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Review

Coronary artery fistulas: Clinical and therapeutic considerations☆

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Abstract

Coronary artery fistulas vary widely in their morphological appearance and presentation. These fistulas are congenital or acquired coronary artery abnormalities in which blood is shunted into a cardiac chamber, great vessel, or other structure, bypassing the myocardial capillary network. The majority of these fistulas arise from the right coronary artery and the left anterior descending coronary artery; the circumflex coronary artery is rarely involved. Clinical manifestations vary considerably and the long-term outcome is not fully known. The patients with coronary fistulas may present with dyspnea, congestive heart failure, angina, endocarditis, arrhythmias, or myocardial infarction. A continuous murmur is often present and is highly suggestive of a coronary artery fistula. Differential diagnosis includes persistent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect, internal mammary artery to pulmonary artery fistula, and systemic arteriovenous fistula. Although noninvasive imaging may facilitate the diagnosis and identification of the origin and insertion of coronary artery fistulas, cardiac catheterization and coronary angiography is necessary for the precise delineation of coronary anatomy, for assessment of hemodynamics, and to show the presence of concomitant atherosclerosis and other structural anomalies. Treatment is advocated for symptomatic patients and for those asymptomatic patients who are at risk for future complications. Possible therapeutic options include surgical correction and transcatheter embolization. Historical perspectives, demographics, clinical presentations, diagnostic evaluation, and management of coronary artery fistula are elaborated.

Keywords: Coronary artery fistula; Coronary anomaly; Congenital heart disease; Coronary angiography; Continuous murmur

1. Introduction

Coronary artery fistulas are unusual congenital or acquired coronary artery abnormalities in which blood is shunted into a cardiac chamber, great vessel or other structure, bypassing the myocardial capillary network. Coronary artery fistulas are rare, difficult to detect coronary anomalies. They are usually congenital, but acquired forms do occur. Congenital fistulous connections between the coronary system and a cardiac chamber appear to represent persistence of embryonic intertrabecular spaces and sinusoids [1–3]. They are the most common congenital coronary anomalies affecting hemodynamic parameters [4–6]. Their clinical importance usually in the adulthood is due to an increased risk of complications, including heart failure, myocardial ischemia, infective endocarditis, arrhythmias, and rupture [7]. The acquired causes of coronary fistulas include coronary atherosclerosis, Takayasu arteritis, and trauma [8–10].

2. Historical perspective

The first reported case of a coronary artery fistula was in 1865 by Krause [11]. Haller and Little [12] described the diagnostic triad for coronary artery fistulas: an abnormal localization for a to-and-fro murmur thought to be due to a patent ductus arteriosus; a left to right shunt at the atrial or ventricular level; and a large, tortuous coronary artery at

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coronary angiography. Bjork and Crafoord reported the first successful fully corrected coronary artery fistulas in 1947 [13]. The first successful transcatheter closure of a coronary artery fistula was reported in 1983 [14].

3. Demographics

This infrequent abnormality can affect any age. Angiographic series reveal an incidence of 0.3–0.8% [4,7]. Coronary artery fistulas can occur from any of the three major coronary arteries, including the left main trunk [15]. The majority of these fistulas arise from the right coronary arteries or the left anterior descending; the circumflex coronary artery is rarely involved [11,16]. The right coronary artery, or its branches, is the site of the fistula in about 55% of cases; the left coronary artery in about 35%; and both coronary arteries in 5%. Over 90% of the fistulas drain into the venous circulation. Low-pressure structures are the most common sites of drainage of the coronary fistula. These include right-sided chambers, pulmonary artery, superior vena cava, and coronary sinus [17]. Fistulous communication to the left-sided chambers is less frequent [18]. Fistulous drainage occurs into the right ventricle in 41%, right atrium in 26%, pulmonary artery in 17%, left ventricle in 3%, and superior vena cava in 1% [1]. A case has been reported where coronary artery fistula drained into the pericardium causing hematoma [19]. Coronary artery dilatation is usual, and the degree of dilatation does not always depend on the shunt size [7]. When the fistulous communication arises in the distal part of a coronary artery, the artery diameter may remain small. Most fistulas are single communications, but multiple fistulas have been reported [20]. The myocardial blood flow is usually not compromised and shunt through the fistula most often is of small magnitude [21]. The shunt ratio is usually small when the drainage site of the fistula is to the pulmonary artery. A left-to-right shunt exists in more than 90% of cases.

4. Clinical presentation

The clinical presentation of coronary artery fistulas is mainly dependent on the severity of the left-to-right shunt [22]. The majority of adult patients are usually asymptomatic. Unlike adults, a smaller percentage of pediatric patients tend to be asymptomatic [23]. This may be the result of the larger fistulas being likely to cause symptoms and bring attention to the fistulas in pediatric age group. Nevertheless, pediatric patients are usually referred due to electrocardiographic or chest X-ray abnormalities or for evaluation of a loud, superficial continuous cardiac murmur. The clinical presentations include fatigue, dyspnea, orthopnea, angina, endocarditis, arrhythmias, stroke, myocardial ischemia or myocardial infarction [22,23]. Myocardial ischemia/infarction can occur from decreased coronary blood flow distal to the fistula and has been documented in patients with coronary fistulas with no evidence of coronary atherosclerosis. Rarely, pericardial effusion and sudden death can be a presenting feature [3,24]. In coronary artery fistulas the continuous murmur tends to be crescendo decrescendo in both systole and diastole, but louder in diastole. In contrast, most of the other continuous murmurs reach their peak intensity at the time of the second sound. The place where the continuous murmur is loudest depends on where fistula enters in the heart. With entry into the right atrium the murmur is loudest lower along the sternal border, with entry into pulmonary artery it is loudest near the second intercostal space to the left of sternum, and with entry into the left ventricle the murmur is loudest near the apex. Potential complications if a large left-to-right shunt exists are pulmonary hypertension and congestive heart failure; others include rupture or thrombosis of the fistula or associated arterial aneurysm or coronary steal phenomena.

5. Diagnostic evaluation

Many fistulas are small and found incidentally during coronary arteriography. Pediatric patients, even asymptomatic, may present with electrocardiographic or chest X-ray abnormalities [22]. Differential diagnosis includes persistent ductus arteriosus, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect, internal mammary artery to pulmonary artery fistula, and systemic arteriovenous fistula [25]. Various cardiac imaging modalities are utilized for diagnosis and for planning before surgical or percutaneous interventions if closure of the coronary fistula is indicated.

A significantly enlarged coronary artery can usually be detected by two-dimensional echocardiography. Transthoracic echocardiographic imaging is more successful in children in whom optimal acoustic windows are obtained. Continuous turbulent systolic and diastolic flow pattern characterizes the shunt entry site [21,26]. Use of contrast microbubbles to enhance the color Doppler signals assists to define location and extent of coronary artery fistulas [27]. When a coronary artery fistula is present, a dilated feeder vessel with an abnormal flow pattern can be readily identified. However, several limitations have been described with transthoracic echocardiography in patients with coronary artery fistulas [28,29]. Color Doppler may not detect flow in the distal site of coronary artery fistulas. Multiplane transesophageal echocardiography can more accurately define and provide a high quality panoramic view of the origin, course, and drainage site of coronary artery fistulas [7,21,26,30]. Even though transesophageal echocardiography can be a complementary imaging modality in angiographically documented coronary anomalies, it also has limitations; identification of coronary artery fistulas is