Coronary Artery Fistulae

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ABSTRACT: Coronary artery fistulae are abnormal communications between a coronary artery and a cardiac chamber or major vessel (vena cava, pulmonary veins, pulmonary artery). They are usually diagnosed by coronary arteriography. Clinical presentations are variable depending on the type of fistula, shunt volume, site of the shunt, and presence of other cardiac conditions. In this article, we review the literature regarding etiology, incidence, clinical manifestation, image studies, and management. [Am J Med Sci 2006;332(2):79–84.]

It is commonly believed that W. Krause, a German anatomist, first reported a fistulous connection between a coronary artery and a cardiac chamber in 1865. In fact, the Austrian anatomist Josef Hyrtl had already described anomalous coronary arteries with fistulae in 1841.1 The distinguished Canadian physician Maude Abbott described further the pathology of this phenomenon in 1906.

The majority of coronary artery fistulae (CAFs) are congenital and not gender-specific. The anomaly accounts for only 0.4% of congenital heart defects2 and approximately 50% of pediatric coronary vasculature anomalies. Its incidence in the overall population is estimated to be about 0.002%. Twenty percent of people with congenital CAFs have other concomitant cardiac anomalies, most frequently aortic and pulmonary atresia and patent ductus arteriosus. CAF with tetralogy of Fallot has also been reported.3

Congenital fistulae often arise from the right coronary artery system, and the majority of them enter into the right ventricle (Figures 1A and 1B), right atrium, superior vena cavae, coronary sinus, or pulmonary arteries. Fistulae draining into the left heart are uncommon (Figure 2). In a review of all patients who underwent coronary arteriography between 1971 to 1981 at the Cleveland Clinic Foundation, a total of 122 fistulae were identified. Of those, 17% drained into the left ventricle and 6% drained into the left atrium.4 Rittenhous et al.5 reviewed 171 cases of surgical correction of fistulae and found that only 6% terminated in the left atrium or left ventricle. The sites of drainage from different studies are listed in Table 1.

Generally, CAF manifests as a single fistula. Multiple CAFs are rare, with only approximately 20 cases reported as generalized arteriosystemic fistulae originating from all three major coronary vessels.6 Although the etiology is unclear, polymyositis has been reported to be associated with multiple CAFs.7

Coronary artery fistulae can also be acquired due to penetrating and nonpenetrating chest trauma and iatrogenic causes. Traumatic CAF is most common between the right coronary artery and the right side of the heart.4 Gasser et al.1 reviewed iatrogenic coronary fistulae in post-transplant patients. Post-endomyocardial biopsy fistulae occur in 8% of transplant patients, resulting in a single CAF or, rarely, multiple CAFs.8 There are also case reports of iatrogenic CAF related to percutaneous coronary intervention, coronary artery bypass graft surgery, mitral valve replacement, septic myocarditis, closed-chest ablation of accessory pathway, permanent pacemaker placement, transbronchial lung biopsy, and acute myocardial infarction.9–9 Although these are reported to be rare causes, it is likely that the true incidence is underestimated.15

Clinical Presentation

The clinical presentation of CAF varies depending on factors such as the age of the patient, the amount of flow, the resistance of the recipient chamber, and development of myocardial ischemia. It is commonly believed that the majority of adult patients are asymptomatic and that the lesion is incidentally detected on routine examination or is an incidental finding during coronary angiogram. But according to several study series as shown in Table 1, about half of the patients are symptomatic at the time of presentation. Liferthson et al.20 reported on 187 patients with CAF: symptoms occurred in 19% of young patients (≤20 years) and in 63% of adults (≥20 years). In Rittenhous’s study of 171 patients,9 41% had symp-
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Figure 1. Multiple CAF in a patient. Angiogram of the right coronary artery in an anteroposterior (AP) projection with cranial angulation shows a fistula originating in the distal artery and communicating with the right ventricle. A left coronary artery angiogram in an AP projection shows a fistula originating from left anterior descending coronary artery that communicates with the right ventricle.

Figure 2. Coronary fistula from left anterior descending (LAD) artery to left ventricle.

toms, with the most common symptom being dyspnea at rest or on exertion (19%). The average age for the development of symptoms was 18 years. Demirkilic et al.21 reported on 21 patients with CAF with a mean age of 37 years. Dyspnea was the most common symptom (29%).

In addition to dyspnea, which is the most common symptom, other symptoms and sequelae have been reported with CAF. These include fatigue, congestive heart failure, pulmonary hypertension, rupture or thrombosis of the fistula or an associated arterial aneurysm, atrial arrhythmia, upper respiratory infection, hemoptysis, edema, palpitations, pneumonia, and paroxysmal nocturnal dyspnea. Angina pectoris is rare in the absence of arteriosclerotic coronary artery disease. In patients who experience angina, the vast majority are older than 40 years of age despite the assumed congenital origin of the malformation.22 Patients with dual CAFs presenting with angina pectoris usually have other associated cardiac disease, such as significant coronary atherosclerosis, aortic stenosis, or hypertrophic obstructive cardiomyopathy.23,24

Pericardial effusion or sudden death as the initial manifestation of CAF is rare but has been reported.25–27 Dichtl et al.27 reported a young woman who survived a cardiac arrest due to myocardial ischemia with subsequent ventricular fibrillation caused by thrombosis of a CAF deriving from the left main coronary artery. There are also reports of sudden death resulting from rupture of aneurysm.28,29

Physical Examination

The classic physical finding in patients with CAF is a soft, continuous murmur that tends to be crescendo-decrescendo in both systole and diastole but louder in diastole. The murmur is often confused with other conditions, such as patent ductus arteriosus, ventricular septal defect with aortic valve incompetence, aortopulmonary window, or sinus of Valsalva fistula. However, most of the other continuous murmurs reach their peak intensity at the time of the second heart sound. In the case of left coronary artery to left ventricle fistulae, blood flow through the shunt occurs exclusively in diastole with a murmur similar to that heard in aortic insufficiency.30 The location on the chest wall where the murmur is the loudest depends on where the fistula enters the heart. Of note, the sudden occurrence of continuous murmur is virtually diagnostic of iatrogenic post-biopsy fistulae.

The prevalence of a continuous murmur varies in different reports depending on the population studied. A continuous murmur was found in 3% (3 of 101) of adult patients with CAF identified at the time of coronary angiography4 and in 9% (3 of 31) of children with an echocardiographic finding of a clinically silent CAF.31 In contrast, in a group of patients undergoing surgical intervention, Cheung et al. reported that all 41 patients with CAF had a grade 4 to 6 continuous murmur on auscultation.32

Natural History

Fifty percent of patients with large fistulae or multiple fistulae develop complications. Some of these complications can be attributed to the additive effect of other comorbid diseases. Complications include bacterial endocarditis, thrombosis, distal embolization,
aneurysm formation, dissection, rupture, premature atherosclerosis, pulmonary hypertension, myocardial ischemia, or infarction. Bacterial endocarditis occurs in 5% to 10% of cases. Most reported cases are caused by Streptococcus viridans.33,34 Penicillin-resistant Streptococcus mitis was reported as the cause of endocarditis in an asymptomatic 12-year-old boy with CAF.35

The natural history of CAF is variable, with long periods of stability in some and sudden onset or gradual progression of symptoms in others. Spontaneous closure of CAF is uncommon but has been reported. Schleich et al.36 reported seven cases of spontaneous closure of CAF and reviewed 13 cases of spontaneous closure in the literature. Spontaneous closure is likely to occur in infants younger than 2 years if the CAF drains into the right heart, especially the right ventricle.

**Diagnostic Studies**

Coronary angiography remains the criterion standard for the diagnosis. Angiography defines the coronary artery involved, the recipient cardiac chamber, and the site of communication. Recently, transthoracic echocardiography, transesophageal echocardiography, and echo-Doppler have also been employed. Schleich et al.36 first described patients whose follow-up by color Doppler echocardiography permitted monitoring of spontaneous CAF closure by noninvasive techniques. Combined two-dimensional and pulsed Doppler echocardiography demonstrates a dilated coronary artery, turbulent flow in the fistula and the recipient chamber. Transthoracic color Doppler echocardiography with a high frequency transducer and a low Nyquist limit allows for multiple coronary artery–left ventricular microfistulae to be visualized.37 Transesophageal echocardiography is used intraoperatively to identify the precise location of the site of drainage of the fistula, which could not be accurately revealed with preoperative coronary angiography.38 Magnetic resonance imaging and multidetector computed tomography also have become alternative methods to evaluate the anatomy, flow, and function of CAF.39–41

In the vast majority of cases, the amount of shunting compared to the total cardiac output is small and cannot be detected by oxygen step-up during right-side catheterization. Hemodynamic quantification of shunt flow has also been estimated by measurements of coronary artery flow velocity using intravascular Doppler ultrasonography, which provides further insight into the pathophysiology of CAF.42

**Treatment**

Bjork and Crafoord, in 1947, first successfully performed surgical closure of a CAF draining into the pulmonary artery in a patient with a preoperative diagnosis of patent ductus arteriosus. Most symptomatic patients and/or patients with complications from a CAF are treated with surgical ligation or closure. For asymptomatic patients, indications for surgery are similar to those for other left to right shunts (e.g., Qp/Qs > 1.5 or right ventricular volume overload). The majority of small, asymptomatic CAFs in adults do not need surgical intervention. For larger shunts and younger patients, treatment of asymptomatic fistulae remains controversial. Several studies recommend surgery because of risk of future complications.43–46 Macri et al.45 believe that prophylactic surgical closure of the fistula during the pediatric age period (preferably in the preschool period) is the treatment of choice. Based on 28 patients with CAF treated in a 20-year span at Duke University as well as review of additional 258 patients reported in the literature, Lowe et al.43 recommended surgical correction to prevent the development of complications.

Other reports4,32,39,47 describe successful conservative treatment in selected patients with CAF. Jebara et al.47 suggest that coronary artery to left ventricle fistulae tend to close spontaneously or to regress significantly with time and may not require surgical treatment. Cheung and his colleagues92 conclude that for asymptomatic patients with mild shunting, the benefits of surgery are uncertain and there is a potential, although small, risk involved. They recommend regular follow-up and surgical correction if symptoms develop or if the amount of shunting increases.

<table>
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<th>Study</th>
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<th>RV, %</th>
<th>SVC/RA/CS, %</th>
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<th>LV, %</th>
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RV, right ventricle; SVC, superior vena cava; RA, right atrium; CS, coronary sinus; PA, pulmonary artery; LA, left atrium; LV, left ventricle; NA, not available.
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For the treatment of traumatic CAF and iatrogenic CAF, early operative intervention is preferable to delayed intervention. Patients being managed conservatively may develop life-threatening complications.\textsuperscript{15,48}

Operative techniques vary based on the involved coronary artery, the recipient cardiac chamber and the exact site of communication. The usual form of correction is direct ligation performed at the point of entry into the cardiac chamber to avoid the development of myocardial ischemia.\textsuperscript{5} Graffing the distal coronary vessel involved is also recommended to ensure optimal coronary blood flow when direct ligation could cause compromise of distal coronary blood flow to the myocardium.

In 1983, Reiday and colleagues first successfully performed transcatheter closure of CAF.\textsuperscript{49} Since then, a variety of materials have been used, including Gianturco coils, covered stainless-steel coils, detachable balloons, coaxial embolization with platinum micro-coils, double-umbrella devices, the Gianturco Griftka vascular occlusion device, and Amplatzer duct occluder.\textsuperscript{49–58} The selection of occlusion device is based on the anatomic features of the fistula.\textsuperscript{59} Coils have been used in small CAFs, and double-umbrella devices have been used in large CAFs. This procedure avoids the need for surgical intervention, cardiopulmonary bypass, and median sternotomy. Mavroudis et al.\textsuperscript{60} recommend elective coil occlusion in patients who satisfy the following criteria: absence of multiple fistulae, a single narrow drainage site, absence of large branch vessels, and safe accessibility to the coronary artery supplying the fistula. Transcatheter embolization has been associated with uncommon complications, such as transient ischemic electrocardiographic changes, arrhythmias, device embolization, and myocardial infarction.\textsuperscript{61} as well as one reported procedural death due to device recoil into the left main coronary artery.\textsuperscript{62} Close long-term follow-up with coronary angiography and myocardial scintigraphy after transcatheter or surgical closure of CAF is very important, to recognize possible recanalization and complications such as left ventricular aneurysm or myocardial infarction.\textsuperscript{63,64}

Medical management after complete CAF occlusion remains controversial in the literature. Most studies do not suggest anticoagulation treatment. However, in view of the persistence of the fistulous cul-de-sac, coronary artery dilation, and potential for thrombus formation (which may lead to myocardial infarction), aspirin may be indicated. For severe coronary artery dilatation (>10 mm), some authors advocate anticoagulation with warfarin.

Medical management of patients with angina due to CAF is similar to management of angina in the absence of CAF. Beta-blockers or calcium channel blockers are usually recommended. Nitrates must be used cautiously, as they can cause dilation of the fistula and decreased end-diastolic pressure of the recipient ventricle, both of which can lead to increased shunt flow and coronary “steal”.

Prophylaxis for bacterial endocarditis is recommended in all CAF patients and in patients after complete fistula occlusion for at least 1 year.\textsuperscript{65}

Conclusion

Coronary artery fistula is a rare coronary artery abnormality that is difficult to detect clinically because of nonspecific manifestations. Most CAFs are benign but some may produce symptoms that can be life threatening. Coronary angiography demonstrating CAF anatomy is crucial to the management of patients with this condition. Surgical correction is indicated in patients who are symptomatic or those in whom there is a significant left-to-right shunt.

References

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