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# Giant Sinus Node Artery Fistuling to Superior Vena Cava Presenting with "Steal Phenomenon"- An Unusual Case

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## Abstract

Congenital coronary fistulas are rare anomalies first described by Krause in 1865. They can be either symptomatic or asymptomatic. Management strategy for a coronary fistula differs depending on the blood flow it has and symptoms it gives rise to. Nevertheless, it should be taken as a serious health problem that could result in such unpleasant complications as congestive heart failure through left to right shunt, myocardial ischemia through 'steal phenomenon', endocarditis, and aneurysmal rupture. We present surgical management of a young female referred to our clinic with exercise-induced substernal chest pain and numbness in the left upper extremity that were found to be caused by a giant sinus node artery fistuled to superior vena cava.

**Keywords:** Coronary artery fistulae; Vena cava superior; Coronary artery disease; Sinus node artery

## Introduction

Congenital coronary fistulas are rare anomalies with an incidence of 0.1% to 0.2% [1]. First described by Krause in 1865, they occur because of anomalies in the embryology of the coronary circulation [1]. The first surgical procedure was performed by Bjork and Crawford in 1974 [2]. The right coronary artery is the most common site of origin and the right ventricle and atrium are the most common areas of opening [2]. We present surgical treatment of a rare and giant coronary fistula that opens into the superior vena cava through the sinus node artery.

## **Case Presentation**

The patient (female, 31 years of age) referred to our clinic with exercise-induced substernal chest pain and numbness in the left upper extremity. In the clinics she visited in the last year, various treatments were applied to attribute the patient's complaints to extracardiac causes. Patient without positive findings on physical examination had cardiomegaly findings on chest radiograph and electrocardiogram [3,4]. After cardiac catheterization, an extraordinary tortuous arterial structure extending from the right coronary artery to the superior vena cava was observed (Figures 1 and 2). At the perioperative inspection, the aneurysmatic right coronary artery was found to give sinus node artery branch approximately 2 cm after separation from the aorta, and the diameter decreased to 4 mm from there. The diameter of the sinus node artery was 1.5 cm. After running about 5 cm, it opened medially into the superior vena cava by creating a murmur of 4/6 magnitude. A  $2 \times 2$  cm venous aneurysm formation was observed at the opening (Figure 3).

Surgery was performed by standard surgical procedures with median sternotomy and bicaval cannulation. Considering the increased likelihood of rupture, the aneurysmatic proximal 2 cm segment of the right coronary artery was removed (Figures 3 and 4). Saphenous vein graft was interposed between the aorta and distal free end of the right coronary artery (Figure 5). The sinus node artery was also aneurysmatic and tortuous [5]. It was excised completely and the point at which it opened into the superior vena cava was sutured. The operation was performed under cardiopulmonary bypass in case of the possibility of rhythm disturbance. After giving the sinus node artery, the right coronary artery continued about 10 cm in its normal diameter. The postoperative course was smooth, and the patient was discharged on the 5<sup>th</sup> day of hospitalization [6].



Figure 1: Appearance of fistula tract in coronary angiography.

#### Discussion

In the presence of coronary artery disease, angina is a common finding in elderly patients. However, in young patients and especially in females, angina should suggest congenital arteriovenous fistulas. Although coronary fistulas are mostly congenital, they may occur after blunting external thoracic trauma, myocardial infarction, angioplasty or cardiac surgery [7]. Other congenital anomalies accompany coronary fistulas by a frequency of 40%. They become symptomatic after thirty years of age. The angina is caused by coronary 'steal'. Fistulas opening

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Figure 2: CT angiography showing the aneurysmatic right coronary artery and sinus node artery. The right coronary artery is seen to run in normal diameter after giving the sinus node artery branch. (RV=Right ventricle, RA=Right atrium, LV=Left Ventricle, LA=Left Atrium, SNA=Sinus Nod Artery, VCI=Vena Cava Inferior).



Figure 3: Perioperative view of aneurysmatic sinus node artery running medial to the right atrium and fistulizing to superior vena cava, and distal right coronary artery in normal diameter.

into right heart chambers create shunts from left to right. This may cause congestive heart failure in proportion to the size of the shunt. Such symptoms as angina pectoris, fatigue, shortness of breath, palpitation, and findings suggestive of rupture of an aneurysm, emboli, endocarditis, congestive heart failure can be detected. The degree of symptoms is proportional to the amount of physiological coronary stealing phenomenon developed by the fistula tract [8]. The frequency of the continuous murmur, the most frequent physical examination



finding, ranges from 20% to 80% [9]. Almost half of the patients have nonspecific electrocardiographic changes. Cardiomegaly can be observed on the telecardiography due to increased cardiac output [1]. For a definitive diagnosis, selective coronary angiography is required. Thus, the origin and termination of the fistula, the path it follows anatomically, and the affected structures can be seen.

It has been reported that coronary fistulas originate mainly from the right coronary artery, less frequently from the left coronary

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**Figure 5**: Saphenous vein graft interposed between the aorta and distal free end of the right coronary artery after resection of the proximal aneurysmatic segment.

artery, or both. They originate from the right coronary artery with a frequency of 19,7% in the Albeyoglu series, 50% in Levin, 65% in Lowe, and 51% in the Wilde and Watt series [3-5]. Drainage occurs into low-pressure heart chambers. Frequency in decreasing order is as right ventricle (39%), right atrium (33%), pulmonary artery (20%), left atrium, coronary sinus, vena cava superior and bronchial arteries [1,4]. Drainage to the left heart chambers is relatively rare (2%) [1]. Pathophysiological, myocardial ischemia or infarction occurs when the blood to be directed to the left ventricle is directed to the low-resistance heart chambers through the fistula, creating a "steal phenomenon". In fistulas that open to the left ventricle, blood flow is frequently diastolic. Ischemia occurs when the pulse pressure providing coronary flow is lowered by steal phenomenon [3]. One of the potential complications is premature atherosclerosis resulting in intimal damage caused by high-volume blood flow [7]. Our patient had angina symptoms and tachycardia attacks on the front.

#### Conclusion

Treatment of asymptomatic fistulas is controversial. However, most surgeons agree that they should be closed in the presence of a significant shunt or aneurysmal dilatation [1]. Symptomatic people need to be treated. The cardiopulmonary bypass may be required or not [10]. Fistulas that are intramural, short, close to Sinus Valsalva and characterized by an aneurysm are generally closed using cardiopulmonary bypass while extramural and anatomically accessible ones are closed by simple ligation and resection. Closure with the use of percutaneous transcatheter closure devices is becoming increasingly widespread, especially in pediatric patients [11]. Some authors do not consider surgery in asymptomatic patients, while surgery is appropriate for moderate to large-flow fistulas. Small fistulas are likely to spontaneously close as primarily or secondarily. Nevertheless, most authors consider surgical removal of fistulas to set aside complications and sudden death risk, regardless of whether they are symptomatic or not. The reported operative mortality and morbidity are very low. It should be known that fistula complications could increase surgical morbidity and mortality.

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