CASE REPORT

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# Rare case of single coronary artery in a patient with liver cirrhosis

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**Abstract**

We report here the case of a 58-year-old male presented with atypical chest pain, dyspnea and fatigue, with a medical history of liver cirrhosis and undergoing treatment with beta-blocker. The clinical exam was normal. The 12-lead electrocardiogram (ECG) showed normal heart rate, without repolarization changes. Transthoracic echocardiography revealed no wall motion abnormalities of the left ventricle, moderate tricuspid regurgitation with mild pulmonary hypertension and left ventricular hypertrophy. The biochemical markers for myocardial infarction were negative. He underwent coronary angiography that revealed a single coronary artery originating from the right coronary sinus of Valsalva.

***Keywords***: single coronary artery, cirrhosis, chest pain, congenital.

## Introduction

Single coronary artery (SCA) is one of the rarest congenital anomaly of the coronary arteries. It is described as a single coronary ostium that provides perfusion for the entire myocardium arising from the aortic trunk [1–3]. The prevalence of SCA in current literature is approximately 0.024% [1] to 0.066% [2] in patients referred for coronary angiography.

In the past 100 years, different classifications were used for SCA based on angiographic and necropsy findings [4, 5]. Based on the anatomical distribution of branches and according to the site of origin, Lipton *et al.* [1] suggested an angiographic classification in the right-type or left-type depending from the origin of the SCA – either from the right or left coronary sinus of Valsalva. These are further classified in three subtypes, depending on the anatomical course of the coronary arteries. SCA arising from the right coronary sinus of Valsalva is extremely rare, especially in the absence of the left coronary artery, and therefore few cases have been reported.

We report a rare case of a 58-year-old male, with a medical history of liver cirrhosis, undergoing treatment with beta-blocker that presented with atypical chest pain, dyspnea and fatigue. The electrocardiogram (ECG) was normal and the biochemical markers for myocardial infarction were negative. We decided to proceed to a coronary angiography that revealed a rare coronary abnormality – a SCA that arises from the right coronary sinus of Valsalva.

## Case presentation

A 58-year-old man without any cardiovascular disease in the past, with medical history of liver cirrhosis for which he was being treated with a beta-blocker, was admitted to the Department of Cardiology of Emergency County Hospital, Craiova, Romania, presenting atypical chest pain and fatigue.

The study was performed according to the tenets of the Declaration of Helsinki and was approved by the University Ethics Committee. The patient was fully informed about the possible consequences of the present study, and he filled in an informed consent form to participate.

Clinical examination revealed average general state, blood pressure was 140/80 mmHg, heart rate 60 beats per minute (bpm), without heart murmurs, peripheral pulse present, no jugular vein distension, vesicular murmur present bilaterally without crackles superimposed.

ECG on admission revealed normal heart rhythm, with a heart rate of 60 bpm, QRS axis of 30 degrees, without alterations of the repolarization phase.

Laboratory data were normal. The analyses of arterial blood gas and the myocardial enzymes levels showed normal findings. The chest X-ray was without any pathological findings. The transthoracic echocardiography was performed in the Department of Cardiology, using Toshiba Ultrasound Aplio CV series with a cardiac transducer PST-50BT with frequency 3–6.2 MHz. Transthoracic echocardiography showed preserved left ventricular systolic function, diastolic dysfunction with delayed relaxation filling pattern, moderate tricuspid regurgitation, mild pulmonary hypertension, left ventricular hypertrophy without segmental motion disorders, the ejection fraction of left ventricle of 60%, the right heart without dilatation.

The patient received treatment with fractionated heparin (Enoxaparin), beta-blocker and nitrate. During hospitalization, he was hemodynamically stable, without repeating chest pain or electrocardiographic changes.

The patient underwent coronary angiography. The procedure was performed in the CathLab of Department of Cardiology, using a Siemens Angiograph. The right radial approach was used, using a 5F sheath. The cannulation of the left coronary artery was attempted with Judkins left 3.5, 4.0, 4.5, 5.0 diagnostic catheters and with Amplatz left catheter without success. Afterwards, the cannulation of the right coronary artery was attempted with Judkins right catheter. The result was unexpected; there was only one SCA with the origin from the right coronary sinus (Figure 1), with a common trunk, that divided into the right coronary artery and left coronary artery with a long left main without any lesions, the left anterior descending artery and the circumflex artery also without significant stenosis (Figure 2, A and B). The right coronary artery was normal. We managed the cannulation of the SCA using also the Judkins left 4.0 catheter. We performed multiple injections in the left coronary sinus and in the ascending aorta with manual injection and automatic injection to show the absence of the left coronary (Figures 3 and 4).

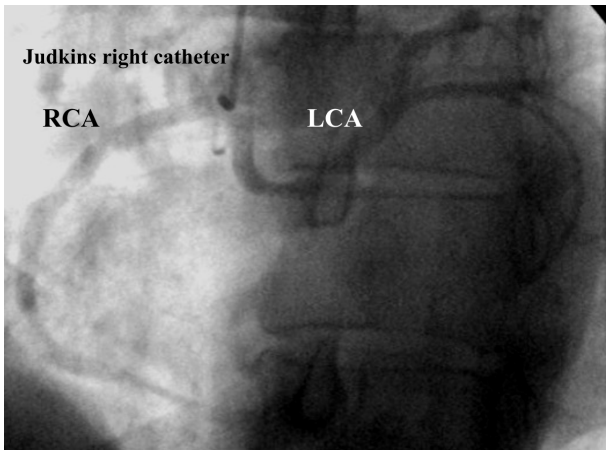


**Figure 1** – ***Single coronary artery from the right sinus of Valsalva. LCA: Left coronary artery; RCA: Right coronary artery.***

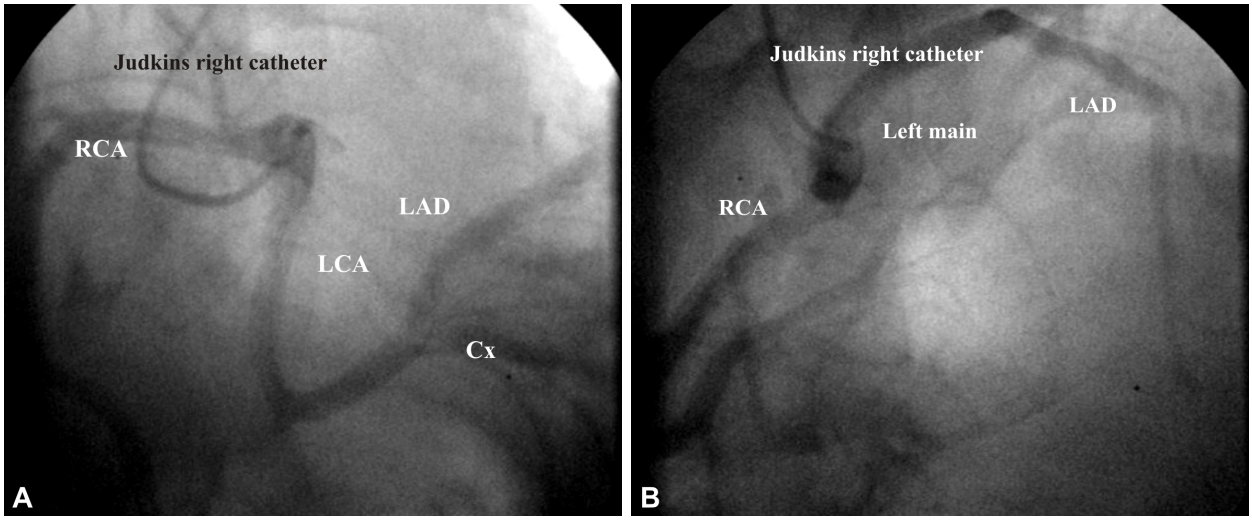


**Figure 2** – ***(A and B) Left main (very long) with left anterior descending (LAD) artery and circumflex (Cx) artery. LCA: Left coronary artery; RCA: Right coronary artery.***

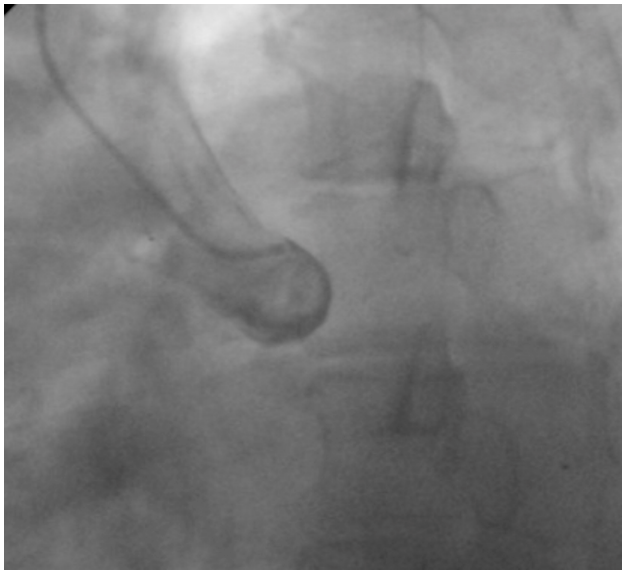

**Figure 3** – ***Left coronary sinus of Valsalva: the normal place of the origin of left coronary artery.***

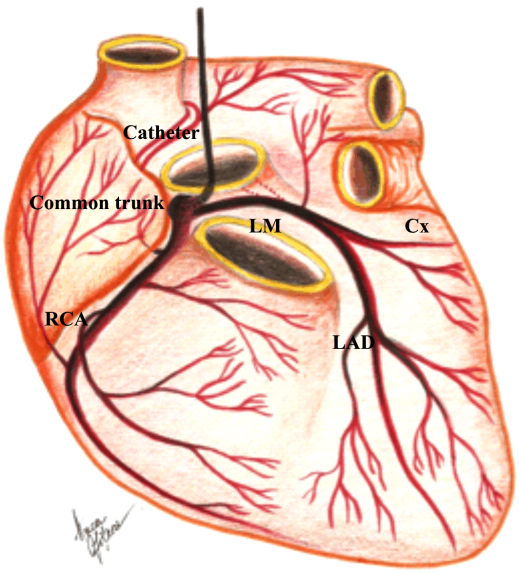


**Figure 4** – ***Schematic representation of the coronary anomaly. Cx: Circumflex (artery); LAD: Left anterior descending (artery); LM: Left main (artery); RCA: Right coronary artery.***

The patient was discharged with good general condition without recurrence of chest pains, no shortness of breath, with the recommendation to continue the medication and periodic reassessment.

## Discussion

The arterial circulation of the heart in very rare cases can be supplied by a SCA, with the origin arising from right, left or posterior sinus of Valsalva [4]. The etiology of the SCA remains uncertain [6]. The SCA may lead in some cases to life threatening conditions, such as angina pectoris, congestive heart failure, myocardial infarction, cardiac arrhythmias, syncope, or even sudden death [1]. Despite neither of those syndromes was found in the present case and the hemodynamic status was normal, we encouraged the follow-up, because the obstruction of the SCA could be fatal.

Coronary angiography provides accurate information in congenital anomalies regarding the origin, the course, and the termination of the arteries. The diagnostic key point of this case was that the arterial supply of the heart was provided by a SCA arising from the right sinus of Valsalva. The knowledge about this rare coronary anomaly is helpful in the differential diagnosis and helps the physician in the future treatment planning.

SCA arising from left, right or posterior sinus of Valsalva is a rare congenital anomaly, with an incidence of 0.024% to 0.06% [1]. From a series of 126 595 subjects investigated by coronary angiography, SCA arising from the right coronary sinus of Valsalva was described in 0.019% of cases [3].

There are several classifications for coronary artery abnormalities. The Lipton *et al.* [1] classification of the coronary abnormalities is based on the origin of the SCA, the anatomical course and its relationship with the ascending aorta and pulmonary trunk (Table 1).

**Table 1 –** ***Lipton et al. [1] classification of single coronary artery***

| | Classification | Description |
|---|---|---|
| Ostial location | R | Right sinus of Valsalva. |
| | L | Left sinus of Valsalva. |
| Anatomical distribution | I | Solitary dominant vessel follows the course of either a normal right or left coronary artery. |
| | II | One coronary artery arises from the proximal portion of the normally located other coronary artery. |
| | III | LAD and Cx arteries arise separately from a common trunk originating from the right sinus of Valsalva. |
| Course of the transverse trunk | A | Anterior to the great vessels. |
| | B | Between the aorta and pulmonary artery. |
| | P | Posterior to the great vessels. |
| | S | Septal type passes through the interventricular septum. |
| | C | Combined type: a combination of diverse routes. |

R: Right (sinus); L: Left (sinus); LAD: Left anterior descending (artery); Cx: Circumflex (artery); A: Anterior; B: Between; P: Posterior; S: Septal; C: Combined.

Type L means that right coronary artery (RCA) has the origin from the left main and type R shows that coronary artery originates from the RCA. These types are also classified from I to III. In class I, the course of the left coronary artery or the right coronary artery follows their normal anatomical course. Class II means that one coronary artery arises from the proximal segment of the opposite coronary artery that has normal origin. Class III: the left anterior descending artery and the circumflex artery have their origins separately from the proximal segment of a normal right coronary artery. The II and III classes are classified also depending on the relation with the pulmonary artery in: type A for anterior course, type B for inter-arterial course (the course of the coronary artery is between the pulmonary trunk and ascending aorta), and type P for posterior course. According to this classification, the type B morphology, when associated with an intramural route, has a high risk of severe clinical consequences [6]. A different classification was proposed more recently by Angelini *et al.* [7], according to the course of coronary artery within the atrio-ventricular groove and the inter-ventricular septum. The type S – septal, when the course is through interventricular septum and type C – the combined type [8].

Anatomical variations among the coronary arteries are well known, most of them describe the presence of a common trunk arising either from the right sinus of Valsalva or from the left sinus of Valsalva [9–11]. In the literature, there are several classifications of the coronary artery anomalies, the origin of the left coronary artery from the right sinus of Valsalva either from the proximal segment of the right coronary artery or as a separate vessel [12]. The first classification of SCAs was Smith's [13]. He classified in three types: Type 1 – a SCA that followed the course of only one coronary artery, Type 2 – SCA divided immediate into left or right branches with normal distribution of blood supply, Type 3 – SCA with atypical distribution of the coronary arteries [13].

Another classification was proposed by Ogden & Goodyer, based on letter "R" – origin from the right sinus of Valsalva or "L" – origin from the left sinus of Valsalva, followed by numbers (1 to 4 for the L and 1 to 5 for the R) [14].

The embryology of the coronary artery is very important. It requires a well-organized array of molecular and morphogenetic events [15]. Recent studies showed that the organogenesis of the coronary arteries could be based on three essential steps: vasculogenesis, arterio-genesis and angiogenesis [16, 17]. Studies in cell lineage tracing and quail chick chimera propose that proepicardial epithelium (PEO) is the source of the coronary arteries. It has a mesothelial origin and is located between the liver and the sinus venosus. The proepicardial cells are involved in the developing of the heart, after their formation the cells migrate and colonize the latter, and contribute to the formation of epicardium. A small part of the PEO separates from the primordial epicardium and develops epicardium-derived cells (EPDCs). These EPDCs fuse, migrate in anterior and posterior way and give rise in the atrioventricular and interventricular grooves to a primitive vascular plexus [18]. Those new organized vessels branch out, elongate and then invade

the aorta [19, 20]. After, the orifices open most often in the right and the left sinus, very rare in the posterior sinus. However, there were described multiple aortal orifices for coronary arteries [21–24].

Our case respects the classification for type R-IIP. In the literature, the most common type of anomalies from type R-II is the sub-group R-IIS, while the coronary anomalies type R-IIP sub-group are very rare.

## Conclusions

Our patient underwent a complete cardiologic examination for his symptoms (atypical angina, chest pain and dyspnea). According to the electrocardiogram, transthoracic echocardiography, to the laboratory results and clinical examination, we suspected coronary artery disease. After we performed the coronary angiography, the diagnosis of atherosclerotic coronary artery disease was excluded, but the patient had a very rare disease: single coronary artery, a congenital anomaly of the coronary system that provides a lower perfusion to the entire myocardium, which can lead to symptoms such as chest pain, angina pectoris or dyspnea.

**Conflict of interests**

Nothing to declare.

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