

# Giant Left Circumflex Artery Aneurysm with Coronary Sinus Fistula

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## Abstract

Giant coronary artery aneurysm with fistula formation is very uncommon. Coronary artery fistulas are rare congenital anomalies and constitute the most common form of hemodynamically significant coronary artery malformation. Given the rarity of condition, no clear cut guidelines exist for the optimal management of giant coronary artery aneurysms. The natural history of coronary artery fistulas is variable. The majority of coronary artery fistulas are asymptomatic in early life. They have long periods of stability followed by either sudden onset or gradual progression of complications in later part of life. We describe management of a case of myocardial ischemia caused by giant circumflex coronary artery aneurysm with fistula to coronary sinus.

**Keywords:** Coronary Artery Aneurysm; Coronary Sinus Fistula; Arteriovenous Fistula

## Introduction

Coronary artery fistulas are a very rare cardiac anomaly. The reported Incidence of coronary artery fistula is 0.1- 0.2% in all patients undergoing coronary angiography [1]. 26% of the patients with coronary artery fistula have associated coronary artery aneurysm. Giant coronary artery aneurysms with diameter exceeding 50 mm are extremely rare. As the aneurysm enlarges, complications related to volume overload, such as congestive heart failure, myocardial infarction due to coronary artery steal phenomenon, thromboembolic and rupture of the aneurysm/fistula, may occur [2]. We describe management of a case of myocardial ischemia caused by giant circumflex coronary artery aneurysm with fistula to coronary sinus.

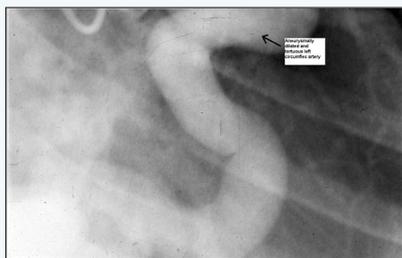
## Case Report

A 50 yr old woman presented with palpitation, exertional chest pain for 6 months. Patient was followed in local health centre for atrial fibrillation and was on digoxin. Physical examination was unremarkable. Chest auscultation revealed a continuous grade III/IV murmur at left 4<sup>th</sup> Intercostal space. An electrocardiogram was normal. Chest X ray showed pulmonary congestion and cardiomegaly. Echocardiogram showed dilated left ventricle with normal left ventricular function, dilated

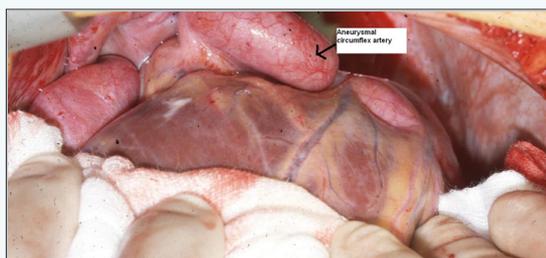
right atrium and right ventricle. Right heart catheterization showed step up in oxygen saturation at the right atrium level and Pulmonary: Systemic oxygen ratio of 2:0. The coronary angiography showed normal coronaries with aneurysmally dilated and tortuous left circumflex artery communicating with markedly dilated coronary sinus (Figures 1 & 2). There were no CT scanning facilities at that time. Patient was taken up for surgical correction because of anginal symptoms and presence of giant left circumflex artery aneurysm with coronary sinus fistula. Median sternotomy was done, palpation over the coronary sinus revealed continuous thrill. The circumflex artery was hugely dilated (4-5 cm) and tortuous (Figure 3).



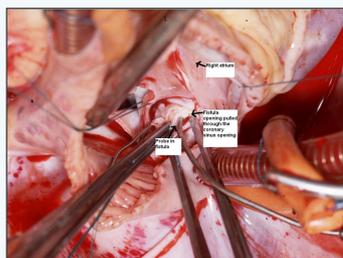
**Figure 1:** Coronary angiogram showing dilated and tortuous circumflex artery.



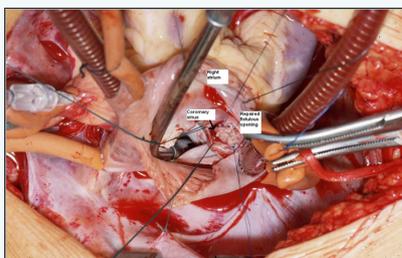
**Figure 2:** Coronary angiogram showing dilated circumflex artery and coronary sinus.



**Figure 3:** Operative picture showing aneurysmal dilated circumflex artery.



**Figure 4:** Operative picture showing fistulous opening pulled through coronary sinus opening.



**Figure 5:** Operative picture showing repaired fistulous opening.

Patient was put on cardiopulmonary bypass. Myocardial protection was achieved by cold blood hyperkalaemic cardioplegia. Right atrium was opened and coronary sinus inspected. Coronary sinus opening was markedly dilated. The fistulous opening could be seen through the coronary sinus ostium and was confirmed by probing (Figure 4). It was pulled out through the ostium and closed with continuous 4-0 prolene suture (Figure 5). Coronary arteries were normal therefore no bypass graft was required. Pt was weaned off from CPB without inotropic support. Intraoperative TEE showed no residual shunt. Post-operative period was uneventful. Follow-up echocardiography showed reduction in the size of circumflex artery aneurysm and patient remained asymptomatic. At fifteen years of follow up, the patient was doing well and has been angina free.

## Discussion

Coronary anomalies are reported in 1-3% of all coronary artery angiography, out of these 10-13% are coronary artery fistulas [3]. Giant coronary artery aneurysms with fistula are extremely rare with only few reported cases in the literature. Majority of coronary artery fistulas are asymptomatic and are found incidentally on coronary angiography. The main causes of coronary artery aneurysm are atherosclerosis, congenital Kawasaki disease, iatrogenic following percutaneous interventions, and following endocarditis [4]. There is no sex or age predilection. The patient's become symptomatic due to coronary ischemia or heart failure. Heart failure is due to volume overload and angina symptoms are due to high flow in the fistula resulting in coronary steal phenomenon with resulting ischemia of area supplied by that coronary artery [5]. Presently, there are no clear cut guidelines in the management of such pts because of the rarity of the condition.

Management of patients with coronary artery fistula depends on symptoms, fistula location, its size and the resulting shunt volume [6]. Lowe and colleagues recommend surgical correction for all pt with giant aneurysm with concomitant fistula [7]. A percutaneous approach may be suitable for small and nonaneurysmal coronary artery fistula. In our case presence of anginal symptoms with associated giant left circumflex artery aneurysm with potential risk of rupture were the reasons for surgical management. In asymptomatic pt with small aneurysm simple observation may be justifiable. Multiple modalities of imaging techniques, including echocardiography, coronary angiography, and high resolution computed tomography can clearly delineate the exact anatomy and location of these tortuous fistulas, and are of great help to the surgeon in planning the best approach to the fistula.

## Conclusion

Coronary artery aneurysm with coronary artery fistula is a rare entity. Complication of coronary artery aneurysm are rupture, angina and with associated fistula, heart failure and infective endocarditis. Asymptomatic cases may be dealt with simple observation, but large symptomatic aneurysm with fistula requires surgical intervention. Surgical procedure for coronary aneurysm with fistula includes resection or plication of aneurysm and isolation of blood supply to the fistula, along with the closure of fistula. Bypass graft is required if the native coronary blood flow is compromised.

## Disclosure

All the authors have declared no competing interest.

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