The Current Status of Coronary Artery Fistula

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Abstract

Coronary artery fistula (CAF) is an unusual coronary anomaly. Most coronary artery fistulas are congenital, but the incidence of acquired CAFs is increasing following the incremental use of intravascular procedures and interventional techniques. The prevalence of CAF is about 0.1-0.8% based on coronary angiography or echocardiography studies. CAFs originate mostly from the right coronary artery and the left anterior descending artery and have proximal involvement. Most of them drain into the right atrium, right ventricle and pulmonary artery. Few of them drain into the left ventricle or left atrium. According to the site of drainage, CAFs have varied physiologic presentations. Clinically, about half of patients with a CAF are asymptomatic. Large fistulas can induce congestive heart failure, angina, myocardial infarction, arrhythmia or pulmonary hypertension. Endocarditis, coronary artery rupture and sudden cardiac death have also been reported. Physical examination usually reveals a continuous murmur. Myocardial ischemia with abnormal ^{201}Tl perfusion image can be detected in large portion of patients with CAF. Coronary angiography is the major diagnostic tool. Cardiac echocardiography, magnetic resonance imaging and multidetector computed tomography are also used for diagnosis. Treatment includes medical therapy, transcatheter closure of the fistula or surgical ligation of the fistula. However, these treatments should be tailored according to the size and location of fistula, and the patient's age and clinical presentation. The characteristics of CAFs in Oriental people were also discussed in this article.（J Intern Med Taiwan 2009; 20: 484-489）

Key Words：Coronary artery fistula, Coronary anomaly, Angina pectoris

Introduction

Coronary artery fistula (CAF) is an unusual vascular anomaly that communicates between one of the coronary arteries and a cardiac chamber or a large thoracic vessel. The first reported case of a CAF was in 1865 by Krause[1]. Its prevalence is about 0.1-0.8% based on coronary angiography and echocardiography studies[2-5]. The true incidence is difficult to evaluate because about half of the cases may be asymptomatic and clinically undectable until an echocardiogram or catheterization is performed. CAFs comprise 14% of congenital coronary artery anomalies[4], and represent 0.4% of all congenital cardiac malformations[6]. Approximately 10-30% of patients with a CAF also have another congenital cardiovascular anomaly[7,8]. The most commonly seen defects include variations of tetralogy of Fallot, patent ductus areteriosus, atrial septal defect, ventricular septal defect and pulmonary stenosis. The majority of CAFs arise

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from the right coronary artery and the left anterior descending artery; the circumflex coronary artery is rarely involved. They most frequently drain into the right ventricle, right atrium or pulmonary artery. Few of them drain into the coronary sinus, left ventricle, left atrium, pulmonary vein or superior vena cava. Generally, most CAFs manifest as a single fistula; cases of multiple fistulas are rare. CAFs can be found at any age and are not gender-specific.

Pathophysiology

According to the site of drainage, CAFs have varied physiologic presentations. Latson et al. described the physiology of CAFs in detail[9]. CAFs that drain into the right side of the circulation create a left-to-right shunt of oxygenated blood back to the pulmonary circulation. Those that drain into the systemic veins or right atrium have a circulatory physiology similar to an atrial septal defect. Those that drain into the right ventricle have a circulatory physiology similar to a ventricular septal defect. Those that drain into the pulmonary arteries are similar hemodynamically to a patent ductus arteriosus. Chronic volume overload of the right heart may lead to pulmonary arterial hypertension. However, there are no literature reports of Eisenmenger's syndrome caused by untreated CAFs. A coronary fistula that drains into the left atrium does not result in a left-to-right shunt, but rather causes a volume load similar to mitral regurgitation. Furthermore, a coronary fistula that drains into the left ventricle produces hemodynamic changes similar to aortic insufficiency.

Occasionally CAFs may exist along with an atherosclerotic stenosis of the same coronary artery. Both disorders can induce myocardial ischemia[10-12]. The location of the coronary fistula with respect to the coronary stenosis may play a significant role in the pathophysiology of myocardial ischemia[13]. In most published reports, the fistula is located proximal to the coronary stenosis. In other cases, the fistula is located distal to the stenosis and may associate with a right-to-left shunt if a significant drop occurs in the coronary arterial pressure distal to the coronary lesion. The mechanism involved in the pathophysiology of ischemia differs depending on the location of the fistula with respect to the stenotic lesion. In the first case, ischemia occurs because of the coronary steal phenomenon associated with decreased coronary flow through the stenosis. In the second situation, ischemia is induced because of a reduction in coronary flow through the stenosis and a right-to-left shunt that allows deoxygenated blood from the right sided circulation to enter the coronary artery.

Etiology and Classification

In the past, CAFs are almost reported as congenital in origin. Congenital fistulous connections between the coronary system and a cardiac chamber appear to represent persistence of embryonic intertrabecular spaces and sinusoids[14-17]. Acquired CAFs have been reported with an increasing frequency in recent years. It is possible that this increase may be due to an incremental use of intravascular procedures and interventional techniques. Acquired CAFs have been reported as a complication of one the following conditions: deceleration accident, percutaneous transluminal coronary angioplasty, endomyocardial biopsy, implantation of permanent ventricular pacing leads, coronary artery bypass graft surgery, mitral valve replacement, septal myectomy or acute myocardial infarction[18-20]. Seventy-six patients (1985-1995) with 96 CAFs were identified from a review of the literature by Said et al.[18]. They reported a congenital origin in 64% of these 76 cases and an acquired cause in 36%.

Latson et al.[9] proposed a way to classify the size of CAFs. They consider that trivial or small fistulas result in little or no dilation of the proximal

coronary artery from which they arise, and are themselves no larger at any point than twice the normal expected proximal coronary artery diameter. Fistulas that, at any point, are larger than two times but less than three times the expected proximal normal coronary artery diameter, or that are associated with a similar range of dilation of the proximal associated coronary artery, are considered to be medium-size fistulas. Fistulas that are more than three times the proximal coronary artery diameter are considered large. This classification may be useful in making clinical decisions.

Clinical Manifestation

Most CAFs are found incidentally during cardiac catheterization. The clinical manifestations vary according to the size of the fistula, drainage site and patient's age. About half of them are asymptomatic, but angina, myocardial infarction, heart failure, arrhythmia, endocarditis and aneurysm rupture have been reported. Said et al. reported that in 76 patients with CAFs, 55% of the patients were asymptomatic, 34% had chest pain, 13% had congestive heart failure and 1% had arrhythmia[18]. Symptoms and complications of CAFs are less common in children but more significant in adults. In the literature review and analysis by Liberthson et al. in 1979, symptoms occurred in only 19% of young patients (＜20 years), whereas 63% of patients over the age of 20 had either symptoms or a complication due to the fistulas[21].

The most common physical finding is a heart murmur. The typical murmur of a moderate or large CAF is continuous murmur that tends to be crescendo-decrescendo in both systole and diastole, louder in diastole, however. Differential diagnosis includes persistent ductus areteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, ventricular septal defect with aortic valve incompetence, aortopulmonary window, and systemic arteriovenous fistulas. Isolated systolic or diastolic murmurs have occasionally been reported[22]. The site of maximal intensity of the murmur is related to the site of drainage. Signs of pulmonary plethora and cardiomegaly at X-ray and ECG signs of right ventricular volume overload could be noted if there is large volume of flow through a CAF that produces a left-to-right shunt. Myocardial ischemia with abnormal ^{201}Tl perfusion image can be detected in large portion of patients with CAF[23]. The absence of ^{201}Tl perfusion defect in patients with CAF may be due to micro-fistula without evident steal phenomenon of coronary blood flow.

Coronary angiography is the major diagnostic tool. It can demonstrate the size, anatomy, number, origination and termination site of the fistulas. Cardiac echocardiography is also useful for diagnosis[24-26]. Magnetic resonance imaging and multi-detector computed tomography are also used to evaluate the anatomy, flow and function of CAFs[27-29].

Racial Differences

Most CAFs were reported from studies in Caucasian people. Chiu et al. presented the first report of Oriental CAF patients with long-term follow-up and with a large number of patients[30]. From September, 1992 to August, 2007, 152 CAFs were detected in 28210 coronary angiograms from 125 Chinese patients (incidence: 0.4%). Of the 125 patients, 58% of CAFs originated from the left anterior descending artery and 29% of CAFs originated from the right coronary artery. Most of CAFs (63%) drained to the pulmonary artery. Chiu et al. classified the CAFs into two types: type I in 99 patients with 124 solitary coronary to cardiac chamber or great vessel fistula; type II: 26 patients with 28 coronary artery-left ventricular multiple microfistulas. The incidence (0.09%) of type II CAFs in this report is significantly higher than those previous reported in Caucasian people. This incidence of type II CAFs was 0.015% in one of the largest series of 33600 consecutive angiograms[31].

Type I CAFs predominantly originated from the proximal segments (76%) and type II fistulas all originated from the mid (50%) or distal (50%) segments of the coronary artery. Single-, double-, and triple- CAFs were detected in 79%, 20%, and 1% of patients, respectively. The incidence of bilateral/multiple fistulas was also higher than those reported in Caucasian people. Coexistent coronary lesions were noted in 41% of patients. Fistula-related symptoms included stable angina in 55, myocardial infarction in 2, heart failure in 2, sudden death with ventricular fibrillation in 1, and syncope in 1. Twenty-four (20%) of patients had coexistent congenital anomalies. Unlike those reported CAFs in Caucasian people, the most common coexistent congenital anomaly was myocardial bridge. Most patients received medical treatment because of mild symptoms. Only 9 patients underwent coronary intervention or/and surgery for CAFs.

Treatment

The management strategy of patients with CAF depends on the size of the fistula, presence of symptoms, the anatomy of the fistula, the patient's age and whether the patient has other associated cardiovascular disorders. Small CAFs are usually asymptomatic, and may close spontaneously[32-34]. Patients with a small CAF have a good long-term prognosis and should be treated conservatively. Medical treatment with either beta blockers or calcium channel blockers is suggested[35,36]. Prophylaxis for bacterial endocarditis is recommended in all CAF patients and in patients after complete fistula occlusion for at least 1 year[37].

There appears to be good consensus that all symptomatic patients should undergo closure of medium or large CAFs. Said et al. suggested surgery or percutaneous transcatheter embolization to treat patients that have moderate or large CAFs with Qp/QS $\geqq 2$[18]. The first successful surgical closure was reported by Biörck and Carfoord in 1947[38], whereas the first therapeutic embolization was performed in 1974 by Zuberbuhler et al.[39]. Because transcatheter closure of CAF is associated with a much shorter recovery time and avoids a scar, it is considered the procedure of choice when fistula closure is indicated. Catheter closure can be performed with a variety of techniques, including detachable balloons, stainless steel coils, controlled-release coils, controlled-release patent ductus areteriosus coils, patent ductus areteriosus plugs, regular and covered stents, and various chemicals[14,40-46]. Surgical ligation should be reserved for patients who have a complex and distally located fistula, or have adjacent vessels at risk. In addition, surgical ligation may be preferred when correction of other congenital defects or coronary artery bypass grafting is required[47]. Mortality related to surgical closure or transcatheter closure of isolated CAFs is low ($<1\%$)[48]. Incomplete closure has been seen in ~10% of patients treated by catheter techniques or surgical ligation[44,49]. Until now, there is limited longitudinal information about the long-term prognosis of patients with CAFs following surgical or transcatheter treatment.

In conclusion, the incidence of acquired CAF is increasing following the incremental use of intravascular procedures and interventional techniques. The major clinical presentation of CAF depends on the size of the fistula. Therapy should be tailored according to patient's age, size and anatomy of the fistula, and other associated cardiovascular disorders. The incidence of bilateral/multiple fistulas or coronary artery-left ventricular fistulas seems to be higher in Oriental people than in Caucasian people. However, this issue needs more studies to declare.

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冠狀動脈瘺管的目前現況

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摘　要

冠狀動脈瘺管是一種少見的冠狀動脈異常。大部分爲先天性，但隨著介入性設備及技術使用的增加，後天性的原因逐漸增加。本症的盛行率在心導管或心臟超音波的研究中約爲0.1-0.8%。本症好發於右冠狀動脈及左前降枝動脈，且以近端侵犯爲主。大多數會引流至右心房、右心室或肺動脈，少數會引流至左心房或左心室。根據瘺管引流部位之不同，會有不同的生理學表現。臨床上約半數之冠狀動脈瘺管不會產生症狀。較大之瘺管可造成心衰竭、心絞痛、心肌梗塞、心律不整、或肺動脈高壓。其他像心內膜炎、冠狀動脈破裂、猝死也有文獻報告過。理學檢查常可聽到連續性雜音。鉈201心肌灌注掃描可用來幫助診斷心肌缺血。冠狀動脈血管攝影爲主要診斷工具。心臟超音波、核磁共振及多切面電腦斷層也常被用來幫助診斷。治療可分爲內科治療、經心導管關閉瘺管或外科瘺管結紮，但必須根據瘺管之大小、位置、病人年齡及臨床症狀來做不同的考量。冠狀動脈瘺管在東方人的表現也將在文章中予以探討。