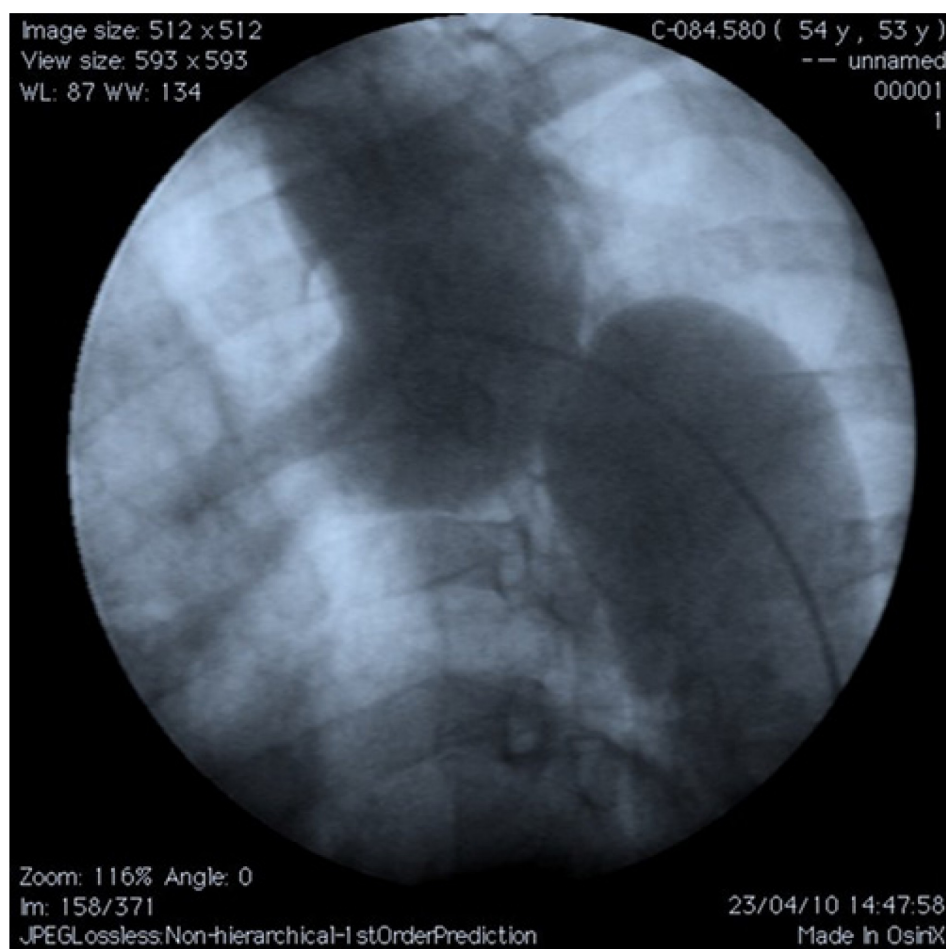


**Figure 1B** Electrocardiogram showing ST segment depression mainly in leads I, II, aVL, and aVF.

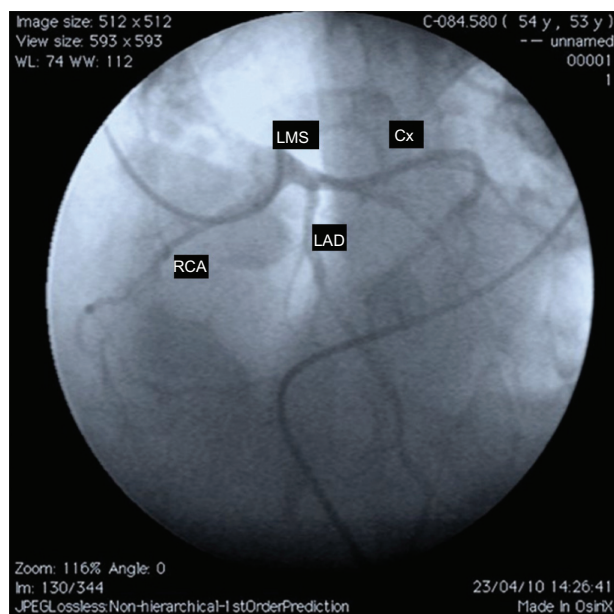


service for further investigation. Physical examination was unremarkable, with no dyspnea, lung congestion, signs of increased venous pressure, gallops, murmurs, or rubs. His heart rate was 87 beats per minute and blood pressure was stable at 110/70 mmHg. The ankle-brachial index in his right leg was 0.76. His daily medications included aspirin 200 mg, atenolol 50 mg, and atorvastatin 20 mg. An electrocardiogram on admission demonstrated sinus rhythm, with a PR interval of 0.12 seconds, a QRS interval of 0.10 seconds, AQRS +30, with no repolarization abnormalities or evidence of injury. The chest X-ray showed widening of the aortic shadow. Laboratory investigations showed a normal full blood count, and normal liver and kidney tests. The patient underwent coronary angiography through the right femoral artery, which revealed aortic coarctation (Figure 2) and a right dominance pattern, with an abnormal right coronary origin, arising from the left coronary artery branch, with no obstructive lesions (Figure 3). No extrinsic compression was observed and the remaining coronary arteries showed

no evidence of relevant atherosclerotic obstruction. Left ventricular function was preserved, with no segmental hypokinesia. Although coronary angiography did not show any significant obstructive lesions, a CT coronary angiogram (using a clinical scanner with 64 data channels) was performed, which revealed an anomalous right coronary course, passing between the aorta and the exit tract of the right ventricle (interarterial, Figure 4), and confirmed the aortic coarctation (Figure 5). A transthoracic echocardiogram showed no further malformations. Based on the positive findings of the exercise test and imaging studies which revealed the malignant course of the right coronary artery, as well as the aortic coarctation, surgical correction of the aortic aneurysm (resection with end-to-end anastomosis) and reimplantation of the coronary ostium were indicated. The surgical procedure was performed successfully with no complications. The patient was discharged from hospital on the fifth postoperative day taking atorvastatin 20 mg/day and atenolol 25 mg twice daily. Three months after the



**Figure 2** Angiogram demonstrating aortic coarctation.



**Figure 3** Coronary angiography revealing a single coronary ostium. The right coronary arises from the left coronary main branch.

**Abbreviations:** LMS, left main stem; Cx, circumflex artery; RCA, right coronary artery; LAD, left anterior descending artery.

procedure, the patient resumed his normal daily activities with no signs or symptoms of myocardial ischemia.

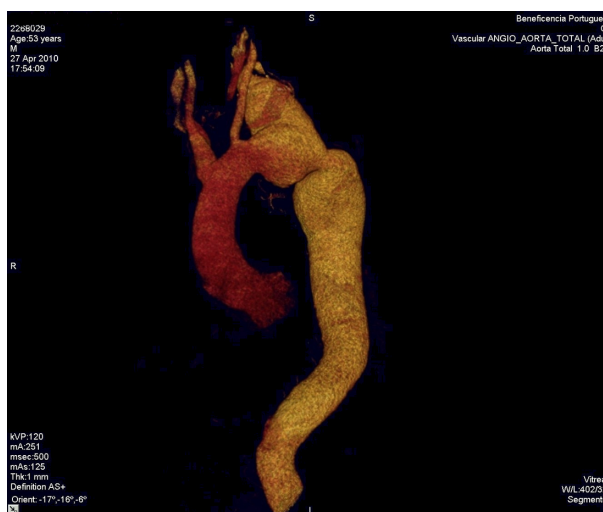
## Discussion

The single coronary ostium is a clinical entity that runs a benign and asymptomatic clinical course in 85% of



**Figure 4** CT angiogram showing the right coronary course between the aorta and pulmonary artery.

**Abbreviations:** LMS, left main stem; Cx, circumflex artery; RCA, right coronary artery; LAD, left anterior descending artery; RC, right coronary.



**Figure 5** CT angiography (three-dimensional reconstruction) confirming the aortic isthmus coarctation.

cases.<sup>6</sup> However, around 15% of individuals may develop myocardial ischemia of varying degrees as a consequence of anomalies of the right coronary artery, which is most commonly diagnosed incidentally on angiography requested to investigate a range of and sometimes nonspecific chest complaints.<sup>2</sup> These anomalies are divided into two major groups, according to whether or not they result in exercise-induced ischemia as a consequence of extrinsic compression arising from their anomalous course between the aorta and the outflow tract of the right ventricle. Reduced blood flow through a compressed artery can result in myocardial ischemia and its clinical manifestations, which range from typical angina to malignant and fatal arrhythmias, sometimes followed by sudden death.<sup>7</sup> The association of coronary anomalies with aortic coarctation is a very rare finding and both left<sup>8</sup> and right<sup>9</sup> coronary anomalies have been described in association with this great vessel malformation. Anatomopathological studies suggest that in individuals who present cardiac ischemia as a result of the anomalous course of the right coronary artery (arising from the contralateral sinus), restriction of blood flow occurs in rare and isolated episodes. The consequential accumulative ischemic damage leads to islands of necrotic tissue and myocardial fibrosis, paving grounds to electrical instability and potentially lethal high-rate arrhythmias.<sup>5</sup> Possible ischemia-related factors are the ostium slit-shape formed by the origin of the anomalous vessel, which can be compressed during ventricular systole, as well as compression of the anomalous vessel that runs through the aorta and the pulmonary artery, especially during physical exertion, when these two major vessels dilate.

The role of extrinsic compression by these major vessels is still elusive, as long as the hydrostatic pressure within the pulmonary artery is lower than the coronary perfusion pressure.<sup>2</sup> Studies with intracoronary ultrasound were quite consistent in demonstrating that in this kind of anomaly. The role of the extrinsic compression by these major vessels is still a matter of debate. Although the hydrostatic pressure within the pulmonary artery is lower than the coronary perfusion pressure,<sup>2</sup> studies with intracoronary ultrasound were quite consistent in demonstrating that in this kind of anomaly, the ectopic coronary artery follows, right after its origin, a transmural course, which is the site of greatest proximity between the aorta and the pulmonary artery. In this intramural segment, the external and internal coronary walls are thin and delicate, predisposing to compression of the vessel lumen in pace with the cardiac cycle, with phasic exacerbation during ventricular systole. Individual variances with respect to systolic compression may explain the different clinical and prognostic behaviors observed amongst subjects presenting with this condition. In this intramural segment, the external and internal aortic walls are thin and delicate, predisposing to compression of the vessel lumen in pace with the cardiac cycle, with phasic exacerbation during ventricular systole. Individual variances with respect to systolic compression may explain the different clinical and prognostic patterns observed in subjects presenting with this condition.

In most affected patients, the anomaly follows a benign course, without any major clinical repercussions. Even amongst the sudden death survivors, about 55%–93% describe no evidence of previous signs or symptoms consistent with the disease. About 10% of subjects present with variable degrees of dyspnea, palpitations, dizziness, syncope, and angina. Sudden death typically occurs in young patients during or after episodes of intense physical activity. The remaining symptoms are often observed in young adults and are associated with the development of systemic arterial hypertension, suggesting that stiffness of the aortic wall may represent a protective factor against sudden death in this subgroup of patients.<sup>10</sup>

Once the diagnosis of the anomaly is established, patients should avoid intense and competitive physical activities, and pharmacological therapy that includes a beta-blocker should be initiated promptly.<sup>2,11</sup> Surgical procedures may also be indicated for this type of anomaly. Conventional revascularization, ostium reimplantation on the aortic root, excision of the common wall between the aorta and the anomalous vessel, and the creation of a new ostium at the

end of the intramural segment of the vessel are all plausible alternatives. Martins et al describe a patient with an anomalous origin of the right coronary who underwent myocardial revascularization with right internal thoracic artery grafting to the proximal right coronary artery segment and proximal ligation of the right coronary artery, with a good postoperative outcome.<sup>10</sup> Repair surgery can be obtained by right coronary artery reimplantation, or by using a graft for this coronary.<sup>12</sup> It is necessary to perform a re-evaluation of the treated vessel before the patient is considered able to resume his normal physical activities.<sup>2,11</sup>

## Conclusion

Technological advances in investigative imaging procedures have made the diagnosis of coronary anomalies quite accurate. Once a single ostium-type coronary anomaly is detected, it is mandatory to undertake CT angiographic evaluation of the coronary course. This measurement allows correct identification of patients in the subgroup of potentially malignant interarterial anatomy, who have the worst prognosis. The association of an anomalous right coronary artery with aortic coarctation has rarely been described and is a critical situation where early diagnosis and prompt intervention are essential to avoid serious and even fatal complications.

## Disclosure

The authors report no conflicts of interest in this work.

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