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Review Article

Coronary artery fistula: a review

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ABSTRACT

Coronary arterial fistulas are abnormal connections between the coronary arteries and the chambers of the heart or major thoracic vessels. Although first described in 1841, the true incidence is difficult to evaluate because approximately half of the cases may be asymptomatic and clinically undetectable. This review will discuss the history and prevalence of coronary artery fistulas and their morphology, histology, presentation, diagnosis, treatment options, and complications.

1. History and prevalence

As the aorta exits the left ventricle, two coronary arteries originate from its root to supply the muscle and tissues of the heart. The left coronary artery originates from the left aortic sinus, whereas the right coronary artery originates from the right aortic sinus. As the right coronary artery descends it branches to give a sinuatrial nodal branch, right marginal branch, and a posterior interventricular branch. On the other hand, the left coronary artery descends and gives an anterior interventricular branch (Fig. 1) and a circumflex branch.

A coronary artery fistula, however, is an abnormal connection that directly links one or more coronary arteries to a heart chamber or to major thoracic vessels without an interposed capillary bed [1]. Fistulas that arise from a coronary artery and then terminate into a chamber of the heart are known as coronary-cameral fistulas, while those terminating into a vein are coronary arteriovenous fistulas [2]. Coronary artery fistulas occur equally in both sexes and have a 0.002% incidence among the general population [3,4], accounting for 0.1% of coronary anomalies with 2–17% occurring bilaterally [1,5].

While relatively uncommon in the population, coronary abnormalities attributed to coronary artery fistulas have been recognized for hundreds of years. The first description of a coronary artery fistula occurred in 1841 by the Austrian anatomist Josef Hrytl [4,6]. Despite this initial discovery, many accredit German anatomist W. Kraus's 1865 description of an aberrant connection between a coronary artery and a heart chamber as the first reported coronary artery fistula [4,6]. Much of the statistical data regarding the incidence, etiology, presentation, and treatment of coronary artery fistulas is derived from cohort data, both retrospective and prospective, as well as numerous case studies. These resources categorize them as either congenital or acquired anomalies. Congenital coronary artery fistulas are more common but only account for 0.4% of congenital cardiac abnormalities [4,7].

Acquired coronary artery fistulas develop from injurious events, iatrogenic or traumatic in nature, with a predominance of the former. Several cases report the appearance of acquired fistulas both postoperatively and after certain medical conditions, including the following: heart transplants, mitral valve replacements, percutaneous coronary interventions, septal myectomies, closed-chest ablations, coronary artery bypass grafts, permanent pacemaker placements, acute myocardial infarctions, and transbronchial lung biopsies [4,6,8]. In such cases, the sudden postoperative appearance of a continuous murmur is indicative of iatrogenic fistula formation [4], whereas acquired coronary artery fistulas due to traumatic injury can occur in both thorax penetrating and non-penetrating accidents [4,9]. In these, the most common point of origin is the right coronary artery, and the most common termination is within the right chambers of the heart [4,10].

Similar to humans, fistulas in mammals, especially cattle, usually occur between the coronary arteries and the ventricles of the heart

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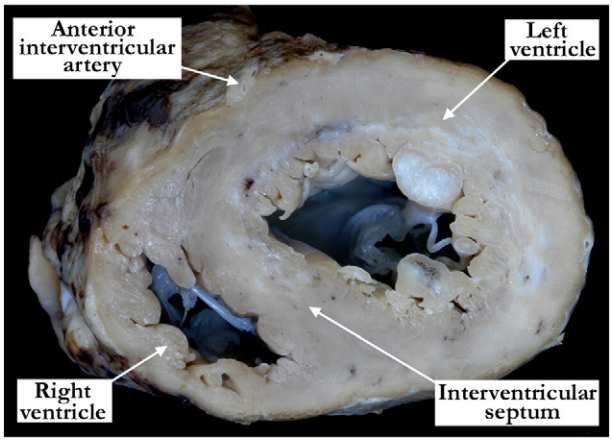

Fig. 1. This figure is a lower cross-sectional view of a normal heart exhibiting key anatomical landmarks: left and right ventricles, interventricular septum, and anterior interventricular artery (a branch of the left coronary artery). Source: Diane Spicer, Department of Pediatrics–Cardiology, University of Florida, Gainesville, Florida.

[11]. In Michaelsson and Ho's study, the right ventricle was the most common termination of these fistulas. In addition to cattle, coronary artery fistulas also occur in canines. This type of fistula is uncommon in dogs, just as they are in humans, and present with analogous symptoms, complications, and treatment options [12].

2. Morphology

While the exact percentage of morphological origins and terminations of coronary artery fistulas differ, the consensus is that fistulas are typically found on the right side of the heart. Studies show that the point of origin in 52–60% of coronary artery fistulas is the right coronary artery, 30% at the left anterior interventricular (left anterior descending) artery, and 18% at the left circumflex artery [2,13,14]. Regardless of point of origin, nearly 90% of fistulas drain to the right chambers of the heart [14]. In 2006, Qureshi et al. stated that, in the right heart, drainage occurs most frequently to the right ventricle, in about 40%, followed by the right atrium (Fig. 2), coronary sinus, and pulmonary trunk [14] (Fig. 3). As these studies indicate, any of the three major coronary arteries can be the feeding artery for the coronary fistula, and depending on the artery and point of origin, the tortuosity and dilation of the fistula will vary. Coronary artery fistulas that branch from a main coronary artery at a proximal location are more dilated when compared to other distally located coronary artery

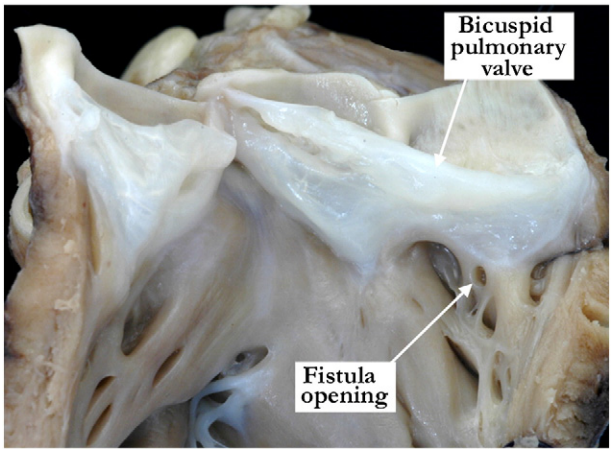

Fig. 2. This figure is taken from a dissected right atrium showing the termination of a coronary artery fistula into the wall of the atrium. Source: Diane Spicer, Department of Pediatrics–Cardiology, University of Florida, Gainesville, Florida.

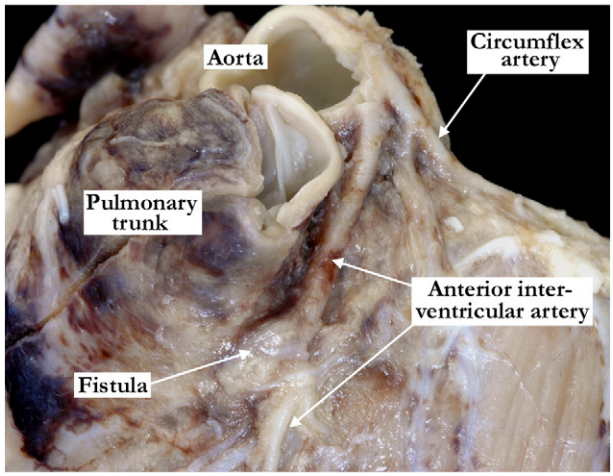

Fig. 3. This figure is an anterior view of the heart showing the anterior interventricular artery as it gives a fistulous communication terminating in the pulmonary trunk. Source: Diane Spicer, Department of Pediatrics–Cardiology, University of Florida, Gainesville, Florida.

fistulas [15]. For example, fistulas that originate proximally and drain into the right atrium are much more dilated but less tortuous. The most anatomically complex coronary artery fistulas are those that originate at the right coronary artery and drain into the coronary sinus. These tend to be extremely large and tortuous [4].

As with the point of origin, the drainage site introduces a certain degree of anatomic variation [16]. In 1974, a study by Ogden et al. was the first to categorize coronary artery fistulas based on their drainage site in the atriums, classifying them into 3 types.

Type 1 coronary artery fistulas can either terminate into the right or left atrium, and these fistulas are physiologically similar to each other but different from those terminating in the high-pressure ventricles. The thick walls of the ventricles allow for the closure of the fistula during ventricular systole whereas the thinner walls of the atriums have no effect on the fistula opening. Type 1 classically has short, dilated arterial branches that extend from the right coronary artery and terminate in either the right appendage (Fig. 4) or the right atrial vestibule (Fig. 2). Right-atrium-terminating Type 1 coronary artery fistulas usually only involve the right coronary artery (Fig. 5) whereas those terminating in the left atrium mainly involve the left coronary branches [16] (Fig. 5).

Type 2 coronary artery fistulas are the most frequently encountered and are more variable than Type 1, since both the left (Fig. 6) and the right (Fig. 6) coronary arteries can feed the fistula. From its point of origin, the fistula usually travels posteriorly between the

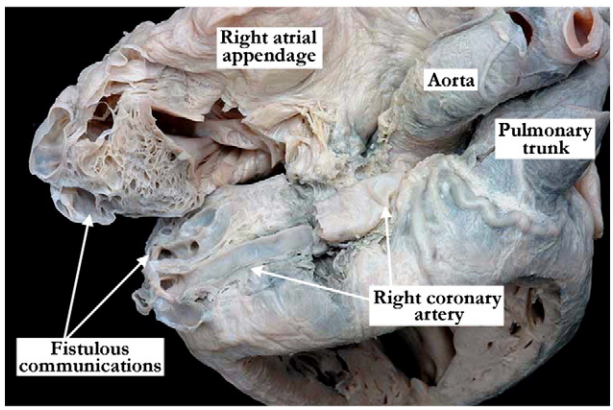

Fig. 4. This figure is taken from a dissected atrial heart showing a fistulous communication from the right coronary artery to the right atrial appendage. Source: Diane Spicer, Department of Pediatrics–Cardiology, University of Florida, Gainesville, Florida.