Coronary Artery Fistulae

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Coronary artery fistulae and coronary cameral fistulae are rare anomalies that are discovered incidentally in patients undergoing coronary angiography. This article reviews the classification, management, and complications of these fistulae, and discusses a variety of presentations.

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KEY WORDS

Coronary artery fistulae • Coronary cameral fistulae • Coronary angiography

Coronary artery fistulae (CAF) are rare anomalies that are either congenital or acquired.[1] The connection may lie between one of the coronary arteries and a cardiac chamber, called coronary cameral fistula (CCF) or any segment of the systemic or pulmonary circulation (coronary arteriovenous fistula [CAVF]).[2] Clinically significant fistulae are the ones that cause severe left to right shunt.[1]

Incidence

The incidence of coronary anomalies was reported as 1.3% in a study by Yamanaka and Hobbs.[3] In this study, 1686 patients underwent coronary arteriography at the Cleveland Clinic Foundation (Cleveland, OH) from 1960 to 1988, of whom 13% had CAF (87% were found to have anomalies of origin and distribution). The results also showed other serious anomalies such as ectopic coronary origin from the pulmonary artery or from the opposite aortic sinus, a single coronary artery, and large coronary fistulae. Coronary anomalies are generally discovered incidentally at the time of catheterization, as most of them do not have any clinical signs or symptoms.

Classification

Congenital Fistulae

Primary congenital anomalies can be classified as hemodynamically significant or insignificant. Myocardial perfusion is affected in

hemodynamically significant primary anomalies of coronary arteries, whereas it is unaltered in hemodynamically insignificant anomalies. Hemodynamically significant anomalies occur in less than 1% of the adult population undergoing angiography.[4-7]

Acquired Fistulae

Some of the causes of acquired fistulae that have been reported in the literature include trauma, coronary artery bypass graft, atherosclerosis, and Takayasu arteritis.[8] A few other rare causes have been recognized, including hypertrophic and dilated cardiomyopathy, acute myocardial infarction, and permanent pacemaker placement.

Clinical Features

CAF is usually seen in the first decade of life with equal incidence among boys and girls.[9] Most patients are asymptomatic. Those with symptoms usually present with a continuous murmur; in such patients, fistulae terminate in the right heart chamber or the pulmonary artery and mimic a patent ductus arteriosus. Symptoms such as irritability, diaphoresis, pallor, tachypnea, and tachycardia may be suggestive of angina in infants.[9-12]

Morphology of Fistulae

More than 90% of CAF drain to the right side of the heart, but their origination from the right and left coronary arteries exists in almost equal proportion. Congenital CAVF involving both coronary arteries is rare. A precapillary fistula is considered to be the most common fistula connecting a major coronary artery to a cardiac chamber, coronary sinus, pulmonary artery, or superior vena cava.[5-7,9] According to a study by Levin and colleagues,[4] the frequency of fistulae arising from the right coronary artery accounts for approximately 50% of all cases, those from the left coronary artery account for 42%, and those from both arteries accounts for 5%. Most fistulae drain toward the venous side (90%); 41% drain to the right ventricle, 26% to the right atrium, and 16% to the pulmonary artery.[5-7,9]

CAF draining to the left atrium and left ventricle are rare.[13,14] Few case reports of generalized coronary arteriosystemic (left ventricular) fistula have been reported in the literature.[15,16] A case of four fistulae originating from three coronary vessels with an aneurysm draining into the left ventricle and the main pulmonary artery was reported,[17] and multiple case reports with associated valvular abnormalities have been reported as well.

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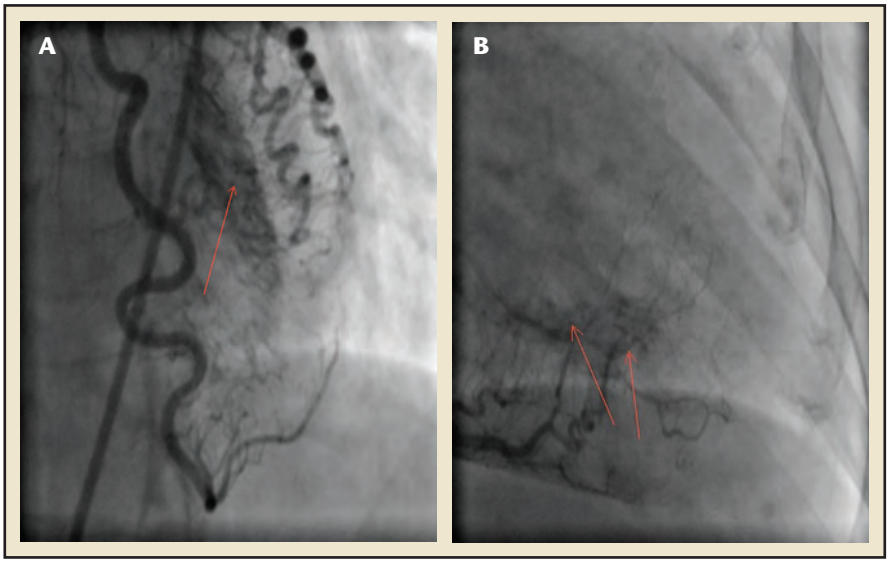
Diagnosis

CAF are mostly diagnosed by coronary angiography. Currently, improved results have been obtained by coronary computed tomography angiography, which delineates the course of these fistulae more accurately and noninvasively.[18] Other means of diagnosis include Doppler and two-dimensional echocardiogram,[19] myocardial contrast echocardiography (especially for identifying extremely small coronary fistulae),[20] and nuclear magnetic resonance imaging (especially in children).[21] Transesophageal echocardiography can have a corresponding role to angiography, as the exact location of the drainage site can be delineated.[22]

Coronary Cameral Fistulae

CCF are discovered incidentally in patients undergoing coronary angiography and are very rare. Most fistulae are single connection and drain into the venous circulation. Most patients with CCF present without symptoms. Different types of fistulae have been described, such as arterioluminal, arteriosinusoidal, and an arteriocapillary

Figure 1. (A) Coronary cameral fistula arising from the left coronary artery (*red arrow*). (B) Coronary cameral fistula arising from the right coronary artery (*red arrows*).



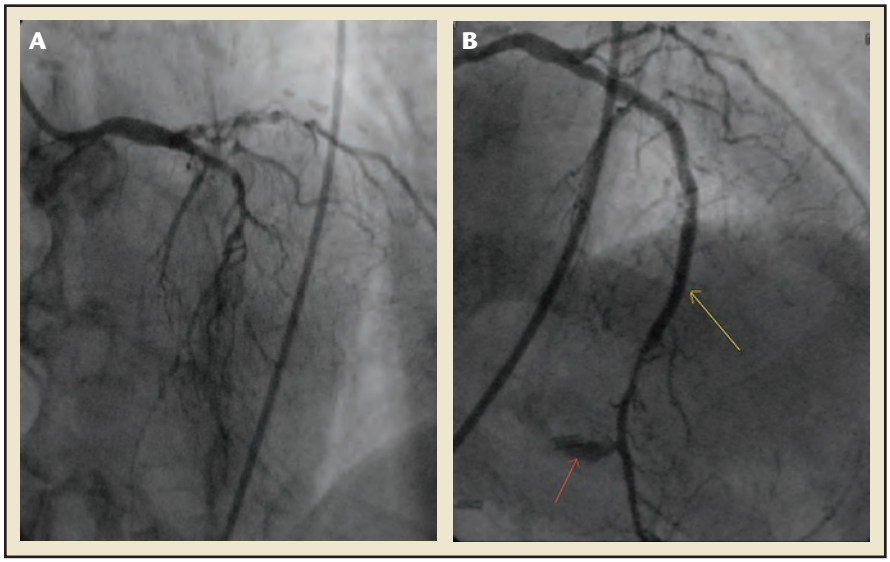


Figure 2. (A) A patient with a history of coronary artery bypass grafting who later underwent percutaneous coronary intervention. **(B)** Similar to the patient seen in **(A)**, this patient underwent percutaneous coronary intervention (*yellow arrow*) and showed a prominent coronary cameral fistula (*red arrow*). The etiology of congenital versus fistula secondary to coronary artery bypass graft was unclear. There is no evidence of pericardial effusion.

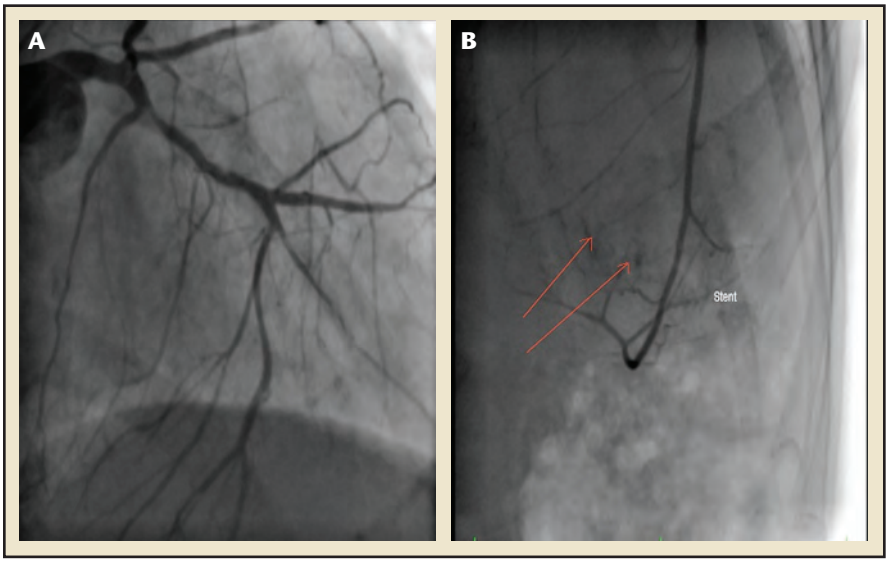


Figure 3. (A) A patient with significant stenosis in the left anterior descending artery. **(B)** A patient with coronary cameral fistula visible after stent placement (*red arrows*).

variant. In arterioluminal fistulae, a connection exists with the respective cardiac chamber, and in arteriosinusoidal fistulae, arterial blood connects through a sinusoidal network with the cardiac chambers.[23]

In the majority of the cases of CCF, the connection exists mostly with the right-sided chambers of the heart, with the remaining cases draining to the left-sided chambers, or both.[24] In one of the studies conducted by Roberts,[25] the majority of the fistulae were found to arise from either the right or left coronary artery, with a very small number of cases in which a connection arose from both coronary arteries. A case of CCF arising from both the coronary arteries is seen in Figure 1.

Complications associated with CCF include myocardial infarction,[26] congestive heart failure, arrhythmias, formation of aneurysm, and rupture of affected vessels.[27] One of the feared complications includes infectious endocarditis, which may occur due to turbulent blood flow, but this is not well proven. Older patients are usually at risk of such complications, because over a period of time there is an enlargement of the fistula due to abnormal hemodynamics. Sometimes the CCF becomes obvious after a chronically stenosed vessel is revascularized, as illustrated in Figures 2 and 3.

Management

It has been reported with good success that catheter-based closure of the fistulous connection is the nonsurgical treatment option for closure of coronary fistulas,[28-31] whereas ligation of the fistula is considered the surgical treatment of choice, as demonstrated by Björk and Crafoord in 1947[32]; this can be performed either from within the cameral chamber or externally. If coronary circulation has been compromised during the procedure, a coronary artery graft may be placed. Coil embolization, use of an AMPLATZER™ (St. Jude Medical, St. Paul, MN) vascular plug, and polytetrafluoroethylene-coated stent deployment are other treatment options that can be used.[33]

Coil embolization may be used in a non-CAF, as shown in Figure 4, in one of our 25-year-old patients with arteriovenous fistula in the right lung and massive hemoptysis. It was considered inoperable from a surgical standpoint. We successfully performed a coil embolization, as demonstrated in Figure 4.

If there is a need to close the fistula, it may be achieved by using an appropriately sized coronary balloon followed by a covered stent as needed. Figure 5 illustrates a case in which we tried to close a fistula

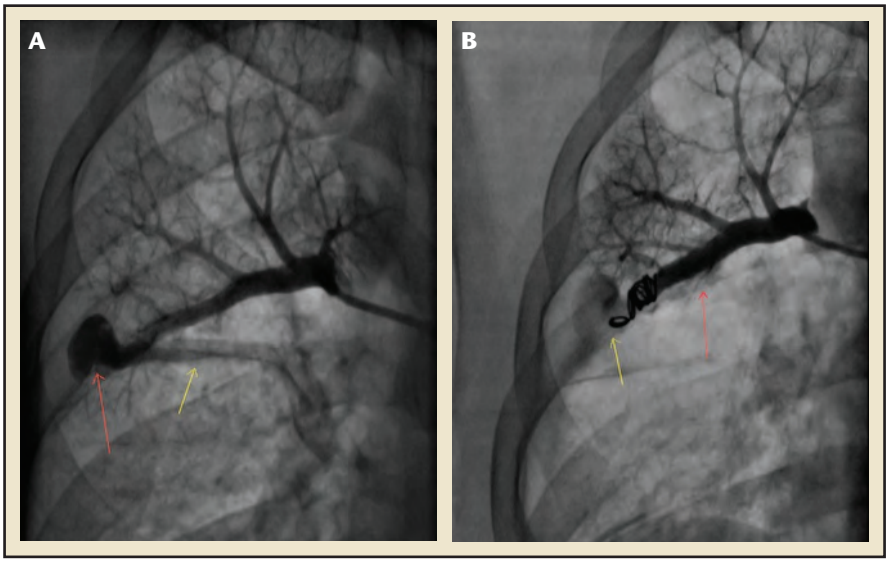
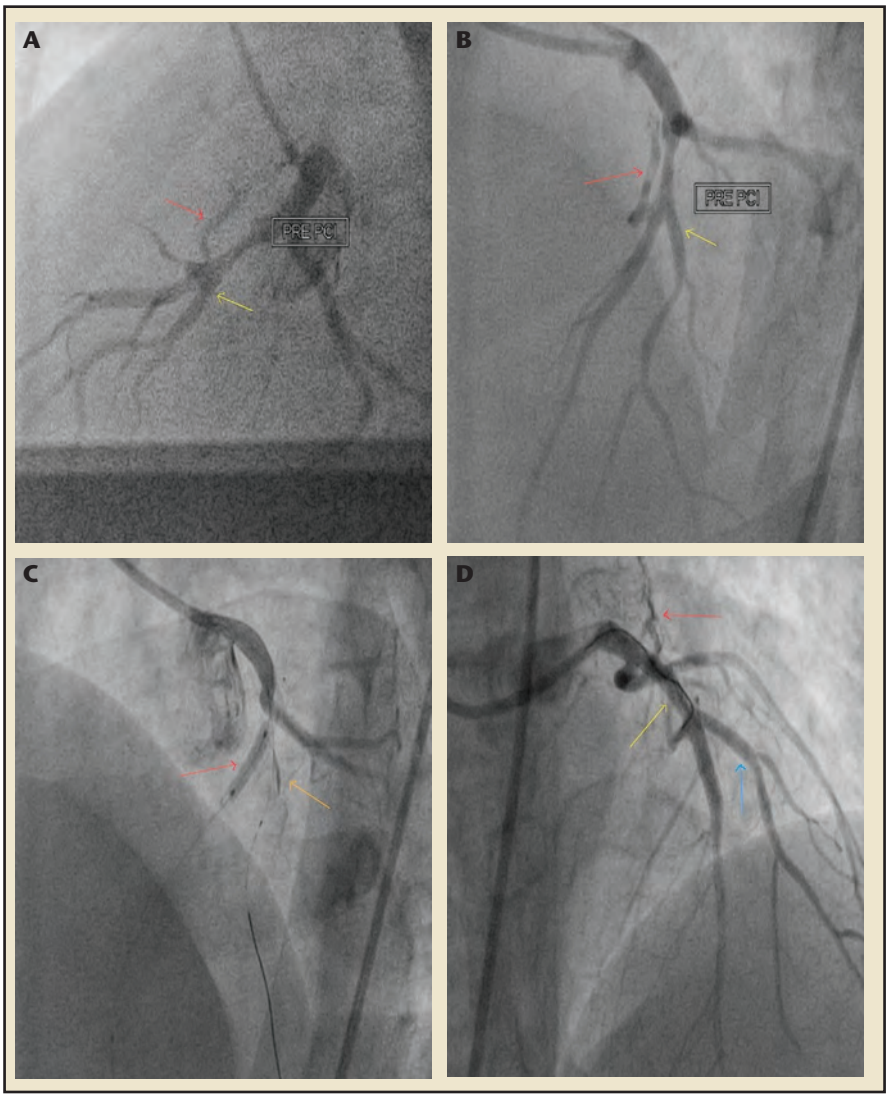


Figure 4. **(A)** Before coil embolization in an arteriovenous fistula in the right lung (*red arrow*). The venous phase of the fistula is also seen (*yellow arrow*). **(B)** After coil embolization in an arteriovenous fistula in the right lung showing obliteration of the venous phase of the fistula (*red arrow*) with near obliteration of the fistula (*yellow arrow*).



originating from the left anterior descending artery (LAD) after the patient was considered inoperable. However, just after balloon occlusion, a large diagonal branch was also compromised. This technique of checking with initial balloon occlusion enabled us to avoid the use of covered stent, and only a drug-eluting stent was used to treat a significant LAD lesion. This might otherwise have caused significant ischemia in the diagonal artery distribution.

Patients presenting with symptoms require closure, whereas in those without symptoms, no clear recommendations have been given. Noninvasive measures include close echocardiographic or angiographic follow-up in order to identify enlargement of the feeding vessel. Large fistulae have the propensity to dilate over time, and therefore increase risk of thrombosis, endocarditis, or even rupture. With this in mind, closure is advised for all but the small fistulous connections. Surgical ligation is the conventional management for large CAVF.

Prognosis

The prognosis of patients with symptomatic CAF is associated with considerable morbidity and mortality.[34] Valente and associates[35] reported a high risk of long-term morbidity after closure of CAF that primarily drain into the coronary

Figure 5. **(A)** A large fistula (*red arrow*) originating from the left anterior descending artery just opposite a large diagonal artery (*yellow arrow*). PRE PCI, before percutaneous coronary intervention. **(B)** A large fistula (*red arrow*) originating from the left anterior descending artery just opposite a large diagonal artery (*yellow arrow*). PRE PCI, before percutaneous coronary intervention. **(C)** Coronary fistula during balloon inflation (*red arrow*). Because there was a complete occlusion of a large diagonal branch (*yellow arrow*), a covered stent could not be deployed. **(D)** A drug-eluting stent (*yellow arrow*) was used to treat a significant left anterior descending artery stenosis, after which both the fistula (*red arrow*) and the diagonal branch (*blue arrow*) remained patent.

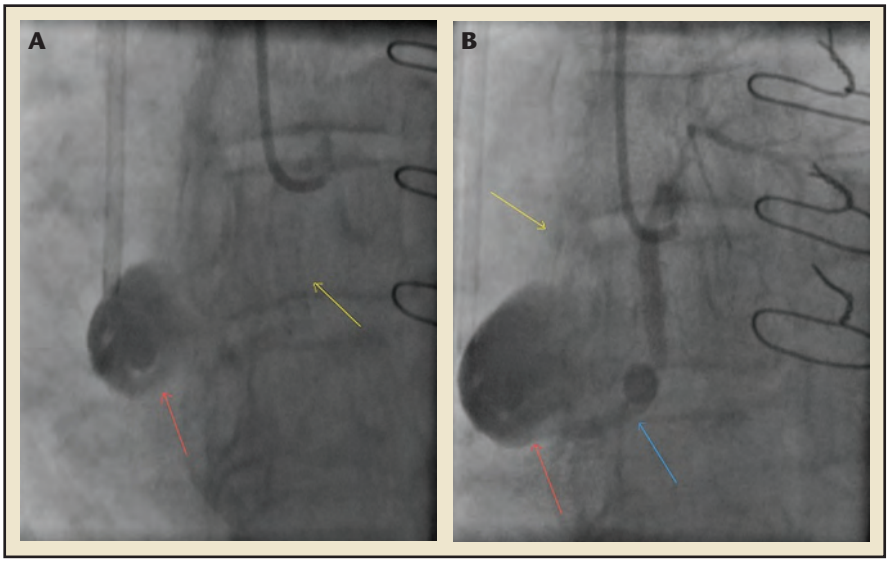


Figure 6. **(A)** Fistula secondary to right coronary artery stent infection (*red arrow*). The stent is also shown (*yellow arrow*). **(B)** Fistula secondary to right coronary artery (RCA) infection (*red arrow*). The connection between the RCA and the fistula is shown (*blue arrow*). A dialysis catheter is also shown (*yellow arrow*).

sinus; long-term medical intervention such as anticoagulation therapy should be considered in these patients.

Complications

One of the most common complications of CAF is cardiac failure. Approximately 75% of CAF patients who present in the fourth decade of life have symptoms of cardiac failure.[9] Bacterial endocarditis is also a known complication of CAF,[36] in addition to rupture of an aneurysm of a fistula,[37] pericardial hematoma,[38] angina, myocardial ischemia, and arrhythmia. Figure 6 demonstrates a case of a CAF secondary to an infected right coronary artery stent.

Conclusions

CAF are uncommon but serious entities that can have serious implications on the long-term health of patients. Proper diagnosis and treatment can avoid serious complications. Percutaneous treatment may offer a simpler and noninvasive mode of treatment in select patients. ■

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MAIN POINTS

- Coronary artery fistulae (CAF) and coronary cameral fistulae (CCF) are rare cardiac anomalies that are discovered incidentally in patients undergoing coronary angiography. They are either congenital or acquired.
- CAF are mostly diagnosed by coronary angiography. Currently, improved results have been obtained by coronary computed tomography angiography. CCF are discovered incidentally in patients undergoing coronary angiography and are very rare; most patients with CCF present without symptoms.
- The prognosis of patients with symptomatic CAF is associated with considerable morbidity and mortality. One of the most common complications of CAF is cardiac failure.
- Catheter-based closure of the fistulous connection is the nonsurgical treatment option for closure of coronary fistulas; ligation of the fistula is considered the surgical treatment of choice. Patients presenting with symptoms require closure; however, no clear recommendations have been given regarding asymptomatic individuals. Noninvasive measures include close echocardiographic or angiographic follow-up in order to identify enlargement of the feeding vessel.

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