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CASE REPORT



## Congenital anomaly of coronary artery: absence of left circumflex artery

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### ABSTRACT

The prevalence of congenital coronary artery anomalies is approximately 1% in the general population. They are a common cause of sudden death in younger persons. Congenital absence of the left circumflex artery is usually a benign condition but can cause symptoms of exertional angina. We present a case of a 59-year-old female who presented with complaints of chest pain. She was evaluated by the cardiology service. An invasive angiogram identified the absence of the circumflex artery, a large right coronary artery, and large septal and diagonal branches of the left main coronary artery possibly as a compensatory mechanism to supply blood to the LCx territories. It is important to define coronary anatomy as anomalies dictate which cardiac intervention should be attempted in cases of ischemia.

### ARTICLE HISTORY

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### KEYWORDS

Left circumflex artery; antiplatelet; congenital anomaly; invasive angiography; chest pain

### 1. Introduction

Congenital coronary artery anomalies (CCAA) are a common cause of sudden cardiac death especially in young patients, with a prevalence of approximately 1% among the general population [1]. The reported incidence of absence of the left circumflex artery (LCx) is between 0.003% and 0.067% [2,3]. In the absence of obstructive coronary artery disease, the congenital absence of the LCx is usually considered a benign condition, however it can cause symptoms of syncope, and exertional angina.

### 2. Case report

The patient is a 59-year-old female with a history of hypertension who presented to the emergency department with complaints of precordial chest pain, radiating to the left arm and neck with no aggravating or relieving factors. The patient had a family history of coronary artery disease with her father being diagnosed at 42 years of age. The patient's physical exam was unremarkable. Troponins were negative. The patient was started on aspirin, a statin and a beta blocker. She was evaluated by cardiology who had initially performed a stress test which was abnormal. The patient had an echocardiogram that reported an ejection fraction of 60%-65%. The patient underwent invasive angiography (IA) revealing an absence of the circumflex artery, a large right coronary artery (RCA) with 30% stenosis, and ostial left anterior descending (LAD) with 60% stenosis. (Figures 1 and 2) The patient was offered

the option of intravascular ultrasound (IVUS) guided atherectomy of the ostial LAD lesion, single vessel bypass of the left internal mammary artery to LAD, or maximizing medical therapy. The patient opted for optimizing medical therapy; therefore she was discharged on dual antiplatelet therapy, statin, beta blocker, ranolazine, and isosorbide mononitrate.

### 3. Discussion

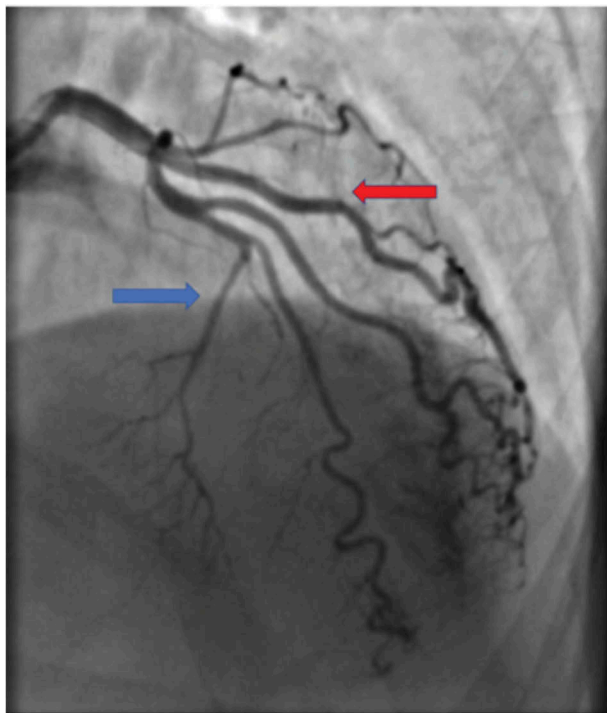
The prevalence of congenital coronary artery anomalies in patients undergoing coronary angiography is 0.3% – 5.6% and approximately 1% are identified on routine autopsy [1]. Congenital absence of the left circumflex artery is an extremely rare occurrence. More commonly, anomalies involving the LCx include originating from RCA as one of its branches or sharing a common ostium with RCA and starting its course from the proximal right sinus of Valsalva [1].

Congenital absence of the LCx occurs due to the failure of the LCx development in the left AV groove. Some experts consider this a normal variant wherein the LCx in this specific condition originates from the distal RCA to perfuse the posterolateral and lateral wall [1]. In other cases, patients usually have a dominant RCA providing compensatory blood supply to the LCx territory in its absence, however in our case the patient had a compensatory dilation of the RCA along with dilation of septal and diagonal branches of the left main artery.

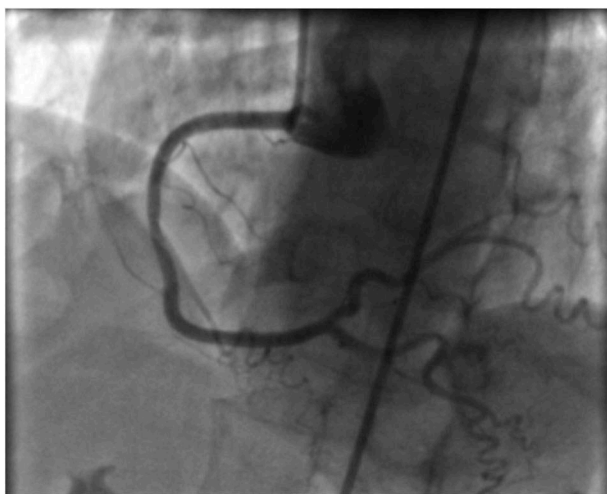
There are very few case reports reporting the absence of the LCx artery. The most common presenting

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**Figure 1.** Invasive Angiogram identifying left anterior descending artery with septal (blue arrow) and diagonal (red arrow) branches.



**Figure 2.** Invasive angiogram showing right coronary artery.

symptom leading to the diagnosis of congenitally absent LCx artery was chest pain [4]. It is mostly a benign condition but can present with symptoms in approximately 20% of patients [5]. It has been reported that congenital absence of the LCx might cause patients to present with syncope, episodic rapid heartbeats, and chest pain. It may also be associated with systolic click syndrome [6]. It can cause symptoms of exertional angina. It has been hypothesized that symptoms on exertion are possibly secondary to a ‘steal’ phenomenon. This phenomenon arises in situations of increased metabolic demands in the LCx territory leading to the diversion of blood flow to these regions resulting in ischemic changes in the LAD or RCA

territories which can mimic an acute coronary event [7]. This can manifest as perfusion defects on stress thallium study [6]. Other potential factors that can alarm the physician for further evaluation of anomalous coronary artery include ischemic symptoms at a young age, family history of premature sudden cardiac death, and lack of other risk factors.

Sudden cardiac death in young patients (<30 years old) with CCAA is most likely due to the anomalous vessel rather than an atherosclerotic disease process [8].

The currently available diagnostic modalities for this rare entity include transthoracic echocardiogram (TTE), which can detect wall motion abnormalities in the LCx artery territory, cardiac magnetic resonance imaging (MRI), coronary computed tomography angiography (CT), and invasive angiography (IA).

The incidence of coronary artery anomalies on TTE was reported at 0.17%, IA at 2.02% and CT at 7.85% [5,9]. Cardiac MRI use is limited due to its inability to visualize small vessels and lack of widespread availability [1].

As CCAA obscures the normal coronary anatomy, therefore, increasing the risk of damage to anomalous vessels during cardiac interventions, it is wise to detect these CCAA beforehand [5]. There is no specific treatment for the absence of LCx; however, appropriate revascularization therapy must be considered when patients with CCAA present with ischemic symptoms. In our case, due to the absence of the LCx and its territories being supplied by the LAD and RCA, even a 60% ostial LAD lesion was considered to be significant for offering IVUS guided atherectomy or single vessel coronary artery bypass graft. The ‘steal’ phenomenon responsible for causing ischemic symptoms may lead to a myocardial infarction involving large areas of myocardium if the LAD develops critical stenosis.

#### 4. Conclusion

Congenital absence of the LCX is an uncommon coronary artery anomaly; however, it is useful to identify CCAA as it may depict the type of cardiac intervention required for reperfusion.

#### Disclosure statement

No potential conflict of interest was reported by the authors.

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