



## Case Report

**Inferior Myocardial Infarction and Total AV Block in a Patient with Single Ostium in the Right Sinus of Valsava (A Rare Congenital Coronary Anomaly)**Imam Mahbub Zam Zami<sup>1\*</sup><sup>1</sup>Department of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia.

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## ABSTRACT

A 51-year-old male suffered from a STEMI inferior and a total AV block. Echocardiography shows hypokinetic wall motion at the inferior and inferoseptal that result in decreased EF. Coronary angiography revealed a single ostium of coronary artery without any stenosis. Cardiac CT revealed a single coronary artery arises from a single ostium from RCC and divided into RCA and LCA. There was an inter-arterial course of proximal RCA and proximal LCA between aorta and pulmonary artery. There was an acute angle take off of RCA from aorta. Inter-arterial course and acute angle take off of coronary artery from aorta result in kinking and narrowing of coronary artery that contributes to myocardial infarction. There is a mismatch between myocardial demand which is increased during exertion and myocardial oxygen delivery that decreased during exertion. A surgical anterior pulmonary transposition is the suggested choice of therapy.

**Introduction**

Single coronary artery (SCA) is a rare congenital anomaly reported to lead to ischemia in the absence of atherosclerosis. Single coronary artery (SCA), the presence of only one coronary trunk arising from the aorta, is a rare congenital coronary artery anomaly with a prevalence of 0.024% to 0.066% [1]. Various subtypes of SCA are described [2,3]. It is reported to lead to angina pectoris, myocardial infarction and even death, even in the absence of associated atherosclerosis.[4,5]

**Case Presentation**

A 51-year-old man was referred by our hospital with a recent diagnosis of inferior STEMI onset 48 hours and total AV block. The patient experiences chest pain and weakness while playing badminton.

Echocardiography showed a hypokinetic wall motion at inferior and inferoseptal and the decrease of systolic function (EF by Teich 52%). The diagnostic coronary angiography revealed a single ostium of coronary artery without any stenosis. The cardiac CT revealed a coronary artery originates from one ostium in the right coronary cuspis (RCC) and divides into two branches; they are the right and left coronary artery (RCA and LCA). LCA then was divided into small LAD and small LCX. There is an inter-arterial pathway of the LCA between the pulmonary artery and the aorta. RCA take off angle was almost zero degrees from the aorta, while the LCA take off angle was still 45 degrees from the aorta with a small caliber LAD. There was no significant stenosis.

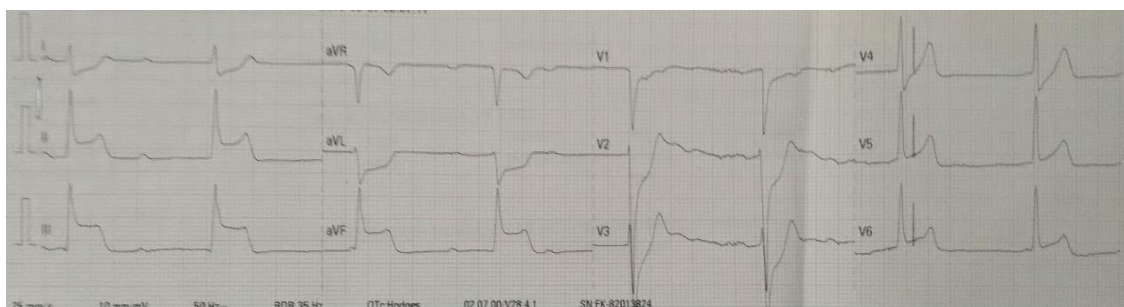


Figure 1. ECG of the patient shows an inferior STEMI and total AV block



Figure 2. Cardiac CT

Picture from top left to bottom right:

- Single ostium coronary
- LCA branches run inter-arterial between the aorta and pulmonary artery
- There is no slit ostium image which shows narrowing of the LCA which runs inter-arterial between the pulmonary artery and the aorta
- There was no stenosis on RCA, the PDA branch came from RCA which showed dominant right
- RCA take off angle was almost zero degrees from the aorta, while the LCA take off angle was still 45 degrees from the aorta with a small caliber LAD image

The STEMI and TAVB then spontaneously resolved. Patient was in stable and good condition. Before discharged, he was then subjected to an exercise stress test (EST) to determine functional capacity. EST was stopped because a limiting chest pain at 7.0 METS. He was refused to surgery. Daily activity was limited according to the functional capacity obtained from the EST results.

## Discussion

Single coronary artery is an extremely rare congenital anomaly with a single coronary ostium from the aortic trunk. The two arteries can either share a proximal common trunk with an ectopic origin of one of the arteries from the opposite sinus or one of the arteries can originate from a distal segment of the contralateral artery.<sup>[6]</sup>

In this case we define the anomaly into Lipton R-IIB type. SCA cases are classified by Lipton et al. and modified by Yamanaka and Hobbs.<sup>[1]</sup>

First, depending on the coronary ostial location, these are classified as “R” type (with origin from right sinus of Valsalva) and “L” type (with origin from left sinus of Valsalva). Further classification is based on anatomical course of the coronary arteries into type I, II and III. In R-II type, only RCA arises from aorta. However, after its origin, RCA gives origin to left main coronary artery (LCA), which reaches its normal position on the left side and then further divides into LAD and LCX (which are usually smaller in caliber).

The final designation depends on the course of the anomalous vessel as it crosses to other side of the heart. “A” denotes course anterior to great vessels (i.e. aorta and pulmonary artery). “B” denotes course between pulmonary artery and aorta. “P” denotes course posterior to the great vessels. “S” is used when part of the course is through inter-ventricular septum (also called as myocardial bridge). “C” stands for combination of diverse routes <sup>[7]</sup>. Types “B” and “S” are considered as potentially “malignant” types, since coronary artery can get compressed between the aorta and pulmonary artery (in type B) and between myocardial muscle fibers (type S), with the potential to cause sudden ischemic events and death. With exercise or physical activity, the blood flow through major vessels such as aorta and pulmonary artery

as well as myocardial contractility increases, predisposing these anomalously coursing arteries for compression.<sup>[8-10]</sup>

In nearly 15% of patients with SCA, myocardial ischemia can develop in the absence of atherosclerosis. During exercise, a mismatch between myocardial demand (which is increased during exertion) and myocardial oxygen delivery, which is dependent upon the single coronary blood flow, which may fail to increase in relation to demand or may even decrease during exertion. This limited or diminished coronary blood flow is probably the result of one or many high risk anatomic and physiologic factors: (a) flap closure of the slit-like deformation of the coronary ostium; (b) acute (non-orthogonal) angle of take-off and kinking of the coronary artery as it exits from the aorta; (c) hypoplasia and/or stenosis of the intramural segment, particularly at the level of the valvar commissure.<sup>[6,11,12]</sup>

Angiography can definitely distinguish between the intramural and the intraseptal course and the coexistence of atherosclerotic <sup>[13,14]</sup>. The presence of a MACE was associated with a coronary minimal lumen area  $\leq 4$  mm<sup>2</sup>, an area stenosis  $\geq 50$  %, a longer intra-arterial course ( $>10$  mm) and a smaller width of proximal segment assessed by CTA <sup>[15,16]</sup>.

Pulmonary translocation has been specifically used in cases where there is a single coronary ostium and no intramural segment as in this case. Both branch PAs are fully mobilized and right branch PA is transected and moved anterior to the aorta. The right PA is reattached and a pericardial patch is added, as necessary. This moves the main PA both anteriorly and leftward, relieving compression on the inter-arterial portion of the anomalous artery.<sup>[17,18]</sup>

In patients with SCA who have symptoms such as chest pain, syncope, ventricular arrhythmia, SCD, or aborted SCD, and/or the presence of ischemia on stress testing should be restricted from competitive sports and should undergo surgical correction [16]. Cardiopulmonary Exercise Testing (CPET) is beneficial after the diagnosis of a coronary anomaly has been established. Postoperatively, it is also used before allowing the patient to return to competitive sports [19]. Resumption of competitive sports should usually be allowed 3 months after the intervention if they have no inducible ischemia and no symptoms.[16]

## Conclusion

From the result of the research, it was found that a single coronary artery type Lipton RII-B may cause inferior myocardial infarction and total AV block without atherosclerotic process.

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