## Congenital absence of the left circumflex artery

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**Abstract** Congenital absence of the left circumflex artery is a rare coronary anomaly with few reported cases in literature. These patients are usually diagnosed incidentally when they undergo either a conventional or a computed tomography (CT) coronary angiography to rule out an underlying coronary artery disease. Coronary CT angiography is a useful noninvasive imaging modality which can be used to diagnose and confirm coronary anomalies reliably. In this article, we report a case of a 55-year-old female who was incidentally found to have a congenitally absent left circumflex artery after a workup was initiated for nonspecific chest pain.

Keywords: Absence and congenital, absent, left circumflex

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#### **INTRODUCTION**

Congenital absence of the left circumflex artery is a rare coronary anomaly with an incidence of 10 in 1495 (0.0067%) patients,<sup>[1,2]</sup> which may be misdiagnosed as complete occlusion of the left artery.<sup>[3]</sup> It is invariably associated with right dominant circulation. Patients with congenital absence of left circumflex artery can present with variable symptoms ranging from dyspnea on exertion to acute-onset myocardial infarction. In this article, we describe the case of a 55-year-old female presenting with nonspecific chest pain but with a strong family history of coronary artery disease (CAD) referred for a computed tomography (CT) coronary angiography which revealed the absence of the left circumflex artery and focal severe stenosis of the mid-right coronary artery. Catheter angiography performed later confirmed the absence of the left circumflex artery. Thus, CT coronary angiography provides a road map to the cardiologist to differentiate between an occluded left circumflex artery from an absent

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left circumflex artery, thus avoiding an iatrogenic creation of a false passage.

#### **CASE REPORT**

A 55-year-old hypertensive female with dyslipidemia and a strong family history of CAD presented with nonspecific chest pain. Electrocardiography (ECG) revealed changes of left ventricular hypertrophy in the form of S wave in lead V1 and R wave in lead V6 measuring more than 35 mm. There were no ECG changes to suggest myocardial infarction. Two-dimensional echocardiography was performed, which confirmed changes of left ventricular hypertrophy. A CT coronary angiography was advised and performed as a screening tool to rule out any significant CAD. It revealed focal short segment severe stenosis of the mid-right coronary artery due to a concentric mixed density wall plaque and nonvisualization of the left circumflex artery. Nonvisualization of the left circumflex artery was attributed to congenital absence [Figures 1 and 2]. A conventional

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coronary angiogram performed later confirmed the CT findings [Figure 3]. Percutaneous transluminal coronary angioplasty was performed for the stenotic segment of the right coronary artery. The patient improved symptomatically postangioplasty.

#### DISCUSSION

Congenital absence of the left circumflex artery to be present is an extremely rare anomaly of the coronary arteries. A report by Srinivasan et al.[4] indicated this condition in only 0.067% of 1495 patients undergoing multidetector CT (MDCT). The congenital absence of the left circumflex artery results from the failure of left circumflex artery development in the left atrioventricular groove. More commonly encountered anomalies involving this artery include left circumflex artery originating from the proximal right sinus of Valsalva, often sharing a common ostium with right coronary artery, or as a proximal branch of the right coronary artery.<sup>[5]</sup> This anomaly is defined as a benign condition and has no significant clinical symptom. However, it can cause angina-like symptoms, particularly on exertion. Most reported cases of this condition are claimed to have experienced chest pain on exertion. The imaging findings of congenitally absent left circumflex artery show no demonstrable left circumflex artery in the left atrioventricular groove [Figure 1b]. This finding is associated with the superdominant right coronary artery continuing into the left circumflex artery territory and prominent multiple diagonal branches from the LAD to supply the lateral wall of the left ventricle, as seen in the literature [Figure 1c].<sup>[4]</sup> CT findings in our case were nonvisualization of the left circumflex artery in the left atrioventricular groove associated with prominent diagonals, dominant right coronary artery, and its posterior left ventricular and posterior descending branches.

Recently, the American College of Cardiology also included CT coronary angiography as a first-line tool for known or suspected anomalies, provided appropriate criteria are used,<sup>[6,7]</sup> Although invasive angiography remains a gold standard in patients with a high pretest probability of CAD.<sup>[7]</sup> Ghadri *et al.* compared CT coronary angiography and invasive angiography for the prevalence of coronary anomalies. The reported prevalence of coronary anomalies on noninvasive CT coronary angiography was 7.85% compared to 2.02% on invasive angiography, which highlights CT coronary angiography as an important tool for the assessment of suspected coronary angiography detected 100% of coronary anomalies as compared to 53% of anomalies on invasive angiography, further



**Figure 1:** Top-view three-dimensional computed tomography coronary angiography (a and b) shows a single branch continuation of the left main artery as the left anterior descending artery (thin arrow) with an absent left circumflex artery in the atrioventricular groove (thick arrows). (c) Prominent diagonal arteries D1 and D2 are seen (thin arrows)



Figure 2: Volume-rendered Computed tomography coronary angiography images. (a) Nonvisualization of the left circumflex artery (thick arrow). (b) Dominant right coronary artery (thick arrow) with its prominent posterior descending artery and posterior left ventricular branches (thin arrows)



**Figure 3:** Coronary angiography (a) right anterior oblique view confirms the absence of the left circumflex artery from the left main coronary artery (yellow arrow). (b) Dominant right coronary artery with its prominent posterior descending artery and posterior left ventricular branches (yellow arrows) and focal moderate stenosis in its midsegment (red arrow)

supporting its use as a better tool for coronary anomalies. <sup>[6]</sup> In addition, CT coronary angiography also offers highly accurate description of coronary anomalies because it can present a three-dimensional image and reliably delineate the origin, course, and termination of coronary arteries and their relationship to cardiac and noncardiac structures.<sup>[6,8]</sup>

Shi et al.<sup>[9]</sup> have reported on the accuracy of MDCT to evaluate coronary artery anomalies. In this study, MDCT and coronary angiography were evaluated in a blinded fashion for their accuracy of anomalous artery origin and path detection, and the results were compared in a secondary consensus. One hundred percent of coronary anomalies were correctly displayed on MDCT. Coronary angiography alone achieved a correct identification of the abnormality in only 53% of cases (P = 0.016). Kacmaz et al.<sup>[10]</sup> have reported on the sensitivity of MDCT in patients who had a coronary artery anomaly demonstrated by conventional coronary angiography. The result was a 100% diagnostic sensitivity for the detection of the origin and course of the anomalous coronary artery. We agree with Kacmaz et al.[10] and Baskurt et al.[11] that CT coronary angiography should be a first-choice imaging modality for the investigation of known or suspected coronary artery anomalies.

Furthermore, these overviews can help the surgeon in preoperative planning of bypass surgery.<sup>[12]</sup>

#### CONCLUSION

The congenital absence of the left circumflex artery is an extremely rare anomaly of coronary arteries. CT coronary angiography is the best modality to detect congenital coronary anomalies because it provides a road map to the cardiologist to differentiate between 100% occluded left circumflex artery from absent left circumflex artery, thus avoiding iatrogenic creation of a false passage. Thus, CT coronary angiography is a highly accurate and sensitive tool in evaluating such coronary anomalies.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### **Conflicts of interest**

There are no conflicts of interest.

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