Int Cardiovasc Res J.2015;**9**(3):183-186.icrj.21014

International Cardiovascular Research Journal

www.ircrj.com

A Young Boy with Coronary Cameral Fistula

Mohammad Arifur Rahman [1,*], Afzalur Rahman [1], Mohammad Mahbubur Rahman [1]

[1] National Institute of Cardiovascular Diseases, Sher e bangle nagar, Dhaka, Bangladesh

ARTICLE INFO

Article Type:
Case Report

Article History:
Received: 03 Jul 2014
Accepted: 09 Feb 2015

Keywords:
Fistula
Heart Murmurs

ABSTRACT

Coronary-Cameral Fistula (CCF) is an anomalous connection between a coronary artery and cardiac chamber. Most CCFs are discovered incidentally during angiographic evaluation for coronary vascular disorder. Here, we report a 16-year-old boy with exertional breathlessness for 3 years. There was a continuous murmur at the right para sternal border in the 4th and 5th intercostal space. Echocardiography showed Right Ventricular Hypertrophy (RVH) with strain pattern. Besides, transthoracic echocardiography demonstrated a normally functioning left ventricle, but dilated right atrium and ventricle. The right ventricle also showed hypertrophy and trabeculation. Coronary angiography demonstrated a direct connection between the right ventricular cavity and the right epicardial coronary artery. However, the left coronary arterial system was normal. The patient was treated by ligation of the fistulous connection by off-pump surgery.

►*Implication for health policy/practice/research/medical education:*
Coronary cameral fistula is a very rare condition. Patients usually present with breathlessness and chest pain or they may remain asymptomatic. Enthusiastic physicians should be aware of such type of condition so that it is not missed. It is a correctable congenital heart disease. Early diagnosis can save lives by corrections either by cardiac interventions, such a coil embolization, or by surgery.

1. Introduction

Coronary Cameral Fistula (CCF) is a very rare condition which may be congenital or acquired due to trauma or intervention. In this condition, coronary artery terminates into cardiac chamber. Most commonly, the right coronary artery drains into the right ventricle. This condition is normally diagnosed incidentally while evaluating a patient presenting with chest pain or breathlessness. An expert echo cardiographer can suggest the condition, but coronary angiogram confirms the diagnosis. Although most patients are asymptomatic, it can lead to symptoms, such as angina pectoris and breathlessness (1). Here, we report a case of CCF with breathlessness. Selective coronary angiography revealed diffuse CCF involving the right coronary artery emptying into the right ventricle.

2. Case Presentation

A 16-year-old boy was admitted in National Institute of Cardiovascular Diseases (NICVD) with exertional breathlessness for 3 years. It was NYHA grade I initially, but class II for the last 3 months. No diurnal or seasonal variation of his breathlessness was detected. His breathlessness was associated with palpitation, which was relieved by rest. He did not have any chest pain or haemoptysis. He had given h/o repeated Respiratory Tract Infection (RTI) since childhood. On examination, the patient was ill looking. His pulse rate was 98/min, regular, and collapsing in nature, his blood pressure was 120/30 mmHg, and his Jugular Venous Pressure (JVP) was not raised. Besides, his whole precordium was pulsatile. The apex beat was in the left 5th intercostal space, medial to the midclavicular line, thrusting in character. Left parasternal heave was also present and P2 was palpable. There was a systolic thrill present in parasternal area of the right 4th and 5th intercostal space. Moreover, the 1st heart sound was normal, while the pulmonary component of the second heart sound was loud. There was a continuous murmur at the right para sternal border in the 4th and 5th intercostal space, grade 4/6, without any radiation, which was best heard with the diaphragm of the stethoscope with held breath in expiration.

*Corresponding author: Mohammad Arifur Rahman, National Institute of Cardiovascular Diseases, Sher e bangle nagar, Dhaka, Bangladesh,
Tel: +88-017-11666030,
E-mail: drarif79@yahoo.com

Other system examinations revealed no abnormalities.

A standard 12 lead ECG showed Right Ventricular Hypertrophy (RVH) with strain pattern (Figure 1). In addition, Chest X Ray Postero Anterior view (CXR P/A) showed cardiomegaly with right ventricle type apex. Transthoracic echocardiography demonstrated a normally functioning left ventricle, and dilated right atrium and ventricle. Besides, the right ventricle showed hypertrophy and trabeculation. Imaging with color flow Doppler demonstrated blood flow throughout diastole from the epicardial surface into the right ventricular cavity through the hypertrophied segment of myocardium (Figure 2). The mitral and aortic valves both opened well with trivial tricuspid regurgitation and the pulmonary arterial systolic pressure estimated from a trivial jet of Tricuspid Regurgitation (TR) was approximately 30 mmHg (Figure 3). Cardiac computed tomography coronary angiogram and right coronary angiography demonstrated a direct connection between the right ventricular cavity and the right epicardial coronary artery (Figure 4). Nonetheless, the left coronary arterial system was normal. Surgical repair of the CCF was done (Figure 5).

3. Discussion

Coronary fistulae with the cardiac chambers (cameral fistulae) are rare congenital vascular anomalies reported

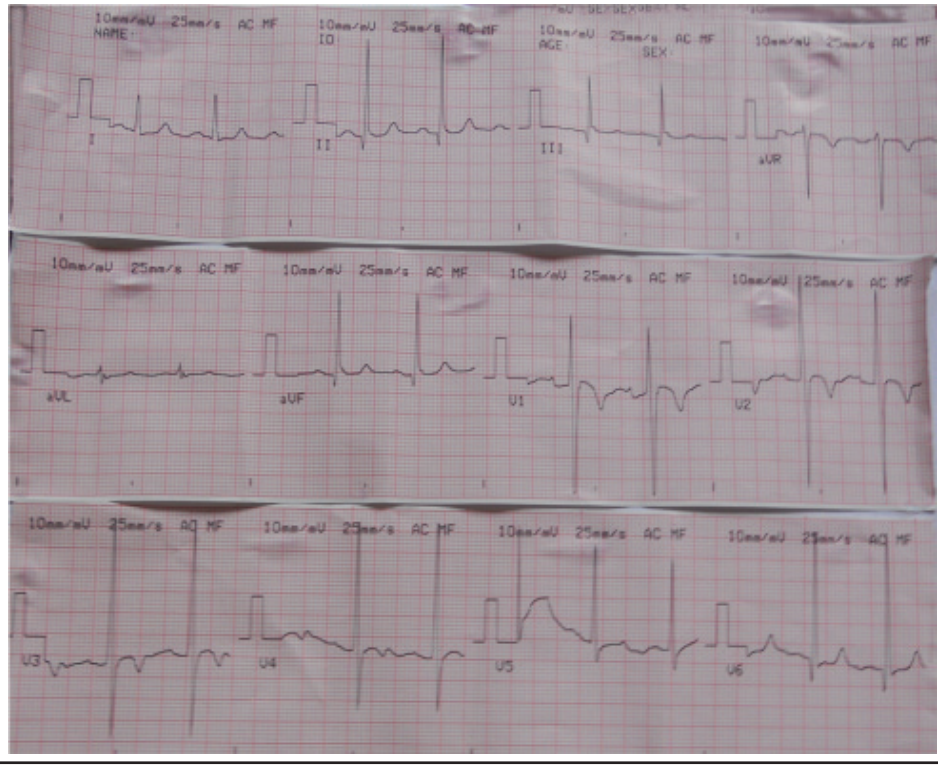

Figure 1. ECG Showing RVH with Strain

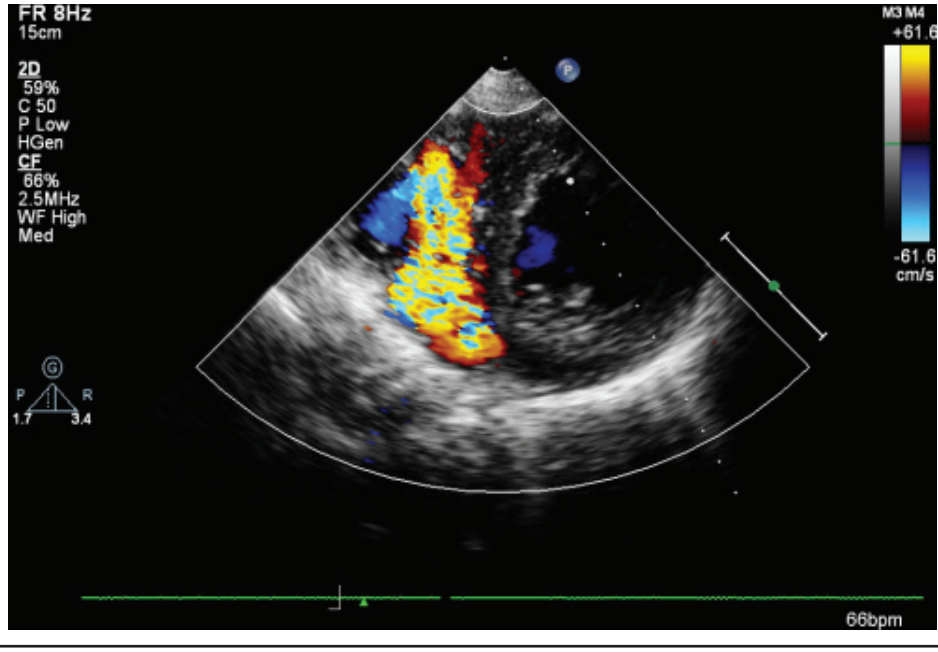

Figure 2. Continuous Flow through Fistula

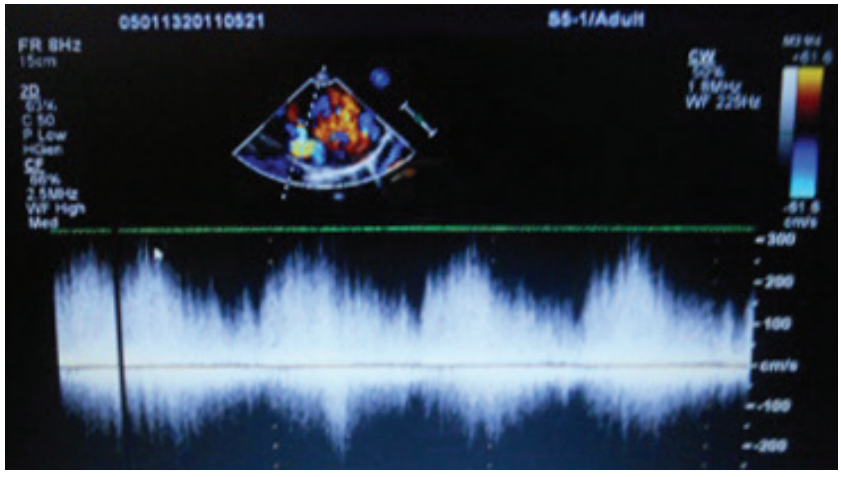

Figure 3. Doppler Spectral Tracing of Continuous Flow through the Fistula

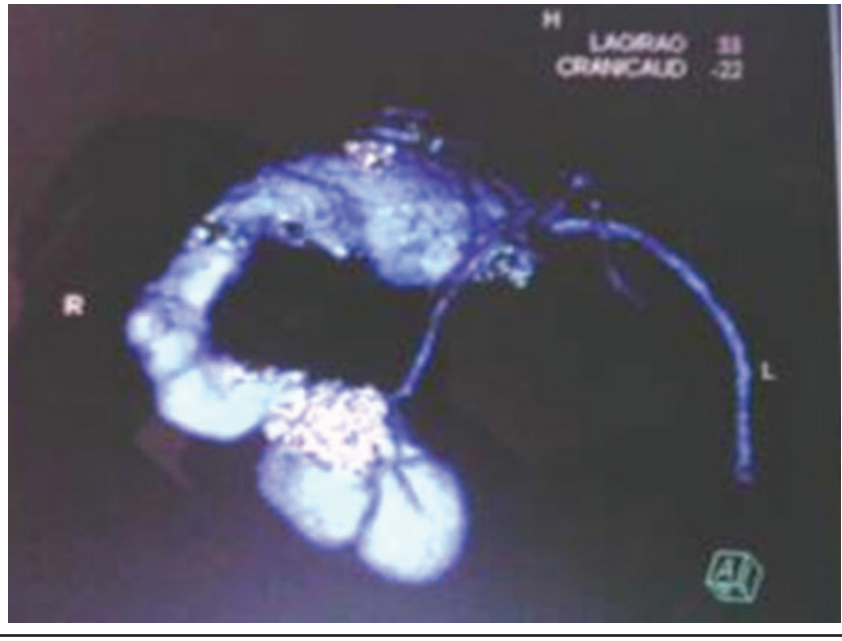

Figure 4. Cardiac CT Angiogram of CCF Draining to the Right Ventricle

to be found in approximately 0.08 - 0.3% of unselected patients undergoing diagnostic coronary angiography (2, 3). CCFs are also seen in 0.1% of patients undergoing coronary angiograms. Major sites of origin of fistula are the right coronary artery (55%), left coronary artery (35%), and both coronary arteries (5%). Besides, the major termination sites are the right ventricle (40%), right atrium (26%), pulmonary arteries (17%), less frequently the superior vena cava or coronary sinus, and least often the left atrium and left ventricle.

Coronary artery left ventricular fistulae is exceedingly rare with the incidence rate of 1.2% (4). Cardiac catheterization with coronary angiography remains the gold standard for diagnosis of coronary artery fistulae. It can demonstrate the size, anatomy, number, origination, and termination site of the fistulae. Cardiac echocardiography is also useful for diagnosis. Magnetic resonance imaging and multidetector computed tomography are also used to evaluate the anatomy, flow, and function of CCF (5, 6). Clinical presentation generally depends on the hemodynamic significance of the anomaly and most commonly, coronary artery fistulae are asymptomatic and are found incidentally (7). Anginal symptoms may be the presenting feature, particularly in patients with multiple fistulae. In the patients with a single fistula, on the other hand, exertional dyspnoea is more likely to predominate. Inducible ischaemia has been well demonstrated in these patients and is thought to occur as a result of left to left shunting, causing a coronary steal phenomenon and diastolic overload (8). CCF may also cause myocardial infarction (9), congestive heart failure, arrhythmias, and aneurysmal formation. Rupture of the affected vessels may occur, as well (10). Furthermore, turbulent blood flow across the fistula may give rise to development of infective endocarditis. This complication is more common in older patients.

Cameral fistulae are recognized to be associated with regional hypertrophy. In this case, it is unclear whether hypertrophy is a cause or effect of the abnormal coronary anatomy (11). The appearance of blood flow within the myocardium may cause diagnostic uncertainty. The trabeculated appearance of the left ventricle on transthoracic echocardiography in the present case raised the suspicion of left ventricular non-compaction; however, inspection of the color Doppler imaging clearly revealed a sustained

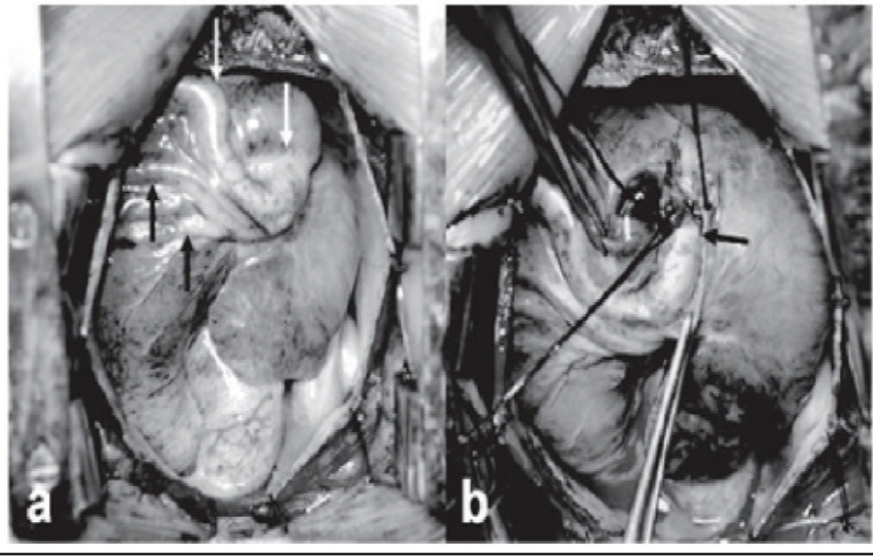

Figure 5. Intra-Operative Photographs Showing: (a) The Origin of the Single Giant Left Coronary Artery and Dilated Right Coronary Artery (White Arrows) and the Initial Left Coronary Branches (Black Arrows) and (b) Fistula from the Right Coronary Artery to the Right Ventricle (Black Arrow) Being Ligated Off-Pump

flow of blood from the epicardial surface to the LV cavity throughout diastole more suggestive of a coronary fistula. In this case, the diagnosis was established by conventional coronary angiography though the coronary anatomy may alternatively be delineated using computed tomography coronary angiography (12). Complications if untreated are myocardial ischemia, arrhythmias, endarteritis, aneurysm rupture, and thrombus formation. The best way to manage cameral fistulae is uncertain largely due to the rarity of the condition. Patients in whom focal fistulae with large shunts exist may benefit from closure of the shunt and if this is to be performed, it is probably best done as early as possible (13). Indications for closure are increased/increasing L ->R shunt, left ventricular volume overload, myocardial ischemia, left ventricular dysfunction, CCF, and prevention of endocarditis. Coil embolisation can be done by Gianturco, Jackson, and Flipper coils (Cook), GDC/IDC coils (F3 catheter), embolisation devices, and occlude devices or plugs such as Cera vascular plug system. Yet, multiple fistulae of the sinusoidal type are unlikely to be amenable to surgical correction and can be treated with beta-blocker (14, 15).

Acknowledgements

There is no acknowledgement.

Authors' Contribution

Mohammad Arifur Rahman developed the original idea, prepared the case, and did the necessary evaluation. Afzalur Rahman supervised the case and did the necessary correction. Mahbubur Rahman helped doing ECG, ECHo, and analysis of the reports.

Financial disclosure

The authors declare that there is no conflict of interests.

Funding/Support

There is no funding/support.

References

1. Brooks CH, Bates PD. Coronary artery-left ventricular fistula with angina pectoris. *Am Heart J*. 1983;**106**(2):404-6.
2. Vavuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. *Cathet Cardiovasc Diagn*. 1995;**35**(2):116-20.
3. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn*. 1990;**21**(1):28-40.
4. Ho H, Cheung C, Jim M, Lam L. Coronary–cameral fistula. *Heart*. 2005;**91**(12):1540-.
5. Lessick J, Kumar G, Beyar R, Lorber A, Engel A. Anomalous origin of a posterior descending artery from the right pulmonary artery: report of a rare case diagnosed by multidetector computed tomography angiography. *J Comput Assist Tomogr*. 2004;**28**(6):857-9.
6. Parga JR, Ikari NM, Bustamante LN, Rochitte CE, de Avila LF, Oliveira SA. Case report: MRI evaluation of congenital coronary artery fistulae. *Br J Radiol*. 2004;**77**(918):508-11.
7. Lee J, Choe YH, Kim HJ, Park JE. Magnetic resonance imaging demonstration of anomalous origin of the right coronary artery from the left coronary sinus associated with acute myocardial infarction. *J Comput Assist Tomogr*. 2003;**27**(2):289-91.
8. Vanchon J. Les fistules congenitales coronaro-ventriculaires gauches. *Ann Cardiol Angiol*. 1983;**32**:21-5.
9. Stierle U, Giannitsis E, Sheikhzadeh A, Potratz J. Myocardial ischemia in generalized coronary artery-left ventricular microfistulae. *Int J Cardiol*. 1998;**63**(1):47-52.
10. McLellan BA, Pelikan PC. Myocardial infarction due to multiple coronary-ventricular fistulas. *Cathet Cardiovasc Diagn*. 1989;**16**(4):247-9.
11. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. *Circulation*. 1979;**59**(5):849-54.
12. Monmeneu JV, Bodi V, Sanchis J, Chorro FJ, Llopis R, Insa L, *et al*. [Apical hypertrophic myocardiopathy and multiple fistulae between the coronary vessels and the left ventricle]. *Rev Esp Cardiol*. 1995;**48**(11):768-70.
13. Manghat NE, Morgan-Hughes GJ, Marshall AJ, Roobottom CA. Multidetector row computed tomography: imaging congenital coronary artery anomalies in adults. *Heart*. 2005;**91**(12):1515-22.
14. Chia BL, Chan AL, Tan LK, Ng RA, Chiang SP. Coronary artery-left ventricular fistula. *Cardiology*. 1981;**68**(3):167-79.
15. Sheikhzadeh A, Stierle U, Langbehn AF, Thoran P, Diederich KW. Generalized coronary arterio-systemic (left ventricular) fistula. Case report and review of literature. *Jpn Heart J*. 1986;**27**(4):533-44.