

# Left Anterior Descendent Coronary Artery Fistula to Main Pulmonary Artery with Triple Vessel Disease: A Report of Two Cases

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## Abstract

Coronary artery fistula is a rare anomaly of coronary artery. It represents connection between one or more of coronary arteries and cardiac chamber or great vessel. It can also cause significant hemodynamic changes. To report cases of left anterior descending coronary artery (LAD) fistula to main pulmonary artery (MPA) with concomitant triple vessel disease. Case 1 was 59-year old male presented with intermittent chest discomfort for a year. Echocardiogram showed severe MR with cleft at A1 and A2 and coronary angiogram result showed LAD fistulation to MPA. Case 2 was 57-year old male presented with chest discomfort and at the left shoulder. Echocardiogram showed trivial MR and coronary angiogram showed triple vessel disease along with tortuous fistulation of LAD to MPA. Both patients underwent teflonpledget-reinforced direct suturing of fistula origin and 3-grafts coronary artery bypass graft. Patient no 1 also underwent mitral annuloplasty and valvuloplasty. Post-operative period was uneventful and both patients were discharged after 17 days and 5 days respectively. Short term follow-up showed improvement of symptoms and no residual fistulation. In short term follow-up teflonpledget-reinforced direct suturing of coronary fistula origin result satisfactory. Larger study and further follow up is necessary.

**Keywords:** Coronary artery fistula, direct pledget suturing, coronary artery disease, triple vessel disease, mitral regurgitation.

## Introduction

Coronary artery fistulae are rare anomaly of coronary artery and usually occurs in isolation. It is defined as a connection between one or more of coronary arteries and cardiac chamber or great vessel<sup>1</sup>. The prevalence of coronary artery fistula detected with computed tomography (CT) angiogram was found to be 0.19%-0.9% while conventional angiographic findings was 0.05-0.25%<sup>2,3</sup>. Most are asymptomatic but some may cause hemodynamically significance problem. It may lead to serious complications including myocardial ischemia, heart failure, arrhythmias, thromboembolism, superior vena cava syndrome and even death<sup>4</sup>.

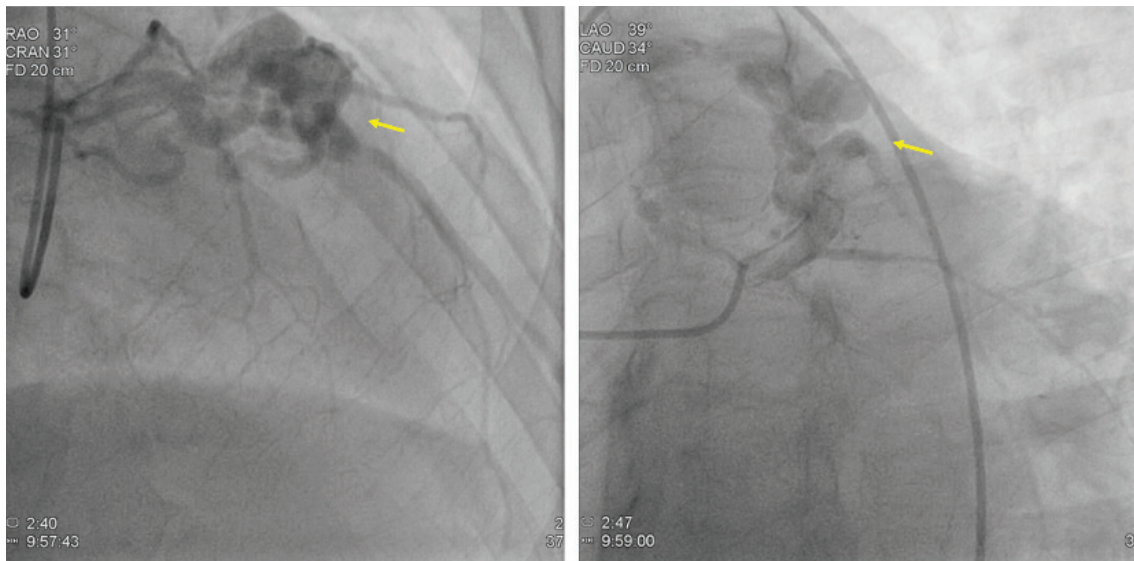
## Case Description

### Case #1

A 59-year old male was referred from a secondary

referral hospital. He presented with intermittent chest discomfort for about a year. Chest discomfort was aggravated with exertion and relieved at rest. He had controlled hypertension but no history or diabetes mellitus or stroke. Patient also had no history of smoking or alcohol consumption.

Physical exam showed grade IV/VI systolic murmur at apex radiated to axilla. Echocardiogram findings were severe MR (Carpentier Type I), dilatation of left atrium, normal systolic function of left ventricle (LV), concentric LV hypertrophy and ejection fraction of 67%. Coronary angiogram showed tortuous and dilated vessel arising superiorly from left anterior descending (LAD) coronary artery (Figure 1a and 1b). Computed tomography angiography (CTA) was also performed. CTA finding showed tortuous, dilated connection between LAD and main pulmonary artery, 2.3 cm in diameter.



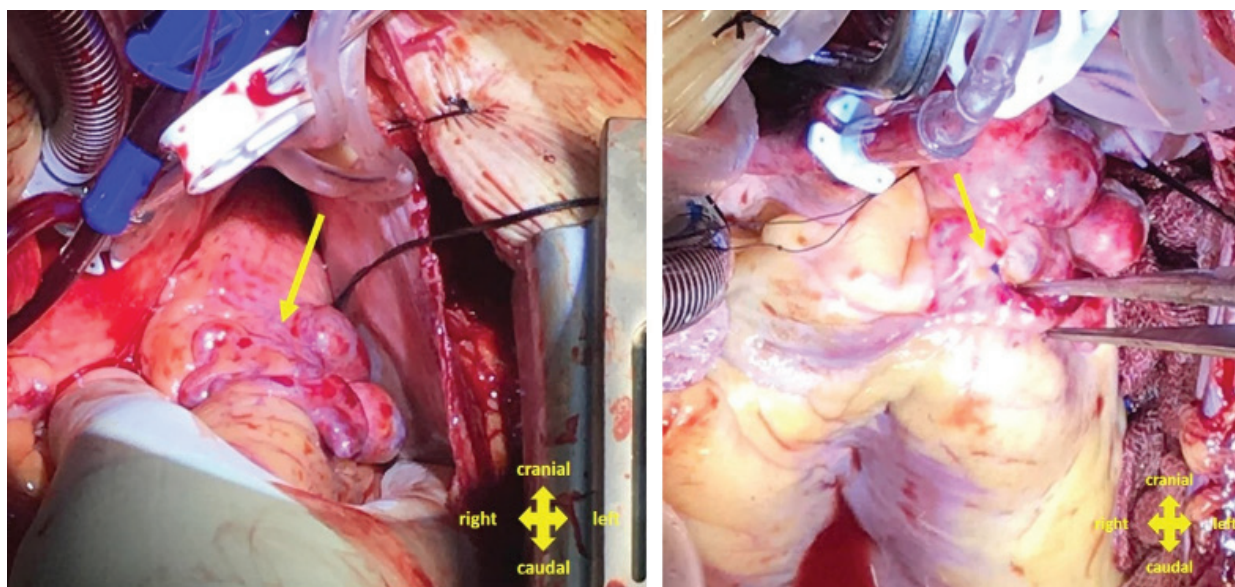
**Figure 1. Coronary angiogram showing dilated and tortuous vessel arising from left anterior descending coronary artery (yellow arrow). 1a)Cranial right anterior oblique projection; 1b)Caudal left anterior oblique projection.**

Surgical repair was then carried out for this patient. Operative finding was tortuous and dilated fistula originated from left anterior descending (LAD) coronary artery (Figure 2a) draining to main pulmonary artery (MPA) along with calcification at proximal to mid right coronary artery (RCA), chronic total occlusion (CTO) in distal RCA, calcification at mid left circumflex (LCx) artery, calcification at anterior mid-LAD and annular dilatation of mitral valve and chordal rupture of P2 and P3, cleft at A1 and A2 of mitral valve.

Three procedures were performed for this patient. Ligation of coronary artery fistula was first carried out, followed by mitral valve repair and then coronary artery bypass graft. Mammary graft was performed later after mitral valve repair.

Coronary artery fistula was identified and then ligated at proximal and distal end of the fistula (Figure 2b). Origin of fistula was ligated using Teflon pledget reinforced direct suture as well as distal end of the fistula

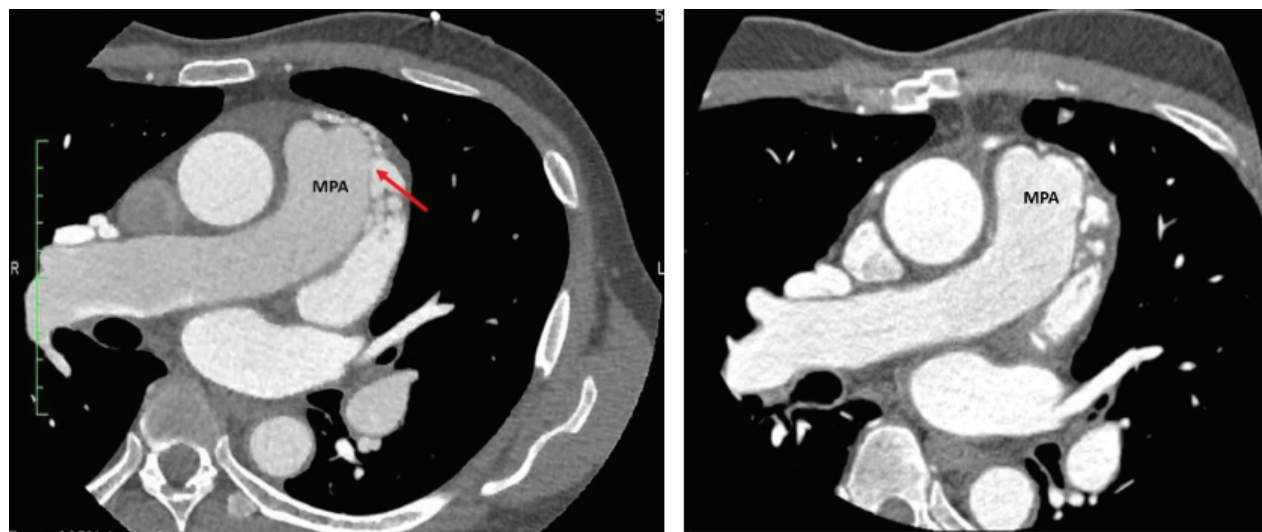
just before MPA. Coronary bypass graft (CABG) using left internal mammary artery and two saphenous vein conduits and mitral valvuloplasty and annuloplasty were also performed. Two saphenous vein conduits and left internal mammary artery were grafted, one vein conduit to Right Posterior Descending Artery, (distal), one vein conduit to distal Right Coronary Artery, and left internal mammary artery to distal left anterior descending (LAD) coronary artery. Valvuloplasty was performed by triangular resection of P2 and closure of cleft between A1 and A2 using 6-0 Prolene. Physio ring was implanted for annuloplasty. Physio ring was stitched using 12 non-pledget suture with 2-0 Premicron® suture. Duration of cardiopulmonary bypass was 256 minutes and duration of aortic cross clamp was 185 minutes. Post-operative blood pressure was 118/68 mmHg, heart rate 100 bpm, central venous pressure 9 cmH<sub>2</sub>O and 99% of peripheral oxygen saturation. Post-operative hemodynamic support was 1 mg/hour of Nitroglycerin and 5 gammas of Dobutamine.



**Figure 2. 2a) (left) Operative findings yellow arrow shows dilated and tortuous segment arising from LAD; 2b) (right) Yellow arrow shows ligated proximal end of coronary artery fistula.**

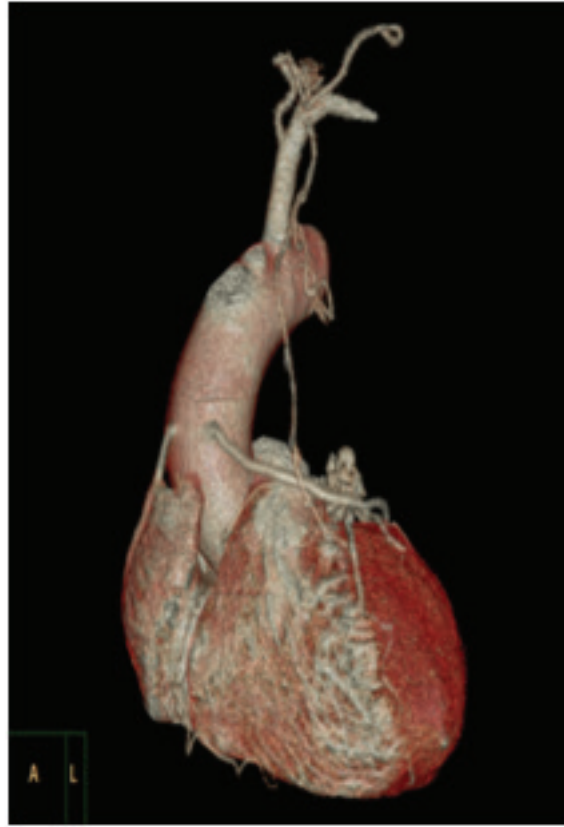
After surgery patient was admitted to ICU for four days and then discharged on post-operative day 17 with no cardiovascular related complication.

After one-year follow-up patient underwent echocardiography and cardiac MSCT Scan. Echocardiogram showed normal cardiac chamber and kinetic, as well as normal left and right ventricular function and ejection fraction was 82%. No obvious fistulation into pulmonal trunk was shown in cardiac MSCT scan (Figure 3b). Multiple patent graft was also shown. (Figure 4).



**Figure 3. 3a) (left) CT angiogram demonstrated connection of fistula to main pulmonary artery (MPA) (red arrow); 3b) (right) Axial slice of cardiac MSCT angiogram one-year post operatively showed no obvious fistulation to main pulmonary artery (MPA).**





**Figure 4. Three-dimensional computed tomography image of after one year follow up showed multiple patent grafts (SVG to RPDA, SVG to distal RCA, LIMA to LAD).**

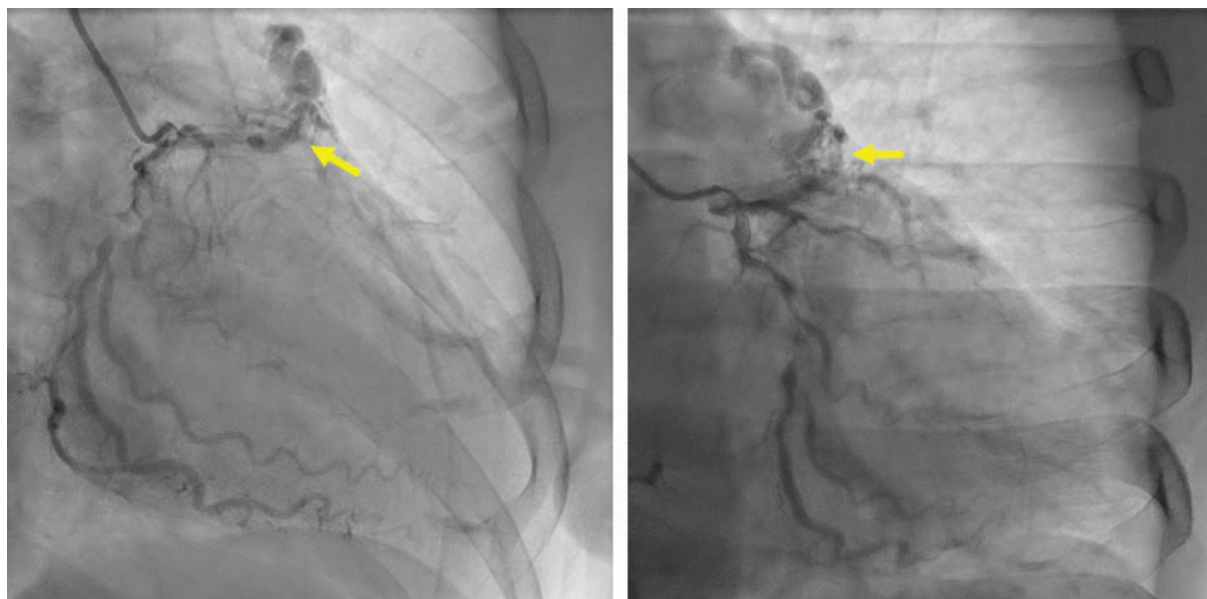
## **Case #2**

Fifty-seven-year-old male presented with symptoms of discomfort around left area of the neck. Discomfort was felt for almost a year. Patient then had himself check to neurologist and then referred to cardiologist before then referred to our hospital.

No abnormalities were found during physical examination. Echocardiography showed normal cardiac chambers and valves, with 64% ejection fraction and normal systolic and diastolic function. Fistulation from

proximal LAD was shown and connect to MPA on cardiac catheterization (Figure 5a and 5b). Chronic total occlusion at proximal of LAD and proximal of RCA, 99% stenosis at distal of LCx were also shown.

During operation, tortuous fistulation from LAD to MPA was shown (Figure 6). The aforementioned fistula was then ligated by Teflon-pledged-reinforced direct suture at the LAD side. Coronary artery bypass graft was also performed which are left internal mammary artery to LAD, saphenous vein graft to distal of RCA, and saphenous vein graft to distal part of LCx.



**Figure 5a and 5b. Fistulation from LAD to MPA was shown on cranial right anterior oblique projection and right anterior oblique projection of coronary catheterization (yellow arrow) along with chronic total occlusion of proximal LAD and 99% stenosis at distal of LCx.**

Aortic cross clamp time of operation was 79 minutes and 127 minutes of cardiopulmonary bypass time. No problem was encountered during operation and patient was hemodynamically supported by 0.5 nano of nitroglycerine and 50 nano of norepinephrine.

Post-operative echocardiogram was performed and trivial mitral regurgitation and trivial tricuspid regurgitation was shown. Left ventricular systolic function was 65% by Teich and 67% by biplane.

Patient was then admitted to intensive care unit and later admitted to elective ward after 3 days. On the seventh day of care patient was then discharge without any significant complications. Short-term follow-up showed improvements of symptoms without any limitations of daily activities.

### Discussion

Coronary artery fistula can occur congenitally or acquired. Most coronary artery fistula are congenital. Acquired coronary artery fistula are extremely rare and usually iatrogenic, posttraumatic, or caused by Takayasu arteritis, Kawasaki disease or chest irradiation<sup>5,6,7</sup>. The feeding artery of the fistula may drain from a main coronary artery or one of its branches and is usually

a dilated and tortuous artery terminating in one of the cardiac chambers of vessel<sup>1</sup>. It may also present with aneurysm of the vessel itself<sup>3</sup>. The fistula may originate from right coronary artery (52%), left anterior descending coronary artery (30%) and circumflex artery (18%)<sup>8</sup>. Fistulas originated from both coronary arteries have also been reported<sup>9,10,11</sup>. The types of coronary artery fistula are coronary to pulmonary artery (76.8%), coronary to bronchial artery fistula (8.9%), coronary artery to cardiac chamber fistula (8.9%), combined coronary to pulmonary and coronary to bronchial artery (3.6%), and coronary artery to superior vena cava fistula (1.8%)<sup>3</sup>. Another congenital anomaly like single coronary artery has been reported concomitant with coronary artery fistula<sup>12</sup>.

Regarding findings in these cases, coronary artery fistula may present with concomitant coronary artery disease. No direct relationship was reported in literature about coronary artery fistula and coronary artery disease, but in most cases coronary artery fistula was found during coronary angiogram for diagnosis of coronary artery disease. Inoue et al reported coronary-pulmonary artery fistula with coronary artery disease related to Kawasaki disease<sup>6</sup>.

Mitral regurgitation found in case #1 was probably related to ischemic heart disease caused by coronary artery disease. In patients with coronary artery disease especially coronary disease involving left anterior descending coronary artery or left circumflex artery or right posterior descending artery (depending on dominance) for the supply papillary muscles<sup>13,14</sup>. No literature was found regarding relationship of mitral regurgitation and coronary artery fistula, yet coronary artery fistula may complicate into mitral valve endocarditis<sup>15</sup>.

Coronary artery fistula may produce hemodynamic changes. When the fistula drains to the right side of the heart, the volume load is increased to the right heart as well as to the pulmonary vascular bed, the left atrium and the left ventricle. When the fistula drains into the left atrium or the left ventricle, there is volume overloading of these chambers but no increase in the pulmonary blood flow. The size of the shunt is determined by the size of the coronary chamber into which the fistula drains<sup>1</sup>.

Coronary arterial fistula is usually asymptomatic in the first two decades, especially when they are hemodynamically small. Even though most patients are asymptomatic, it may also cause serious consequences i.e. myocardial ischemia, thromboembolism, cardiac failure, arrhythmias, rupture, and endocarditis<sup>16,17</sup>. Spontaneous rupture of the aneurysmal fistula causing hemopericardium has also been reported<sup>1</sup>. It is also reported to be associated with bronchiectasis and hypoplasia of pulmonary artery.

Diagnosis of coronary artery fistula can be made by two-dimensional echocardiography and color doppler while coronary angiography and cardiac catheterization are necessary investigations to decide the exact location and thus the mode of intervention to close the fistula<sup>18</sup>. Coronary Computed Tomography Angiography (CTA) is also useful for making diagnosis of coronary artery fistula<sup>3</sup>. Gundogdu et al recommend multi detector computed tomography to be used to precise delineation of coronary fistulas<sup>19</sup>.

Without concomitant heart condition, the indications for treatment of coronary arterial fistulas include the present of a large or increasing left-to-

right shunt, left ventricular overload, myocardial ischemia, left ventricular dysfunction, congestive heart failure and for prevention of endocarditis/ endarteritis. Volume overload of any heart chamber producing secondary valve disease, heart failure, significant left-to-right shunt, disturbed cardiac rhythms and evidence of ischemia necessitate early closure. Indications for interventions in asymptomatic patients are controversial but presence of increased left atrial diameter and Qp/Qs ratio more than 1.3 need early interventions to prevent further complication. Surgery for asymptomatic patients remains controversy but some surgeons advocate operative treatment of coronary artery fistula due to the possibility of sudden death without any warrant symptoms<sup>9</sup>.

Coronary artery fistula may be closed surgically or using coil embolization via trans-catheter approach<sup>20</sup>. Saboo *et al.* point out indications for transcatheter closure and indications for surgical management. Transcatheter closure is indicated when a CAF is proximally located with single drainage, no complicated communications and single drainage site. Transcatheter closure can be performed using coil embolization and may followed by stent implantation if patient has concomitant coronary artery disease<sup>21</sup>. Surgical management is indicated when there are distal fistulas; possible accidental occlusion of important vessels by embolization, multiple complex communications; tortuous arteries, prominent aneurysm, wide drainage site and drainage to coronary sinus<sup>5</sup>. Approach for surgical management may be median sternotomy, left anterior mini-thoracotomy or left parasternal approach<sup>22,23,24</sup>. Some reports also stated that surgical closure is the gold standard for treatment with excellent prognosis<sup>4,18</sup>. In these cases we performed telfonpledget reinforced direct suture to close fistulation of coronary artery. Telfonpledget was used for reinforcement so that the suture will not cut or severe the vessel or the fistula.

Even though outcome from most surgical closure and transcatheter interventions are satisfactory, recurrence of coronary artery fistula has been reported. The exact mechanism of recurrence is still unknown but Kostis *et al.*<sup>25</sup> pointed out that the pulsatile aortic pressure transmitted to the probable small residual

lumen between the in-folded walls of the vessel at the level of the ligature resulted in reestablishment of the fistula, recommending that division and over-sewing should be done whenever feasible to prevent coronary artery fistula from recurrence. Despite ligation and division recurrence may still be occurred therefore long-term follow-up of patient is essential.

### Conclusion

Coronary artery fistula is a rare anomaly of coronary artery that usually are congenital. Most case are asymptomatic but it may cause hemodynamic changes that may result in harmful complications. Diagnostic tools for coronary artery fistula are echocardiography, coronary angiography and CT angiography. Management of coronary artery fistula can be transcatheter closure or surgical repair. Long term follow-up is essential since recurrence may occur. In our cases surgical management with by Teflon-pledget-reinforced direct suture of coronary artery fistula of left descending coronary artery to pulmonary provide satisfactory outcome with uneventful result and remarkable recovery of patients.

**Conflict of Interest :** The author declare that they have no conflict of interest.

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