Heart, Lung and Circulation (2018) xx, 1–12
1443-9506/04/$36.00
https://doi.org/10.1016/j.hlc.2017.07.014

REVIEW

Coronary Artery Fistulae: Anatomy, Diagnosis and Management Strategies

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Received 8 November 2016; received in revised form 3 April 2017; accepted 10 July 2017; online published-ahead-of-print xxx

Coronary artery fistula (CAF) is a relatively rare anatomic abnormality of the coronary arteries that afflicts 0.002% of the general population and represents 14% of all the anomalies of coronary arteries. Its clinical relevance focusses mainly on the mechanism of ''coronary steal phenomenon'', causing myocardial functional ischaemia even in the absence of stenosis, hence common symptoms are angina or effort dyspnoea. The suggested diagnostic approach is guided by the patient's symptoms and consists of a number of instrumental examinations like ECG, treadmill test, echocardiography, computed tomography scan, cardiac magnetic resonance and coronary angiography. If it is not an incidental finding, coronary angiography is required in view of the optimal therapeutic planning. Small sized fistulae are usually asymptomatic and have an excellent prognosis if managed medically with clinical follow-up with echocardiography every 2 to 5 years. In the case of symptomatic, large-sized or giant fistulae an invasive treatment, by transcatheter approach or surgical ligation, is usually a reasonable choice, and both strategies show equivalent results at long-term follow-up. Antibiotic prophylaxis for the prevention of bacterial endocarditis is recommended in all patients with coronary artery fistulae who undergo dental, gastrointestinal or urological procedures. A life-long follow-up is always essential to ensure that the patient is not undergoing progression of disease or further cardiac complications.

Keywords Coronary artery fistula (CAF) • Management • Transcatheter closure (TCC) • Diagnosis • Surgery • Coronary artery fistula ligation

Introduction

Coronary artery fistula (CAF) is an anomalous connection that bypasses the myocardial capillary bed between one or more coronary arteries and a cardiac chamber (coronary-cameral fistulae) or, otherwise, that links a coronary artery to a major blood vessel (arterio-venous fistulae). The first case described by Krause dates back to 1865 [1]. Frequently isolated (80%), CAFs could also be associated with other cardiac malformations (20%), mostly atrial or ventricular septal defects, tetralogy of Fallot, patent ductus arteriosus [2].

Epidemiology of CAF

Coronary artery fistula's exact incidence is as yet unknown because the undiagnosed rate still remains high, but it is estimated that, whereas the incidence of coronary anomalies is 0.2 to 1.2% in the general population, CAFs are present in 0.002% of the cases [3,4] and represent about 0.2–0.4% of all cardiac malformations [5] and 14% of all coronary anomalies [6]. Several studies report the presence of CAF in 0.3% of patients presenting with congenital heart disease, in 0.06% of children undergoing echocardiography and in 0.13–0.22% of adults undergoing coronary angiography [7].

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Please cite this article in press as: Buccheri D, et al. Coronary Artery Fistulae: Anatomy, Diagnosis and Management Strategies. Heart, Lung and Circulation (2018), https://doi.org/10.1016/j.hlc.2017.07.014

Ethnic group and gender do not affect the incidence of CAF, and multiple fistulae are present in 10.7–16% of the cases, while a single fistula is much more common, being present in up to 90% of all CAFs. To note, around 75% of incidentally-found CAFs are small and clinically silent [8].

Aetiology of CAF

Although, in the past, CAF was usually congenital, over the years the development and dissemination of interventional and surgical techniques has resulted in a change of its aetiology, with a higher prevalence of the acquired forms [9], which may include those secondary to infective endocarditis, aortic dissection, previous surgery, endomyocardial biopsy, coronary angioplasty and bypass surgery, valve replacement, cardiac transplantation, trauma, permanent pacemaker placement, closed-chest ablation of accessory pathways, neoplasms, iatrogenic management of Kawasaki disease [10].

From the embryological point of view, the congenital forms are due to the persistence of intramyocardial trabecular connections formed by endothelial cells and blood lacunae that are formed initially within the cardiac venous plexus and subsequently with the epicardial coronary arteries [11].

Anatomy and Classification of CAF

Frequently, the feeding artery of the CAF may drain from a coronary artery or one of its branches after a tortuous and dilated course that ends in one of the cardiac chambers or in a vessel. Proximal CAF are frequently large. On the other hand, they are usually smaller and more tortuous if distally located. Fistulae originating from the left coronary artery and draining into the left ventricle are particularly tortuous and represent a possible target for transcatheter closure (TCC) [8].

Multiple feeding arteries may converge on a single CAF, as well as multiple drainage sites which may arise by a single fistula. Multiple fistulae between the three major coronary arteries and the left ventricle have also been reported. In some cases, especially in adults, fistulae may originate from both coronary arteries, which drain into the pulmonary trunk. These CAFs can frequently cause angina and require closure [12]. Coronary artery fistulae arise more frequently from the right coronary artery (~50–60%) and often drain into the right heart (~80%).

Figure 1 shows the rates (%) of the origin of the fistulae from each coronary artery (Figure 1A) and drainage into each heart chamber/great vessel (Figure 1B).

Coronary artery fistulas are unilateral in most cases (more than 80%, both in children and adults), seldom bilateral or multilateral in very few cases, and generally isolated or, in 5–30% of the cases, associated with cardiac defects [13,14].

Pathophysiology, Clinical Presentation and Complications of Coronary Artery Fistulae

The pathophysiological changes caused by a fistula depend, at first, on the resistance that blood flow meets along its course, and the different pressures between the coronary

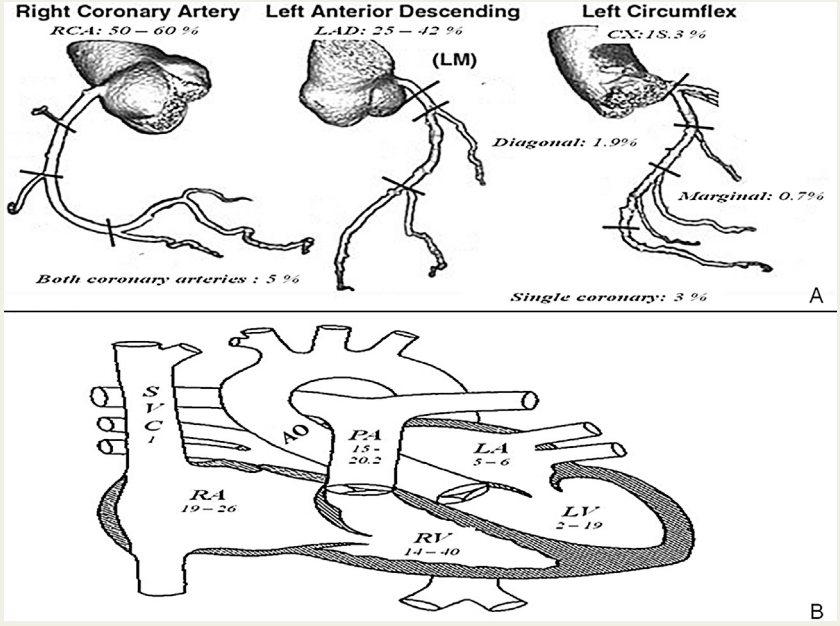

Figure 1 **A)** Rate (%) of origin of the fistulas from each coronary artery. **B)** Rate (%) of the fistula drainage sites into each heart chamber/great vessel. Source: Dodge-Khatami [4].
Abbreviations: RCA, right coronary artery; LAD, left anterior descending artery; LCX, left circumflex artery; LM, left main coronary artery; SVC, superior vena cava; RA, right atrium; RV, right ventricle; AO, aorta; PA, pulmonary artery; LA, left atrium; LV, left ventricle.

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