

Congenital Coronary Artery Fistulas: Report of Three Rare Cases

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Abstract

Coronary artery fistula is a rare congenital anomaly with an incidence of about 0.2 - 0.6% in different reports. It is defined as a direct communication between the coronary artery and any surrounding cardiac chamber or vascular structure which bypasses the myocardial capillary bed. Three interesting cases of coronary artery fistula are reported. Two of the patients were symptomatic. In one case, all coronary arteries (in addition to duplicated LAD) were fistulized into the right ventricle. Diagnosis was made by echocardiographic study and coronary angiography. Surgical correction is discussed. In one case, angiography six months later showed no fistula. Serial echocardiography during follow-up was unremarkable (*Iranian Heart Journal 2008; 9 (1):64 -68*).

Key words: coronary artery fistula ■ congenital heart disease

Congenital coronary artery fistula was first described by Krause in 1865, but Swan and colleagues successfully performed the surgical correction for the first time in 1959.¹ With the extensive use of echocardiographic and angiographic studies, reported cases of coronary artery fistulas, even the symptomatic ones, have increased. In this report, the clinical and angiographic characteristics and surgical correction techniques of three interesting and rare cases are described. Between July 1993 and October 2001, three patients (2 male, 1 female) underwent surgical correction for their interesting forms of coronary artery fistulas at Ghaem Medical Center, Mashad University of Medical Sciences. Two cases were symptomatic and one of them was referred for a heart murmur.

Case 1

A 5-year-old boy complaining of easy fatigability was referred to a pediatric cardiologist. His medical and familial histories were unremarkable.

On physical examination, a continuous murmur (3-4/6 grade) was heard mostly at the left sternal border. Echocardiography showed a fistulous chamber at the apex draining to the right ventricle. Angiography revealed that all major coronary artery branches (posterior descending, PDA; obtuse marginal, OM; left anterior descending, LAD) are engorged and drained to a fistulous chamber at the apex.

This chamber was connected to the right ventricular cavity. Surprisingly, the LAD artery was duplicated (Fig. 1, A and B).

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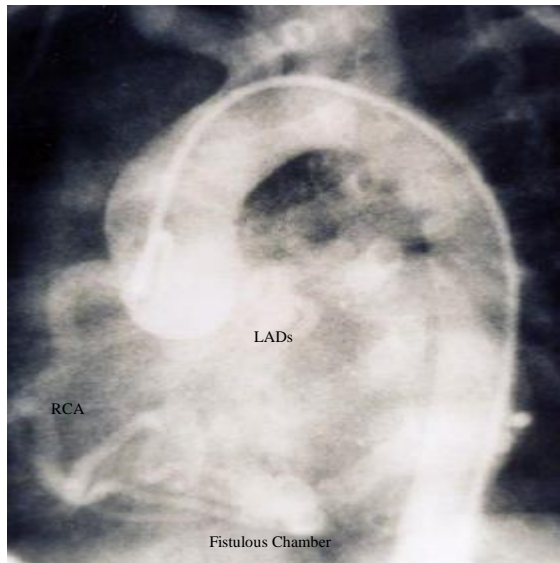


Fig. 1, A. Preoperative angiogram of case 1 showing duplicated left anterior descending, circumflex and posterior descending arteries entering a fistulous chamber located at the apex.

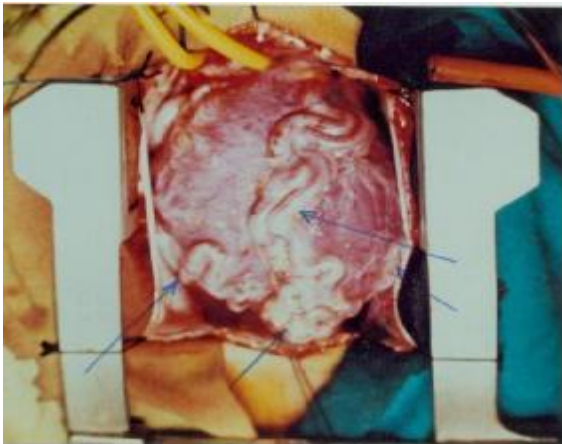


Fig. 1, B. Operative view of case 1, showing duplicated LAD, LCX, RCA and fistulous chamber. It should be mentioned that the apex of the heart was lifted by putting a sponge beneath the heart.

The patient was referred to us and underwent surgical correction with cardiopulmonary bypass, moderate hypothermia and cardioplegic arrest.

By opening the fistulous chamber, all four holes (arterial ends entering the chamber) were closed with 6-0 polypropylene suture reinforced in pericardial pledgets, and the

orifice of the chamber to the right ventricle was closed as well. Finally, the chamber was obliterated with two U-shaped 5-0 polypropylene sutures. Postoperative course was uneventful, without electrocardiographic and hemodynamic changes. He left hospital on the 6th postoperative day. Control echocardiography 6 months later showed no shunt.

Case 2

A 6-year-old boy was referred to our hospital for an evaluation of a continuous heart murmur. The medical and familial histories were unremarkable. In clinical evaluation and physical examination, a machinery murmur (grade 4/6) was detected at the right sternal border. Echocardiography disclosed a coronary artery fistula with the significant left to right shunt. Angiography showed a very engorged right coronary artery associated with a large branch which was drained to the right atrium (Fig 2, A and B).



Fig. 2, A. Preoperative angiogram of case 2 revealing a huge right coronary artery

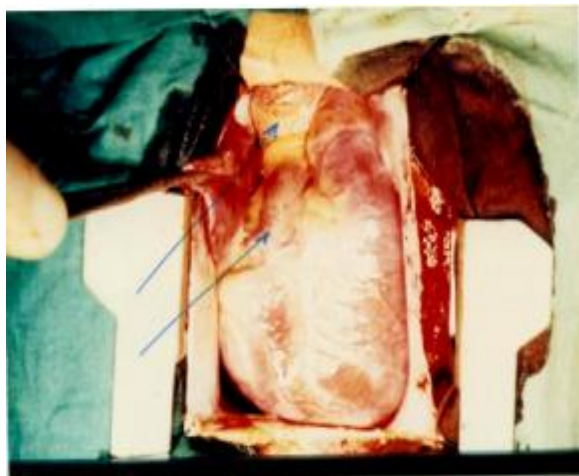


Fig. 2, B. Operative view of case 2, showing the engorged RCA before entering the right atrium

He was operated on with cardiopulmonary bypass, mild hypothermia and cardioplegic arrest. The fistulous arterial branch was followed, which drained just at the junction of the right and left atria. The orifice of this large artery was closed and obliterated by using pericardial pledget and 5-0 polypropylene via right atriotomy. Postoperative period was unremarkable without any electrocardiographic changes. He was discharged on the 5th postoperative day. Three months later, an echocardiographic study showed no shunt.

Case 3

A 17-year-old girl complaining of dyspnea on exertion was admitted to adult cardiology for investigation. On physical examination, a continuous murmur (grade 4/6) was heard at the left sternal border. Echocardiography and electrocardiography were unremarkable except for evidence of a fistula draining into the right ventricle. Angiography showed a large and tortuous LAD artery and two large branches dividing from the medial side of LAD passing the right ventricle and entering two fistulous chambers separately. The fistulous chambers entered the right ventricle (Fig. 3, A and B).

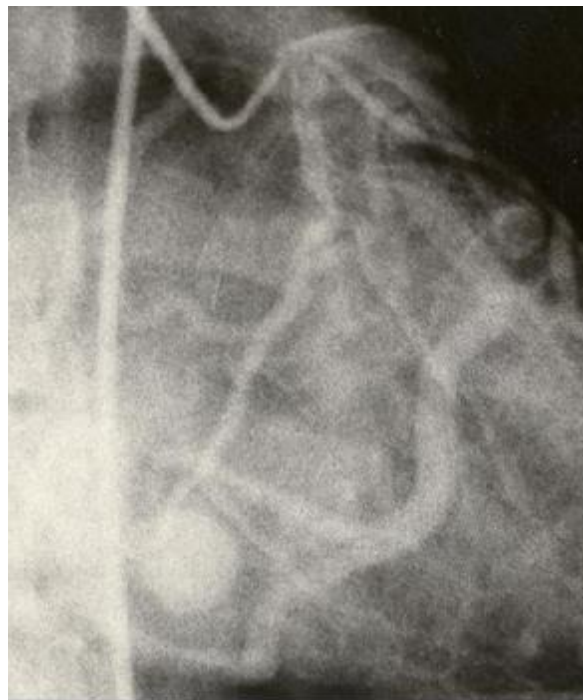


Fig. 3, A. Preoperative angiogram of case 3 showing proximal engorgement of LAD and its abnormal branches entering two separate chambers

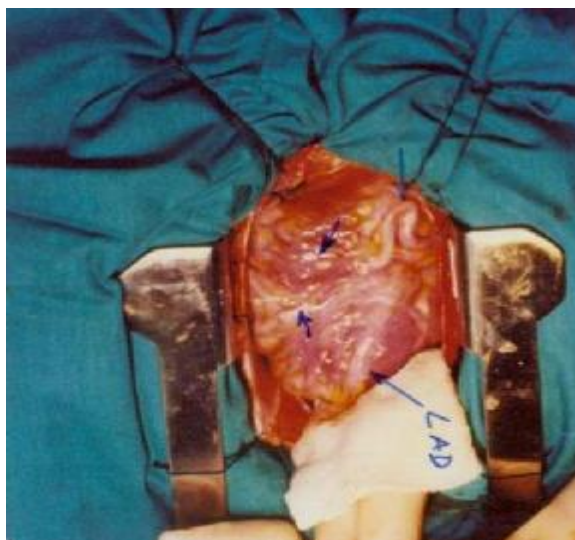


Fig. 3, B. Operative view of case 3, showing the abnormal branch of LAD with two fistulous chambers. The rest of LAD is abnormally small as shown by the arrow.

She was operated on using cardiopulmonary bypass and mild hypothermia. Both branches were ligated at the junction of the chambers. Two chambers were obliterated inside and outside the RV separately. She left hospital on the 6th postoperative day without any complication. Postoperative echocardiography three months later revealed no sign of fistula or shunt. Also, control angiography revealed no shunt six months after the operation.

Discussion

Congenital coronary artery anomalies are rare but can carry a significant risk of myocardial ischemia, myocardial dysfunction, congestive heart failure and sudden death.^{2, 11}

With the popular use of selective coronary angiography, increasing numbers of coronary fistulas have been diagnosed and treated. The reported incidence of the condition was 0.2% to 0.6% of all patients undergoing cardiac catheterization.¹⁴ The series report by Gillebert and associates³ is perhaps the largest with 14000 consecutive patients undergoing coronary angiographic studies: 19 coronary artery fistulas were identified, giving an incidence rate of 0.13%. Rate of aneurysmal formation of fistula is estimated at about 19%.⁸

Most patients considered for operation are asymptomatic and present either because of a continuous murmur or mild cardiomegaly and plethora on chest radiograph.^{4, 5, 9, 12, 13}

Symptoms include dyspnea on exertion and easy fatigability from left-to-right shunt. Angina pectoris is uncommon (about 7%), and myocardial infarction is rare (about 3%).^{4, 11}

It is postulated that these ischemic symptoms are due to coronary artery steal.

Heart failure occurs in 12% to 15% of patients presenting for operation but is much more common in older patients, as is angina^{11, 14} and in cases with fistulous connection to the coronary sinus, or with large shunts. Echocardiography and coronary angiography almost always confirm the diagnosis; but as reported by Hara and colleagues, in complex

coronary anatomy in which angiography could not make the precise diagnosis, the images from 16- slice multidetector row computed tomographic (MDCT) coronary angiography can be helpful in understanding the pathologic anatomy.¹⁰

In our patients, two were symptomatic, but both of them had continuous murmur. In addition to the echocardiographic study, for planning of either surgical repair or coil occlusion by interventional catheterization,¹⁴ all cases underwent selective coronary angiography. It is well accepted that all symptomatic patients should be treated with surgical ligation or closure, and the same applies to those with complications.⁴ For symptomatic patients, most surgeons will operate on those with a significant shunt because of the large volume load on the recipient's cardiac chamber.⁴

Our first case was very rare and probably unreported as of this date. As was mentioned, the duplicated LADs, RCA and CX took part in the fistulous phenomenon and entered the single fistulous chamber. The incidence of bilateral coronary artery fistula has been reported at less than 5%.⁶ In our second case, the RCA was huge and interesting in a 6-year-old boy. The third case also was very unusual, because two abnormal branches dividing from the medial side of the LAD entered the right ventricle via two separate fistulous chambers. We used cardiopulmonary bypass (CPB) intraoperatively because of the importance of the placement of the fistula in all our cases. By marking the precise location of fistula with a stitch before establishing CPB, in two cases with the fistulous chambers, the closure of the fistula and obliteration of chambers was performed. This maneuver was associated with opening the right ventricle and closing the internal hole with over-and-over sutures. In the case of RCA fistula to RA, repair was simple: the fistula and internal hole of RA were closed. Iida and colleagues described the role and usefulness of intraoperative transesophageal echocardiography in order to identify the site of the drainage of coronary

artery fistula and also assess the result of the surgical closure of the fistula as well.¹⁵

All our patients left hospital in good health. Liberthson and colleagues indicated a mortality rate of 4%⁵ in 173 patients. For one of our patients, postoperative angiography showed no evidence of shunt. All our cases have, however, been followed by serial echocardiography without any evidence of fistula up to the present time.

In conclusion, we reiterate that the surgical repair of symptomatic coronary artery fistula or asymptomatic cases with a significant shunt are mandatory and safe.¹⁶

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