

# CASE REPORT

## Left main coronary anomalies of the right ventricular sinus in patients with aortic failure due to the web sub-aortic: A case report

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### Abstract

Coronary artery anomalies are reported to have a higher incidence in young victims of sudden cardiac arrest (4-15%), as compared to adults (1%). Among coronary anomalies, anomalous coronary artery arising from the opposite sinus of Valsalva (ACAOS) remains a major clinical problem which poses daunting challenges. The current paper reports on the first case with main left coronary anomalies with aortic failure due to the web sub-aortic. A 29-year-old woman with orthopnea referred to a doctor. Echocardiography and electrocardiogram (ECG) confirmed sub aortic web with severe aortic insufficiency. Subsequently, the patient was subjected to aortic valve replacement surgery and the subvalvular resection was performed. During the surgery, the aorta was initially opened and the left coronary hole was not found in the coronary sinus. Therefore, only a cardioplegia from the right coronary (artery) hole was administered to the patient. The cusps of the valve and subvalvular web were removed. During the release, the left main artery which apparently originated from the right coronary artery(RCA) was found to be damaged. Therefore, the damaged part of the left main coronary artery was repaired, the valve was placed, and the aorta was repaired. Following deaeration, cross-clamp was removed, and the right heart started to work; nonetheless, the left heart was not engaged in any activity. Therefore, the cross-clamp was attached again to the aorta, and a vein graft on the left anterior descending (LAD) artery was placed on the aorta. After the removal of the cross-clamp, the heart recovered its normal function, and the patient was removed from the pump with low inotropic. Thereafter, she was transferred to the Intensive Care Unit (ICU) of open-heart surgery. The present report aimed to express the incidence of multiple complications, such as recurrence of illness, the optimal time for surgery, the best way of surgery, getting cardiologists' attention (colleagues) during angiography, and knowing the characteristics of these abnormalities.

**Key words:** Aortic stenosis, Aortic valve insufficiency, Coronary sinus, Coronary vessel anomalies, Subvalvular

### Introduction

Congenital coronary artery anomaly is recognized as a major cause of sudden cardiac death (Hirachan). Coronary artery anomalies are reported to have a higher incidence in young victims of sudden cardiac arrest(4-15%), as compared to adults (1%). Among the recognized anatomic variants, anomalous coronary artery

arising from the opposite sinus of Valsalva (ACAOS) remains a major clinical problem which poses daunting challenges (2). According to the literature review, no study has so far been conducted in Iran on aortic failure due to the sub-aortic web with the main left coronary anomalies of the right ventricular sinus. Similarly, in the survey of studies performed outside Iran, no study was found on the existence of these two

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abnormalities. The current article reported on the first case with the main left coronary anomalies with aortic failure following a sub-aortic web. The present study aimed to report the incidence of multiple complications, such as the recurrence of the illness, optimal time of surgery, the best way of surgery, getting cardiologists' attention during angiography, and knowing about the characteristics of these abnormalities. Cardiologist's unawareness of these anomalies during angiography will cause irreparable lesions to patients.

## Cases

Here we report the case of a 29-year-old woman who referred to a doctor with a complaint of orthopnea. Her height was 160 cm and her weight was 36 kg. She had no history of previous illness, surgery, and drug use; nonetheless, she had a history of drug abuse. In the examination, the systolic murmur was heard in the aortic center, and she did not undergo angiography due to her age.

Echocardiography and electrocardiogram (ECG) revealed a subaortic web with severe aortic insufficiency. Subsequently, she was subjected to aortic valve replacement surgery, and the subvalvular resection was performed.

In addition, echocardiography established a definitive diagnosis of severe aortic regurgitation (AR). Therefore, the patient underwent transesophageal echocardiography (TEE). According to the echocardiography of the patient, TEE was not sufficient; therefore, transthoracic echocardiogram (TTE) was performed. The TEE

findings are demonstrated in Table 1. TTE examinations revealed aortic valve failure. In the echocardiography of severe aortic insufficiency and subaortic web and left superior vena cava (LSVC), aortic failure was induced by the subaortic web. After the definitive diagnosis, the patient was subjected to the aortic valve replacement and the resection of the subvalvular web.

During the surgery, the aorta was initially opened and the left coronary hole was not found in the coronary sinus. Therefore, only a cardioplegia from the right coronary (artery) hole was administered to the patient. The cusps of the valve and subvalvular web were removed (Web pathology under the valve in Table 1). The valve sutures were attached to the annulus valve, and aortic root enlargement was performed since the bottom of the metal valve No. 19 was not replaceable. Therefore, the aortic wall was cut to the non-coronary sinus of the annulus. During the release, we found that the left main artery which apparently originated from the right coronary artery (RCA) was damaged. Cardioplegic injection confirmed this state. Therefore, the damaged part of the left main coronary artery was repaired, the valve was placed, and the aorta was repaired. Following deaeration, cross-clamp was removed, and the right heart started working; however, the left heart was not engaged in any activity. Therefore, the cross-clamp was attached again to the aorta and a vein graft on LAD was placed on the aorta. After the removal of cross-clamp, the heart recovered its normal function, the patient was removed from the pump with low inotropic, and she was transferred to the intensive care unit

**Table 1: Web pathology under the valve**

1- Aortic valve: myxoid changes with hyalinization
2- Subaortic web/resection: fibrohyalin tissue with myxoid changes
Echo Lab,+ TTE Report
Clinical dx: SUB aortic web, rhythm: Normal sinus rhythm with a short run of Paroxysmal Supraventricular Tachycardia, HR: 85-90/Min, Bsa=1.36m <sup>2</sup>
*LEFT Ventricle: the severely dilated size and NL function with no LVH, EF=65%, NO RWMA ( RWMA: regional wall motion abnormality), Visible paraseptal band.
*RV: is NL size & function.
*Left atrium: is mildly dilated size
*CS: Dilated coronary sinus (origin:12mm) due to persistent left superior vena cava without atrial septal defect contrast study
*RA: Highly mobile chiari network
*Mitral valve: NL with trace mitral regurgitation
*Aortic valve:
Is 3ricuspid % thickened and mildly calcified NCC % RCC with moderately severe aortic insufficiency, no valvular aortic stenosis
There is thick semi-circular WEB just below the aortic valve with significant left ventricular outflow tract turbulence and mild stenosis, web position 12mm below the aortic valve
*Tricuspid valve: NL Mild TR
*Pulmonic valve: NL/ Mild PI

(ICU) of open-heart surgery. Postoperative echocardiography was normal, and the patient underwent drug therapy. It is worthy to note that case report articles are not obliged to show "Ethical Approval Code. Nevertheless, written informed consent was obtained from the patient.

## Discussion

Several studies have so far been carried out on the anomalous origin of the coronary artery. Nonetheless, the current study is the first reported case of left main coronary anomalies of the right ventricular sinus in patients with aortic failure due to the web sub-aortic. Any genetic defect that contributes to poor formation of the primitive coronary artery may be associated with a defect in the muscular wall of the aorta as concentric stenosis revealed as a sand hour defect (3).

Subvalvular aortic stenosis (SAS), also known as subaortic stenosis, includes various anatomic lesions which are likely to arise either alone or in combination. The following discrete entities have been described in the literature: 1. The thin, crescent-shaped membrane just below the aortic valve: discrete SAS. This represents 75%-85% of SAS cases. 2. Thick fibromuscular ridge. 3. Tunnel or tubular: long, narrow, fibromuscular channel along the Left Ventricular Outflow Tract (LVOT). It is noteworthy that the majority of adult patients with SAS are asymptomatic (4). In this report, a thick semicircular web was detected just below the aortic (AO) valve with significant left ventricular outflow tract (VOT) turbulence and mild stenosis, web position 12mm below the aortic valve.

Left coronary arteries which originate from the right vascular sinus are very rare and of four types. Type one is an antler (percolator): the left coronary artery passes through the anterior polyarteritis artery. Type two is inter-arteries: The left coronary artery is derived from the aorta and pulmonary arteries. Type three is Septal Brigade: The left coronary artery passes through the crystalline peristalsis. Type four is Retro motor: The left coronary artery moves from the posterior aorta (5). In the current report, the patient was diagnosed with a major left coronary of type four retrosynthetic. In other words, the left coronary artery passed through the aorta.

The majority of adult patients with SAS are known to be asymptomatic. Some patients experience no symptoms until they get engaged in such activities as exercise or pregnancy that cause physical stress. Symptoms may include dyspnea,

presyncope, or fatigue. As the obstruction worsens, some patients may develop chest pain or syncope during exertion. In addition, palpitations may occur in some other patients. Moreover, it can lead to congestive heart failure in rare cases. The SAS is initially diagnosed with the auscultation of a systolic ejection murmur which is louder at the left mid-sternal border radiating to the upper sternal border (4). In the present report, the patient with orthopnea was examined, and the murmur was heard in the aortic center. She was admitted with chest pain and dyspnea due to her age (29-year-old). In their study, Sabzi et al. described a rare case of this anomaly that was simultaneously associated with supravulvular aortic stenosis and coronary-pulmonary fistula without the presence of conventional collateral circulation in a 16-year-old boy (3).

Echocardiography is recognized as the diagnostic test of choice for SAS. In a study conducted by Oliver et al., Doppler examination was of great help to the precise identification of the cardiac abnormality leading to LVOT obstruction. This results in the correct assessment of the different anatomic patterns (4). Imaging plays a key role in the evaluation of discrete subvalvular aortic stenosis to distinguish it from other or coexisting causes of LVOT obstruction and direct the timing and type of intervention. Transthoracic and transesophageal echocardiography is well suited to imaging of these patients. Nonetheless, recent advances in 3D echocardiography, computed tomography, and magnetic resonance imaging perform prominent roles in specific situations (6). In this case report, TTE examinations indicated aortic valve failure. Echocardiography confirmed the diagnosis of a subaortic web with severe aortic insufficiency.

The membrane, as is well known, can affect the aortic valve and cause secondary aortic regurgitation which can be addressed only by surgical resection of the subaortic membrane and repair of the aortic valve (7). There exists major controversy over the management of the anomalous origin of the coronary artery. Surgery is a definitive treatment which is even recommended for asymptomatic patients. Only a few patients are treated without follow-up and medical treatments (8). Most of the reports on the risk of reoperation in patients undergoing relief of subaortic obstruction have focused on anatomic subtypes. Ezon et al. reported that  $\geq 2$  studies recommended surgery at diagnosis, regardless of the severity of the obstruction. Brauner et al. suggested that early surgery prevents AR. Nevertheless, the prevention of AR alone is not a

criterion for surgery (4).

In this report, the patient was also candidate for aortic valve replacement surgery. Moreover, the left coronary anomaly was detected during the surgery, and coronary artery bypass graft (CABG) surgery was also performed. According to the clinical and Para clinical evidence, the patient is in good physical condition following three years follow-up. In the case report conducted by sabzi et al., the patient underwent coronary artery bypass grafting with the repair of supravalvular aortic stenosis. The postoperative course was uneventful. The 6-month follow-up revealed a normal diameter of the ascending aorta with symptomatic relief of preoperative chest complaint.

The left main coronary artery of the right ventricular sinus can be diagnosed with angiography. However, angiography was not performed in our patient due to her age (29-year-old), and the coronary anomaly of the right ventricular sinus was detected during the surgery. It should be noted that angiography is preferred in patients with severe aortic insufficiency even at a young age.

## Conclusions

Based on the obtained results, it can be concluded that coronary involvement must be taken into account regardless of the severity of the disease. In addition, all patients may require detailed coronary artery imaging before surgical intervention.

## Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this article.

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