

# Congenital Left Main Coronary Artery Fistula to Right Atrium : A Case Report

Fatemeh Vaziri , MD, Shahla Roodpeyma , MD, Manuchehr Hekmat , MD

## **Abstract**

Coronary artery fistulas are rare abnormalities with an estimated frequency of 0.002% in the general population. The majority of these fistulas arise from the right coronary artery. The left coronary artery is rarely involved. This study presents a 5-year old girl with a left coronary artery fistula to right atrium who underwent a successful surgical operation.

## **Introduction**

Coronary artery fistulas (CAF) are rare congenital anomalies; they connect a major coronary artery directly to a cardiac chamber, coronary sinus, superior vena cava or pulmonary artery. They constitute the most common form of hemodynamically significant coronary malformation with an incidence of 0.002% in the general population and 0.2% in patients undergoing coronary angiography. The most plausible explanation for a congenital CAF is the persistence of embryonic vascular sinusoids in the myocardium (1). The right coronary artery or its branches are the site of fistula in about 55% of cases; the left coronary artery in about 35%; and both coronary arteries in 5%. Over 90% of the fistulas drain into the venous circulation. These include right – sided chambers, pulmonary artery, superior vena cava and coronary sinus. Fistulas drainage occurs into the RV in (41%), RA in (26%), pulmonary artery in (17%), LV in (3%), and SVC in (1%). Most fistulas are single communications, but multiple fistulas have been reported (2). The myocardial blood flow is usually not compromised and shunt through the fistula usually is of small magnitude. A left – to – right shunt exists in more than 90% of cases. The majority of adult patients are usually asymptomatic. Unlike

adults a smaller percentage of pediatric patients are asymptomatic (2). Sometimes the blood in CAF bypasses the myocardial capillary network (steal phenomena) and symptoms range from mild shortness of breath to myocardial ischemia, angina, congestive heart failure, cardiac arrhythmia, rupture or dissection of fistula with or without cardiac tamponade and endocarditis (3,4). The acquired causes of coronary fistulas are rare and include atherosclerosis, takayasu arteritis, and trauma (2). Although selective coronary angiography is capable of identifying the origin of aneurysmal CAF, it is difficult to clarify the relation of the distal site of the CAF to other cardiac chambers because of dilution of a contrast medium or overlapping of adjacent structure. A multi – slice CT angiography might well become the modality of choice for the diagnosis of these rare congenital anomalies (5). It is well accepted that all symptomatic patients should be treated with surgical operation because the surgical risk in most cases appears to be considerably less than the potential development of serious and fatal complication (6).

The aim of this study is to present an asymptomatic pediatric patient with a rare type of CAF who underwent a successful surgical repair.



### Case Report

A 5-year old girl was presented to our clinic for evaluation of heart murmur. She was noted to have a heart murmur at birth which was not investigated. There were no special symptoms related to heart or any other organ in her past medical history. In physical examination the patient was acyanotic, well developed and well nourished. Weight = 16 kg, height = 110 cm, BP= 100/60 mm Hg. Chest auscultation disclosed a grade 3/6 continuous murmur at lower left sternal border. The first and second heart sounds were normal. Pulses were bounding. There was no abnormality on other organs. Chest X ray demonstrated moderate cardiomegaly involving right atrium, and increased pulmonary vascular marking. Electrocardiogram showed sinus rhythm, with normal intervals and axis. Transthoracic echocardiography images revealed abnormal flow into the right atrium (RA) but could not reveal the precise location of the drainage (Figure 1).

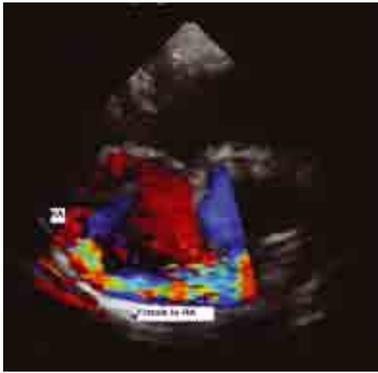


Figure 1- Echocardiography shows the turbulent flow in the right atrium

Cardiac catheterization and angiocardiography were performed for the patient. Courses of venous and arterial catheters were normal. Systemic samples were saturated. There was an O<sub>2</sub> step up of 16% at right atrium level. Pulmonary artery pressure was mildly elevated (mean PA pressure 31 mm Hg). Thoracic aorta injection at lateral view was normal. Left ventricular injection at LAO view showed no VSD. Aortogram at LAO and RAO views showed dilatation of left Valsalva sinus. It was suggested that a fistulous coronary artery arises from LMCA and drain to the right atrium (Figure 2). The multislice CT angiography (Brilliance 64, Philips Medical System) by intravenous injection of contrast medium was performed. The reconstruction of 3D volume – rendered (VR) images showed the following findings: Aortic valve had normal three cusps, followed by their normal Valsalva sinuses. Right coronary artery (RCA) had been arisen



Figure 2-Angiocardiography shows dilatation of the left main coronary artery

from right Valsalva sinus and was normal. The left main coronary artery (LMCA) was markedly dilated and its caliber was 10 mm. The LAD branch had normal course and caliber, measuring 1.6 mm. A dilated anomalous coronary artery had been arisen from left main coronary artery and showed tortuosity on its course. There was a 360° rightward turn of this artery. Then it passed below the right pulmonary artery, anterior to left atrium and right superior pulmonary veins. Above the left atrium it reached to a position just posterior to superior vena cava and vertically entered the right atrium. This was an abnormal coronary artery fistula between the left main coronary artery and right atrium. The most dilated part of this fistulous coronary artery was at its distal end (entrance point to RA) and measured 11 mm. The ascending aorta, aortic arch and descending thoracic aorta were of normal course and caliber. The right ventricular outflow tract, main pulmonary artery, right and left pulmonary arteries and their branches, as well as the four pulmonary veins (normally draining into the left atrium) were of normal caliber and appearance (Figure 3).



Figure 3- CT angiography shows the fistulous coronary artery originate from left main coronary artery and drained in the right atrium

The operation was performed through a standard median sternotomy using cardiopulmonary bypass. The RA was found to be dilated. After the pericardium was opened and the aorta was pulled up, the tract of fistula came into view. It was an aberrant artery separated from the left main coronary artery and passed from posterior side of pulmonary artery and aorta and traversed from the ceiling of left atrium and entered to the medial side of RA. The diameter of this aberrant artery was 5-6 mm and its length was 4-5 cm. two distal ends of fistula were clamped. After a pause of 15 minutes and measurement of O<sub>2</sub> saturation of right side heart the two distal ends of fistula were over sewn with 4-0 prolene suture and ligated.

The postoperative course of the patient was uneventful and she completely recovered after 3- day stay in postoperative ICU. In physical examination the machinery murmur was disappeared. Postoperative echocardiography was normal and showed no abnormal turbulent flow. The patient was discharged from hospital 6 days after admission. In the first postoperative follow-up she was well, with normal findings, and without complaint.

### Discussion

Coronary fistulas are connections between the coronary arteries and another cavity. The majority of these fistulas arise from the right coronary artery, and involvement of the left coronary artery especially its branches like left anterior descending coronary artery (LAD), and the circumflex coronary artery is rare. Our patient suffered from fistula of left main coronary artery. A significantly enlarged coronary artery can usually be detected by 2- dimensional echocardiography. Transthoracic echocardiographic imaging is more successful in children. Although noninvasive imaging may facilitate the diagnosis and identification of the origin and insertion of coronary artery fistulas, cardiac catheterization and coronary angiography are necessary for the precise delineation of coronary anatomy (2). Coronary angiography remains the gold standard for imaging the coronary arteries, but the relation of coronary artery fistulas to other structures, their origin and course may not be apparent and it is not always possible to reveal the complete delineation of CAF including its origin, course, and drainage (7). Multi – slice CT scanner can identify the precise location of the drainage site in their 3D-VR images and on the axial images (5). In our patient transthoracic echocardiography and cardiac cath-

eterization suggested the presence of CAF, and CT angiography clearly showed its origin, course and drainage site. The main indications for closure are clinical symptom especially of heart failure and myocardial ischemia, and in asymptomatic patients with high – flow shunting to prevent occurrence of symptoms or complications, especially in pediatric population (8). Surgical correction is safe and effective, with good results. The vast majority of the fistulas treated with catheter intervention were occluded with coils. Results from the transcatheter and surgical closure show that both approaches have similar early effectiveness, morbidity, and mortality. The safe and effective results of both approaches support the option for elective closure of clinically significant coronary artery fistulas in childhood (2). Surgical treatment of asymptomatic coronary fistulas is usually advised because of their propensity to become symptomatic or to cause complications. The most suitable surgical technique for treating CAF is the closure of both edges (9). Prognosis after successful closure of CAF is excellent. Long – term follow-up is essential due to the possibility of postoperative recanalization.

### References:

- 1) Akcay A, Yasim A, Koroglu S. Successful surgical treatment of giant main coronary artery fistula connecting to right atrium. *Thorac Cardiovasc Surg* 2009; 57: 489-495.
- 2) Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: Clinical and therapeutic considerations. *International Journal of Cardiology* 2006; 107 : 7-10.
- 3) Saezde Ibarra JI, Fernandez – Tarrío R, Forteza JF, Bonnin O. Giant coronary artery fistula between the left main coronary and the superior vena cava complicated by coronary artery dissection. *Rev ESP Cardiol* 2010; 63 (6) : 743 – 745.
- 4) Gandy KL, Rebeiz AG, Wang A, Jagers JJ. Left main coronary artery – to – pulmonary artery fistula with severe aneurysmal dilatation. *Ann Thorac Surg* 2004; 77 : 10810 – 03.
- 5) Shiga Y, Tsuchiya Y, Yahiro E, et al. Left main coronary trunk connecting into right atrium with an aneurysmal coronary artery fistula. *International Journal of Cardiology* 2008; 123 : 28 – 30.
- 6) Kamiya H, Yasuda T, Nagamine H, et al. Surgical treatment of congenital coronary artery fistulas : 27 years experience and a review of the literature . *J Card Surg* 2002; 17 : 173 – 7.
- 7) Iida R, Yamamoto T, Kondo N, et al. Identification of the site of drainage of left main coronary artery to right atrium fistula with intraoperative transesophageal echocardiography. *Journal of Cardiothoracic and Vascular Anesthesia* 2005; 13 (6): 777 – 780.
- 8) Balanescu S, Sangiorgi G, Castelvechio S, Medda M, Inglese L. Coronary artery fistula : clinical consequences and method of closure; a literature review. *Ital Heart J* 2001; 2 : 669 – 76.
- 9) Morgan RJ, Stephenson LW, Rashkind WJ. Fistula from left main coronary artery to right atrium : treatment by combined transaortic and transatrial approach. *International Journal of Cardiology* 1984; 6: 237 – 240.