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## A Jellyfish Shaped Proximal Left Anterior Descending Coronary Artery: About an Intriguing Case

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### Abstract

Coronary artery fistulae are rare connections between the coronary vessels and the cardiac chambers or other vascular structures. We present a case of coronary artery fistulae between the proximal left anterior descending artery (LAD) and the main pulmonary artery. The case was admitted with history of chest pain.

**Keywords:** Fistula; Coronary artery disease; Congenital

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### Background

Coronary artery fistula (CAF) is an abnormal communication between a coronary artery and one of the cardiac chambers or a great vessel, so bypassing the myocardial capillaries. They are usually discovered incidentally upon coronary angiography. Clinical manifestations are variable depending on the type of fistula, the severity of shunt, site of shunt, and presence of other cardiac conditions.

### Case

A 63 years old tabetic and hypertensive man was referred to cardiology clinic of LA RABTA with chest pain. His chest pain was retrosternal and effort-related, was relieved by rest, radiated to left arm. He had no history of diabetic mellitus, hyperlipidemia, and family history of coronary artery disease.

There were no signs of cardiopulmonary insufficiency. Physical examination and heart auscultation revealed nothing unusual. ECG showed a sinus rhythm of 76 beats/min, without repolarization anomalies. The transthoracic echocardiogram demonstrated normal wall motion with an ejection fraction of 55% and heart function valve was unremarkable.

Seen a high probability of coronary artery disease, the patient underwent a coronary arteriogram (**Figure 1**), which revealed a big and complex fistula connection arising from the left anterior descending artery (LAD) which was mildly calcified and draining into left pulmonary artery.

The fistula was serpiginous and jellyfish-like from proximal left anterior descending artery (LAD) and it ends in two ways on the left pulmonary artery. There was a significant stenosis at the mid of LAD. The CT-Scan (**Figure 2**) of coronary arteries confirmed the

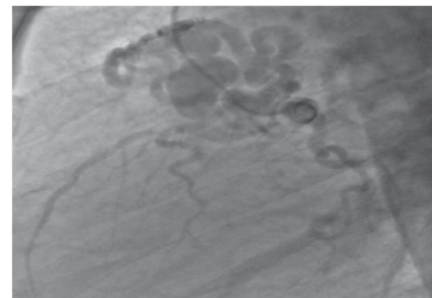
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**Figure 1** Coronary Arteriogram: a big and complex fistula connection arising from the left anterior descending artery.



**Figure 2** CT Scan Axial 5 Chambers View of Heart: aneurysmal calcified and tortuous coronary artery fistulae between proximal segment of LAD and left pulmonary artery.

presence of aneurysmal calcified and tortuous coronary artery fistulae between proximal segment of LAD and left pulmonary artery.

Considering the complexity of this fistulae, we thought that trans catheter repair would not occlude it totally and therefore, surgery would be a more feasible and effective approach. After discussing the risks and benefits of the surgical and trans catheter approaches with the patient, the decision was made to pursue surgical repair.

## Discussion

Current research shows that this is a rare congenital anomaly that occurs in 0.2-0.4% of congenital cardiac anomalies [1]. With the advent of computed tomography (CT) angiography 0.9% of individuals have been incidentally diagnosed with a coronary artery fistula [2].

Fifty percent (50%) of the fistulas were found to arise from the RCA, 42% from the left coronary artery, and 5% from both coronary arteries. The most common site of drainage is the right ventricle (41%), followed by the right atrium (26%) and the pulmonary artery (17%) [3]. Fistula between the left anterior descending artery and the main pulmonary artery, as in the case, is a very rare finding [4].

Fistulas may be multiple feeding arteries to a single drainage point, and multiple drainage sites may exist [5].

Coronary “steal phenomenon” is believed to be the primary pathophysiological problem seen in CAF without outflow obstruction. The mechanism is related to the runoff from the high-pressure coronary vasculature to a low-resistance receiving cavity (e.g., pulmonary vasculature) due to a diastolic pressure gradient

Fistulae can be large (> 250 mm) and dilated or ecstatic, and they tend to enlarge over time [6]. Generally, the symptoms develop depending on the amount of the left-to-right shunt or the presence of coronary steal phenomenon of the fistulae, which usually present in young adults with angina (3–7%), exertional dyspnoea (60%), endocarditis in the fistula (20%), syncope, palpitations, myocardial ischemia and infarction, and manifest in older adults with congestive heart failure (19%), atherosclerosis, and cardiac arrhythmias. Angina pectoris occurs as a result of the coronary “steal phenomenon” where there is blood shunting and perfusion away from the myocardium [7].

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Multidetector computed tomography (MDCT) allows excellent anatomical delineation. The presence or absence of obstruction can be determined with MDCT, and therefore, the likelihood of a coronary steal presentation. A contrast opacification into the receiving chamber/vessel is useful in confirming the CAF entry site and patency of the shunt [8, 9].

Angiography is the main diagnostic technique for the precise diagnosis of the fistulae. Cardiac catheterization provides the hemodynamic evaluation of the fistula and remains the modality of choice for defining coronary artery patterns for structure and flow

Operative management versus embolization would be a feasible alternative for patients who are symptomatic secondary to the coronary artery fistula and remains to be controversial in patients who are asymptomatic

According to the American Heart Association (AHA) guidelines, “percutaneous or surgical closure is a Class I recommendation for large fistulae regardless of symptoms [10]. Surgical treatment is generally reserved for single, large, symptomatic fistulae that are present with angina, cardiac decompression, or complications characterized by high-fistula flow, multiple communications, very tortuous pathways, multiple terminations, significant aneurysmal formation, or need for simultaneous distal bypass [11].

Trans catheter Closure (TCC) avoids surgical intervention and all related complications including surgical stress, bleeding, infections, events related to inflammatory response due to cardiopulmonary bypass, wound healing problems, and general anesthesia adverse events. The TCC technique is indicated when the anatomy of the fistula is favorable for this treatment as vessel tortuosity and lumen caliber appear to be significant limitations in occlusion device delivery, as reported in the small study by Collins et al [12].

## Conclusion

Our case is a good example of a rare congenital anomaly in which coronary artery pathology can remain entirely asymptomatic over many years. Despite the fact that CAF is rare, this diagnosis should be considered in all patients who present with angina, as was evident in this case.

## Conflicts of Interest

No Interest Conflicts.

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