

## CASE REPORTS

### Congenital Absence of Right Coronary Artery without any other Associated Anomalies –A Case Report.

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#### Abstract

A 54 years gentleman presented with central compressive type of chest pain. He was newly diagnosed diabetic, dyslipidaemic. His ECG (electrocardiogram) showed features of anterior and inferior wall ischemia. Echocardiography revealed presence of regional wall motion abnormality. Exercise tolerance test was positive for provokable myocardial ischemia. Angiographic findings includes single coronary artery with congenital absence of RCA (Right coronary artery). Right coronary arterial territory was supplied by the terminal part of left circumflex coronary artery.

**Key Words:** Congenital coronary, Right coronary artery (RCA), Single Coronary Artery.

#### Introduction

Anomalous coronary arteries are congenital cardiac defects with varying symptomatology and controversial clinical significance. These should be considered as a differential diagnosis, particularly when coronary ischemia occurs in a child or young adult. The incidence rate of coronary anomalies in the general population has been estimated to be between 0.2% and 1.3%.<sup>1,2</sup> One of the more serious anomalies involves the right coronary artery (RCA) originating from the left main (LM) coronary artery and coursing between the aorta and pulmonary artery,<sup>3-5</sup> which may result in severe myocardial ischemia due to compression of the artery from the aorta and pulmonary artery.<sup>5-8</sup> Congenital coronary anomalies are the second most common cause of sudden death in young athletes after hypertrophic cardiomyopathy.<sup>9-10</sup>

#### Case Presentation

A 54-year-old hypertensive, newly diagnosed diabetic, dyslipidemic, chronic smoker gentleman was admitted into our hospital with the complaints of central compressive type of chest pain relieved by taking rest or nitroglycerine spray. On examination his pulse was 70 beats/min and regular. He was hemodynamically stable with blood pressure was 120/70 mm Hg and no other abnormality was found in general and systemic examination. His exercise tolerance test (ETT) was done few days back before admission in to our hospital which revealed positive for provokable myocardial ischemia achieved at 7.0 METs .

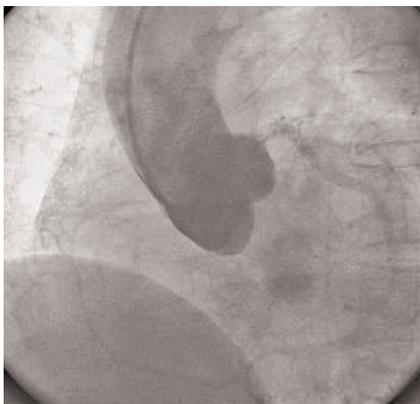
After admission, his electrocardiogram (ECG) showed the feature of anterior- inferior wall ischemia. Echocardiography was done which revealed presence of regional wall

motion abnormality with LVEF - 45%. His lipid profile showed dyslipidaemia with low HDL cholesterol and high LDL cholesterol (150 mg/dl). Considering his typical symptom, and ECG echocardiography and ETT findings, he was sent for conventional coronary angiogram.

#### Evaluation and diagnosis

Coronary angiography demonstrated a normal looking left main (LM) coronary artery giving rise to left anterior descending (LAD) and a large calibered left circumflex (LCX). The left anterior descending artery (LAD) was good sized and totally occluded from its near ostium. Left anterior descending artery (LAD) was lately visible through ipsilateral collaterals. First Diagonal (D1) and second diagonal (D2) were also visible through ipsilateral collaterals. The left circumflex artery (LCX) was dominant and was free of disease. Left circumflex artery (LCX) was being continued as posterior descending artery (LPDA) without significant disease. There was mild discrete eccentric lesion in the distal left circumflex artery (LCX) before origin of the posterior descending artery (PDA). Good sized obtuse marginal (OM) branches were present and free of significant disease. The distal left circumflex artery (LCX) was continued beyond the atrioventricular groove into right coronary artery territory to terminate close to base of the heart. We were unable to find the ostium of the right coronary artery (RCA) using a Judkins Right 3.5 catheter. Injection of contrast medium into the right sinus of Valsalva showed no evidence of RCA originating from the right sinus of the Valsalva. Right coronary artery was congenitally absent which was revealed by root aortogram. Left ventriculography showed apical akinesia with left ventricular ejection fraction of about 45% with LVEDP-35 mm Hg.

So finally the patient was diagnosed as a case of single vessel coronary artery disease and congenital absence of right coronary artery and recommended for revascularization of LAD. (Figures 1 to 3).



**Figure 1.** Aortogram showing absence of origin of right coronary artery.



**Figure 2.** Coronary angiogram of left coronary artery showing continuation of distal left circumflex artery in right coronary artery territory.



**Figure 3.** Coronary angiogram showing distal left circumflex artery in right coronary artery territory.

## Discussion

Coronary anomalies are relatively common, with a reported incidence of 0.2% to 1.3% in the general population.<sup>1,2,11</sup> Most anomalies are clinically insignificant; however, some anomalies have been associated with adverse outcomes like angina, dyspnea, syncope, acute myocardial infarction, and even sudden death. Clinicians face the challenge of precisely identifying these anomalies associated with adverse outcomes. In this context, recognition of an anomalous origin of the coronary artery from the opposite sinus is particularly relevant due to the significantly increased mortality rate for an anomalous left coronary artery arising from the right sinus (57%) and right coronary artery (RCA) arising from the left sinus (25%).<sup>12</sup>

Single coronary artery by origin is rare, with an autopsy incidence of 0.29%.<sup>2</sup> Forty percent (40%) of the anomalies occur in association with other congenital cardiac abnormalities.<sup>3</sup> A right coronary artery (RCA) originating from the left main (LM) coronary artery accounts for only 0.65% of these anomalies.<sup>1</sup> Congenital absence of right coronary artery (RCA) has been described previously in a 38-year-old female with nonobstructive hypertrophic cardiomyopathy where right coronary artery (RCA) arose as an extension of terminal portion of left circumflex artery (LCx)<sup>13</sup> and in another 78-year-old female with obstructive hypertrophic cardiomyopathy where right coronary artery (RCA) arose as an extension of terminal portion of left anterior descending artery (LAD).<sup>14</sup> McMahon et al<sup>15</sup> reported atresia of the proximal right coronary artery (RCA) in a 2-year-old boy that was associated with fistula from left anterior descending artery (LAD) to mid portion of the right coronary artery (RCA). Our case represents congenital absence of RCA without associated structural cardiac abnormalities.

Right coronary artery anomalies are of controversial significance. Virmani and associates found that in 21 patients with this anomaly, 10 died of cardiac causes, of which 5 were reported as sudden cardiac death (e.g. 4 with no signs of atherosclerosis).<sup>16</sup> There are 2 mechanisms of ischemia that have been proposed for these anomalies. First proposed mechanism depicts that the great vessels, which dilate during exercise, may externally compress the coronary artery.<sup>5,6</sup> The second proposed mechanism suggests that valve-like ridges and the acute angulations of the artery as it traverses from the left to the right sinus may induce ischemia.<sup>6</sup>

In our case, the large calibre dominant LCx due to congenital absence of RCA is supplying the RCA territory and as LAD is totally occluded from its near ostium, LCx is performing the role of both RCA and LAD to supply the myocardium of LAD and RCA territory.

The patient was advised for urgent PCI to LAD or CABG. The patient underwent CABG and discharged with no immediate complication.

## Conclusion

Congenital absence of RCA is not a common finding and of course it's an incidental finding in almost all of the cases while doing invasive coronary angiogram. Noninvasive modalities including electrocardiographic-gated multidetector computed tomography (MDCT)<sup>7</sup> is also another excellent means to diagnose and visualize coronary anomalies without any hazard. But as it's not possible to predict the anomaly by clinical evaluation, CT angiography can not be the first line diagnostic tools to assess the coronary arteries especially when the patient had substantial evidence of coronary artery disease like positive stress exercise test and also regional wall motion abnormality with low LVEF assessed by echocardiography. Still then, in developed countries, now a days as per the patient's choice, initial screening of the coronary arteries by MDCT has been almost a fashion due to its large availability and of course the absence of thought for cost and expenditure. In our case, as the pathology of the coronary artery (LAD) was well relevant with the patient's clinical picture, further evaluation of the coronary anomaly was not necessary. But, in other cases where coronary anomalies may have some clinical impact, details of anomalies can be further evaluated by MDCT.

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