

Coronary Artery to Pulmonary Artery Fistula: A Congenital Rare Defect in Adult. How was it Managed?-A Case Report

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Abstract

Coronary artery fistula (CAF) is a rare anatomic abnormality of the coronary arteries that results from abnormal communication between a coronary artery and either the chambers of the heart or the great vessels that bypass the myocardial capillary bed. It is usually congenital, but it can be acquired. Both male and females are affected. Clinical presentation may be asymptomatic or symptomatic and it depends on the type of fistula, size of the fistula, site of the shunt and severity of the shunt. Coronary angiography is the gold standard for the diagnosis of coronary artery fistula. Surgery is indicated when a patient is clinically symptomatic, even in asymptomatic patients those are at risk for future complications.

Keywords: Coronary artery fistula (CAF); Pulmonary artery; Right coronary artery; Coronary angiography.

Introduction

Coronary artery fistula (CAF) is a rare anomaly of the coronary arteries that results from abnormal communication between a coronary artery with a cardiac chamber or the great vessels of the heart. It is usually congenital, but it can be acquired.

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Received Date: 05-29-2023

Accepted Date: 06-12-2023

Published Date: 06-26-2023

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Coronary artery fistula occurs in both sexes. Since there are probably many untreated cases, the exact incidence of Coronary artery fistula is unknown. In general population, the incidence of coronary anomalies is 0.2% to 1.2% where coronary artery fistulas (CAFs) are present in 0.002% of those cases [1,2]. The prevalence of congenital coronary artery fistula is approximately 1% in general population [3] and around 2,000 years ago, it was first described by Galene and Versalius [4]. After being first described by Austrian anatomist Josef Hrytl in 1841, the existence of CAFs has been well documented [5]. An abnormal connection between a coronary artery and a heart chamber was reported by German anatomist W. Kraus in 1865 [6].

These coronary anomalies can arise from abnormal involution, truncus arteriosus division, or a shift in the position of the endothelial buds during the embryonic stage [7,8] and the formation of Coronary artery fistula and the persistence of embryonic sinusoids beyond what is predicted are closely connected [9].

Congenital coronary artery fistula arises from right coronary artery and left coronary artery or its branches and drains into cardiac chambers and pulmonary trunk. In some cases, especially in adults, fistulae may originate from both coronaries' arteries, which drain into the main pulmonary artery or pulmonary trunk. Coronary artery to pulmonary artery fistula is a type of coronary artery fistula in which coronary artery communicates with the main pulmonary artery. Those patients who undergo invasive angiography; coronary artery fistulas (CAFs) are found in ranging from 0.05% to 0.25% [10].

Clinical presentation of coronary artery fistula depends on the type of the fistula, site of the shunt, shunt volume. A little more than half of CAF patients have symptoms. Chest pain at rest, palpitations, exhaustion, and exertional dyspnoea are among the symptoms and may be asymptomatic. Treatment of patients with coronary artery fistulas (CAFs) is based on the presence of symptoms, the anatomy and size of the fistula, and whether it is connected to cardiovascular diseases [11].

Coronary artery fistula can be managed conservatively, surgically or by transcatheter technique. Here the authors report a case of congenital coronary artery fistula which arises from the right coronary artery which drains into main pulmonary artery, a rare defect managed surgically.

Case report

A 49-year-old man got admitted to cardiology unit of our hospital throughout patient department with the complaints of chest pain and tightness of the chest on exertion for about six months which got aggravated for 10-15 days. The pain was retrosternal which radiated to the back of the chest and relieved by rest. Pain associated with sweating, palpitation, shortness of breath but not associated with nausea, vomiting, pre-syncope, or syncope.

The patient was diabetic and hypertensive. The patient had no history of chronic kidney disease, cerebrovascular disease, dyslipidaemia, smoking, family history of coronary artery disease, chronic obstructive pulmonary disease. On physical examination-patient was hemodynamically stable (pulse rate was 76 beats/min, blood pressure was

110/60mmhg, and a respiratory rate was 20 breaths/min). Jugular venous pressure was not elevated. On auscultation-Ejection systolic murmur present at the left sternal border but there was no crackling or wheezing in both lungs. There was no evidence of ankle oedema or hepatomegaly.

The electrocardiogram showed sinus rhythm with biphasic T wave in V₃-V₆ precordial leads. Chest radiograph showed mild cardiomegaly and slightly increased pulmonary vasculature. All biochemical investigations were within normal limits.

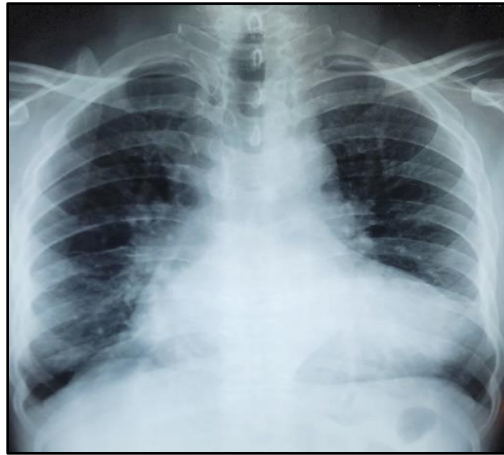


Figure 1: Chest X-ray showed cardiomegaly and increased pulmonary vasculature.

Echocardiogram showed no gross regional wall motion abnormality with normal LV systolic function (EF-50%), concentric LVH with normal heart valve function.

Afterward Coronary angiogram was performed which revealed normal epicardial coronary arteries with coronary fistula arising from proximal RCA and draining into Pulmonary artery (PA).

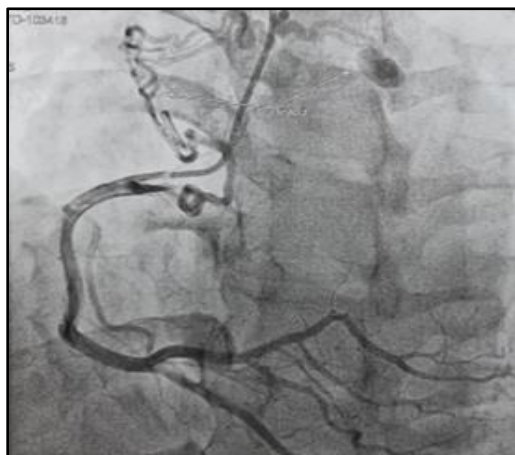


Figure 2: CAG showed RCA to PA fistula.



Figure 3: CAG Showed RCA to PA fistula.

With the evidence of coronary angiography, the patient was selected for surgical intervention. The patient was then sent to the operating room for surgical closure. Surgery was performed with endotracheal intubation under G/A and median sternotomy with the help of cardiopulmonary bypass with moderate hypothermia and

antegrade cold blood cardioplegia. After median sternotomy, pericardiotomy was done. The aorta showed healthy, PA (Pulmonary artery) showed mild tense, RV-Enlarged, LV-Normal, PA pressure measured 19 mmHg. Dilated, tortuous blood vessels were found over the mid and lower part of the ascending aorta.



Figure 4: Per-operative findings: Showed dilated, tortuous blood vessels over the aorta.

On palpation-Thrill was found over the ascending aorta. Anatomy and course of coronary arteries were checked. Cardiopulmonary bypass was established after systemic heparinization with aortic and bicaval venous cannulation (superior and inferior venacava separately). Cross clamp

was applied. Heart was arrested for maintaining myocardial protection by antegrade cold blood cardioplegia. The pulmonary arteriotomy was done and then checked out the opening of the fistula. Intrapulmonary opening of clonal branch of right coronary artery was then found and

closed the opening of the fistula with pledged 5-0 monofilament proline suture. Dilated

collateral branches were also sewed with stitches.

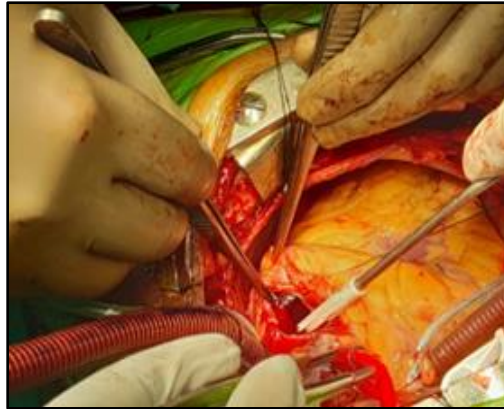


Figure 5: Pulmonary arteriotomy.

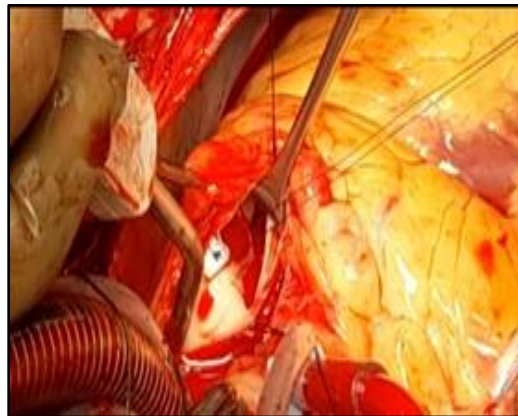


Figure 6: Closure of opening of fistula arising from RCA.

Pulmonary arteriotomy was closed. A heart was deaired and gradually weaned from cardiopulmonary bypass. Decannulation was

done and heparin was reversed with protamine sulphate.

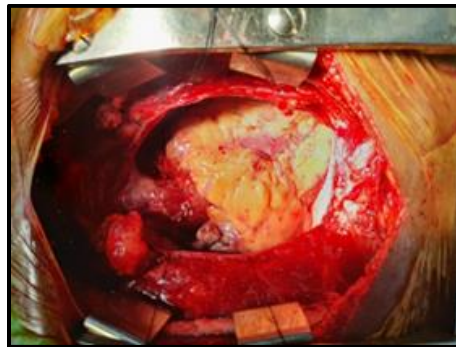


Figure 7: After completion of fistula repair.

After maintaining proper hemostasis, the wound was closed in layers after keeping mediastinal drain tubes in situ. Patient was shifted to CT-ICU with stable hemodynamic condition. The patient was extubated 6th hour after the end time of surgery and the subsequent post-operative period was uneventful. The patient was discharged from hospital on the 7th day after surgery with good condition.

Follow up

At the two-year follow-up, the patient was asymptomatic and doing well and the echocardiography revealed 50% LVEF.

Discussion

The congenital anomaly of a coronary artery fistula is extremely uncommon. Only 0.4% of congenital cardiac abnormalities and 14% of coronary anomalies are caused by it [12]. Coronary artery fistulae can be ecstatic or big and dilated, and they usually get broader over time [13]. Although CAF was previously typically congenital, the development and widespread use of interventional and surgical techniques over time has changed its etiology, increasing the prevalence of acquired forms [14] which may be caused by infective endocarditis, aortic dissection, prior surgery, endomyocardial biopsy, coronary angioplasty and bypass surgery, valve replacement, cardiac transplantation, trauma, permanent pacemaker placement, and others. Most coronary artery fistulas do not cause any symptoms. On average, coronary artery fistulas will remain asymptomatic during childhood. Symptoms usually occur after 20 years of age which include angina, dyspnea, fatigue, murmurs, arrhythmias, endocarditis,

congestive heart failure [15]. Angina and dyspnea are the most common symptoms. An increase in the left-to-right shunt and gradual fistula enlargement are linked to symptoms getting worse [16]. In a review of 363 cases of CAF, it was discovered that RCA, the left coronary artery, and both coronary arteries all contributed to fistulas in 50%, 42%, and 5% of the cases, respectively. The right ventricle (41%) is the most frequent location of drainage, followed by the right atrium (26%), and the pulmonary artery (17%) [17].

Coronary artery dilatation, mural thrombosis, rupture, aneurysm development, atherosclerotic deposition, calcification, intimal ulceration, and rupture are complications of coronary fistula [18]. CAF thrombosis may result in myocardial infarction, as well as atrial and ventricular arrhythmia [19]. The patient with coronary artery fistula is usually diagnosed with sign & symptoms of the patient and some diagnostic tools like ECG, Echocardiography, TEE, CT coronary angiography, Cardiac MRI, and Coronary angiography. Transesophageal echocardiography (TEE) gives origin, course, and termination of the fistula. So, if the surgery is indicated, TEE could be used to provide the exact site of drainage of coronary artery fistula [20]. But Coronary angiography remains the gold standard for the diagnosis of Coronary artery fistula (CAF) because it provides anatomic detail including the origin and termination of the coronary artery fistula [15].

Management of coronary artery fistulas (CAFs) are dependent on size, site of the fistula and whether the patient is symptomatic or not. Coronary artery fistula

can be treated by Conservatively and closure technique (surgical closure or transcatheter closure). The American College of Cardiology and the American Heart Association advocate close monitoring by echocardiography every 3 to 5 years for asymptomatic patients with small CAFs and encourage closure for extremely large CAFs regardless of the existence of symptoms due to the risk of arrhythmias, myocardial ischemia or infarction, and ventricular dilatation. Small or medium asymptomatic CAFs in adults can be closed, but serial echocardiography every three to five years is advised as a follow-up. Anticoagulation can be used to treat small CAFs, whereas aspirin and clopidogrel are needed to treat medium asymptomatic CAFs [21].

Surgery is indicated for symptomatic fistula, high-velocity CAFs, signs of volume overload, distal CAFs, proximal CAFs with large aneurysms, CAFs with tortuous courses or complex communications. Trans-catheter closure (coil embolization or occlude systems) is indicated for proximal CAF with single-site drainage, distal narrowing of the CAF with device access feasibility, non-tortuous vessels. Biorck and Crafoord first reported surgical ligation in 1947 as an effective method of fistula closure [22].

Yam, et al., have succinctly summed up the experience with cardiac fistulae for more than three decades in the management of coronary artery fistulae [23]. They outline three surgical treatment modalities.

Epicardial

The fistula is mobilized from the epicardial surface and then divided and ligated.

Endocardial

The fistula is identified from the lumen of the pulmonary artery or the interior of the cardiac chamber. The fistula tract is then completely obliterated with the closure of entry and/or exit points. Pericardial patch, Dacron patch or suture can be used for closure of the fistula tract.

Trans coronary

An arteriotomy is performed directly proximal to the CAF when the coronary artery near the fistula is dilated, and the fistulous orifice is oversewn from within the lumen.

For both symptomatic and asymptomatic patients at risk for future complications like bacterial endocarditis, thrombosis, distal embolization, aneurysm, dissection, rupture, premature atherosclerosis, pulmonary hypertension, myocardial ischemia or infarction, surgical intervention to close the fistula is advised [24-26].

Here the authors report a case of coronary artery to main pulmonary artery fistula, a rare defect managed surgically (by endocardial techniques) because of high flow, symptomatic (myocardial ischemia) fistula arises from proximal part of right coronary artery.

Conclusion

As a result of increasing experiences and techniques, surgical closure of coronary to pulmonary artery fistula is more effective than other techniques. For this type of fistula, surgical closure is the best treatment choice when used with the support of extracorporeal circulation and myocardial protection.

References

1. Holdright DR, Lindsay DC, Clarke D, Fox K, Poole-Wilson PA, Collins P. Coronary Flow Reserve in Patients with Chest Pain and Normal Coronary Arteries. *Br Heart J*. 1993;70(6):513-9. [PubMed](#) | [CrossRef](#)
2. Dodge-Khatami A, Mavroudis C, Backer CL. Congenital Heart Surgery Nomenclature and Database Project: Anomalies of the Coronary arteries. *Ann Thorac Surg*. 2000;69(3):270-97. [PubMed](#) | [CrossRef](#)
3. Zenooz NA, Habibi R, Mammen L, Finn JP, Gilkeson RC. Coronary Artery Fistulas: CT Findings. *Radiographics*. 2009;29(3):781-9. [PubMed](#) | [CrossRef](#)
4. Mirchandani, Sunil, and Colin KL Phoon. Management of Anomalous Coronary Arteries from the Contralateral Sinus. *Int J Cardiol*. (2005): 383-9. [PubMed](#) | [CrossRef](#)
5. Luo L, Kebede S, Wu S, Stouffer GA. Coronary Artery Fistulae. *Am J Med Sci*. 2006;332(2):79-84. [PubMed](#) | [CrossRef](#)
6. Gasser S, Bareza N, Gasser R, Klein W. Iatrogenic Coronary Fistula in Post-transplant Patients: Pathogenesis, Clinical Features and Therapy. *J Clin Basic Cardiol*. 2003;6(1):19-21.
7. Spicer DE, Henderson DJ, Chaudhry B, Mohun TJ, Anderson RH. The Anatomy and Development of Normal and Abnormal Coronary Arteries. *Cardiol Young*. 2015;25(8):1493-503. [PubMed](#) | [CrossRef](#)
8. Malagò R, Pezzato A, Barbiani C, Alfonsi U, Nicoli L, Caliarì G, et al. Coronary Artery Anatomy and Variants. *Pediatr Radiol*. 2011;41:1505-15. [PubMed](#) | [CrossRef](#)
9. Silverman FN, Landing BH. Abnormal Congenital Fistulous Communications of the Coronary Arteries. *Am J Roentgenol Radium Ther Nucl Med*. 1959;82:392-402. [PubMed](#)
10. Iglesias JF, Thai HT, Kabir T, Roguelov C, Eeckhout E. Transcatheter Coil Embolization of Multiple Bilateral Congenital Coronary Artery Fistulae. *J Invasive Cardiol*. 2010;22(3):142-5. [PubMed](#)
11. Kannan A, Lee JZ, Saleh L, Kanakadandi U, Rigatelli G, Lee KS, et al. How Should I Treat a Coronary Artery to Pulmonary Artery Fistula at an Acute Stent Thrombosis Site? *EuroIntervention*. 2017;13(11):1367-72. [PubMed](#) | [CrossRef](#)
12. Sunder KS, Balakrishnan KG, Tharakan JA, Titus T, Pillai VR, Francis B, et al. Coronary Artery Fistula in Children and Adults: A Review of 25 Cases with Long-term Observations. *Int J Cardiol*. 1997;58(1):47-53. [PubMed](#) | [CrossRef](#)
13. Friedman AH, Fogel MA, Stephens P, Hellinger JC, Nykanen DG, Tweddell J, et al. Identification, Imaging, Functional Assessment and Management of Congenital Coronary Arterial Abnormalities in children. *Cardiol Young*. 2007;17(S4):56-67. [PubMed](#) | [CrossRef](#)
14. Said SA, El Gamal MI, Van Werf TD. Coronary Arteriovenous Fistulas: Collective Review and Management of Six New Cases-Changing Etiology, Presentation, and Treatment Strategy. *Clin Cardiol*. 1997;20(9):748-52. [PubMed](#) | [CrossRef](#)
15. Liberthson RR, Sagar KA, 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. [PubMed](#) | [CrossRef](#)
16. Taleb MM, Sheikh MA, Cooper CJ, Tinkel JL. Multiple Coronary to Pulmonary Artery Fistulas: A Case Report and Review of Literature. *Cardiovasc Interv Ther*. 2012;27:127-30. [PubMed](#) | [CrossRef](#)
17. Papadopoulos DP, Perakis A, Votreas V, Anagnostopoulou S. Bilateral Fistulas: A Rare Cause of Chest Pain. Case report with Literature Review. *Hellenic J Cardiol*. 2008;49(2):111-3. [PubMed](#)
18. Sandhu JS, Uretsky BF, Zerbe TR, Goldsmith AS, Reddy PS, Kormos RL, et al. Coronary Artery Fistula in the Heart Transplant Patient. A Potential Complication of Endomyocardial Biopsy. *Circulation*. 1989;79(2):350-6. [PubMed](#) | [CrossRef](#)
19. Germain AS, Gabriel A, John A, Tubbs RS, Spicer D. Coronary Artery Fistula: A Review. *Cardiovasc Pathol*. 2015;24(3):141-8. [PubMed](#) | [CrossRef](#)
20. Iida R, Yamamoto T, Suzuki T, Saeki S, Ogawa Lida S. The Usefulness of Intraoperative Transesophageal Echocardiography to Identify the Site of Drainage of Coronary Artery Fistula. 2015;101:330-1. [PubMed](#) | [CrossRef](#)

21. Gowda ST, Forbes TJ, Singh H, Kovach JA, Prieto L, Latson LA, et al. Remodeling and Thrombosis Following Closure of Coronary Artery Fistula with Review of Management: Large Distal Coronary Artery Fistula-To Close or Not to Close? Catheter Cardiovasc Interv. 2013;82(1):132-42. [PubMed](#) | [CrossRef](#)
22. Biörck G, Crafoord C. Arteriovenous Aneurysm on the Pulmonary Artery Simulating Patent Ductus Arteriosus Botalli. Thorax. 1947;2(2):65. [PubMed](#) | [CrossRef](#)
23. Yim D, Yong MS, d'Udekem Y, Brizard CP, Konstantinov IE. Early Surgical Repair of the Coronary Artery Fistulae in Children: 30 Years of Experience. Ann Thorac Surg. 2015;100(1):188-94. [PubMed](#) | [CrossRef](#)
24. Jerbi S, Tarmiz A, Fradi S, Brahem A, Beltaifa M, Mlika S, et al. Coronary Artery Fistula: Case Report and Review of the Literature. Ann Cardiol Angeiol (Paris). 2008;58(4):236-9. [PubMed](#) | [CrossRef](#)
25. Ekonomou CK, Papadopoulos DP, Dalianis NV, Stratigis NG, Benos J, Votteas VE. Coronary Fistula from Left Main Stem to Main Pulmonary Artery. J Invasive Cardiol. 2003;15(10):600-1. [PubMed](#)
26. Luo L, Kebede S, Wu S, Stouffer GA. Coronary Artery Fistula. Am Med Sci. 2006;332:79-84. [PubMed](#) | [CrossRef](#)