

# Coronary Artery Connection Abnormality Revealed by Acute Coronary Syndrome: A Case Report and Review of the Literature

H. Nabawi <sup>a\*</sup>, M. Boutgourine <sup>a</sup>, B. Maatouf <sup>a</sup>, I. Rhoujati <sup>a</sup>, M. Eljamili <sup>a</sup>  
and M. Elhattaoui <sup>a</sup>

<sup>a</sup> Department of Cardiology, Mohammed VI University Hospital, Marrakesh, Morocco.

## Authors' contributions

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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Case Study

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## ABSTRACT

Coronary artery connection abnormalities are rare with an angiographic prevalence of approximately 1%. Their symptomatology is very varied, from incidental discovery to sudden death by myocardial ischemia, especially during exercise, hence the importance of early diagnosis and management. Angiographic diagnosis is generally easy, but it is sometimes difficult to specify the initial course of the ectopic vessel, hence the interest of non-invasive imaging, in particular the coronary CT scan. This latter represents the innovative imaging of choice for assessing the anatomical risk and selecting candidates for surgical treatment. We report a case of abnormal connection of the left coronary revealed by an acute coronary syndrome and we present a review of the literature on the different anatomical forms of the pathology and its management.

**Keywords:** Coronary artery connection abnormalities; sudden death; coronary angiography; the coronary CT scan.

## 1. INTRODUCTION

Coronary artery connection abnormalities are rare and polymorphic with an angiographic

prevalence of approximately 1% [1,2]. Their clinical manifestations are diverse from incidental discovery to sudden death by myocardial ischemia especially when the abnormal coronary

\*Corresponding author: E-mail: hindnabawi1@gmail.com;

artery arises from the opposite sinus of Valsalva [3,4]. Thus, the initial course of the ectopic coronary is important to consider, because the prognosis of these abnormalities depends on the anatomic shape [1].

The contribution of non-invasive imaging is described for the positive diagnosis of these anatomical variations, sometimes delicate in coronary angiography, but also for the distinction of benign forms and malignant forms potentially responsible for myocardial ischemia [5,6]. Thus, the difficulties encountered in their management are explained by the numerous anatomical forms described and the lack of data on their follow-up whether treated or not [1,7].

In the present article, we report a case of abnormal connection of the left coronary birth at the right sinus revealed at the age of 80 years on the occasion of a high-risk NSTEMI and we present a review of the literature on the different anatomical forms of the pathology by highlighting the management which remains poorly codified.

## 2. CASE PRESENTATION

80-year-old patient followed for arterial hypertension under treatment, no other specific pathological history or cardiovascular risk factor. Admitted in a cardiology intensive care unit for an acute coronary syndrome without high-risk ST-segment elevation, he presented a constrictive, intense, prolonged retrosternal chest pain radiating to the left upper limb without any notion of dyspnea, palpitations, loss of consciousness or other associated signs. He had a good hemodynamic and respiratory condition with a physical examination without abnormalities.

His electrocardiogram (ECG) was in sinus rhythm and showed a transient ST-segment elevation in the inferior part without Q wave of necrosis with a mirror image in the high lateral part (Fig. 1).

Transthoracic echocardiogram was performed detecting a preserved systolic function and size of the cardiac chambers with disorders of segmental kinetics. The ultrasensitive troponins were high without any other detectable anomalies.

Coronary angiography, performed urgently through the right radial approach, revealed an abnormal artery of the left coronary artery born from the antero-right sinus (Fig. 2) associated with a short tight stenosis of the distal circumflex

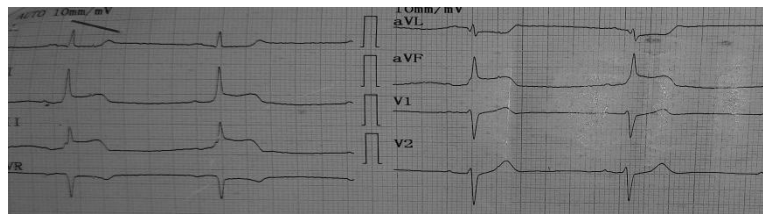
artery (CX), Tight stenosis of the middle right coronary artery (DC), double intermediate stenosis of the distal DC and tight stenosis of the posterior interventricular artery, who underwent angioplasty of the middle right coronary artery with placement of an active stent after predilation [8,9]. The patient was put on medical treatment combining double antiplatelet aggregation, beta-blocker, ACE inhibitor, gastric protection and statin.

The coronary CT scan confirmed the diagnosis showing the abnormal connection by a common trunk with the right coronary artery from the right antero-lateral sinus and describes an inter-lateral aortopulmonary malignant path [10,11].

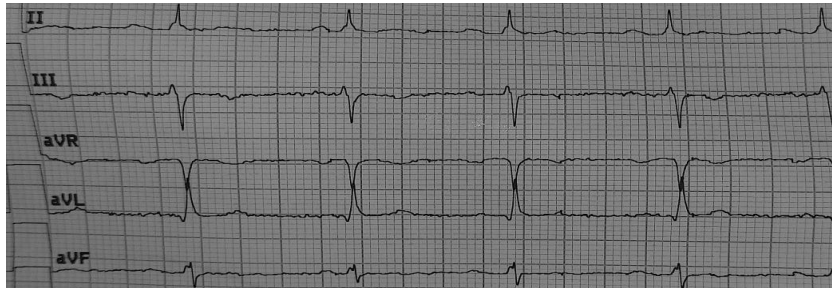
## 3. DISCUSSION

Coronary artery connection abnormalities are the most common congenital coronary anomalies. Their angiographic prevalence is close to 1% [1]. The ectopic connection of the circumflex artery is the most concerned (50% of cases) versus 30% for that of the right coronary and 20% for that of the common trunk or the IVA artery. Indeed, the recognition of this pathology requires to define beforehand what is a normal coronary network by including the anatomical variants, in order not to make an excessive diagnosis [1]. The normal number of coronaries, usually two, may increase to three if the IVA and circumflex arise separately at the level of the left sinus. The place of connection may be debatable when it involves the normal sinus but with an unusual location (abnormally low or high or very close to the contralateral sinus). This makes the angiographic diagnosis between an anatomical variant and a Coronary artery connection abnormally difficult [12].

The main Coronary artery connection abnormalities are represented by the connection in the contralateral sinus or artery, the connection in a normal sinus but in an eccentric position, the abnormally high aortic connection and the so-called single coronary. The majority of these patients are asymptomatic with incidental discovery. The symptomatology consists most often of chest pain on exertion, palpitations, syncope on exertion and sudden death which is the most dreadful complication [3,4,13]. The revelation of these abnormalities by an acute coronary syndrome remains rare apart from an associated atheromatous disease after the age of 40 [1].

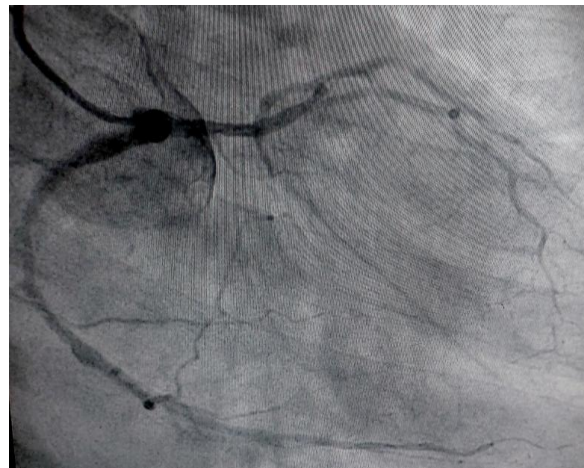


(A)



(B)

**Fig. 1. (A-B): ECG showing a transient ST-segment elevation in the inferior leads without Q wave of necrosis**



**Fig. 2. Coronary angiography showing Abnormal connection of the left coronary birth at the right sinus**

The angiographic diagnosis of this pathology is generally easy, but it is sometimes difficult to specify the initial path of the ectopic vessel. Non-invasive imaging techniques are now essential to determine the relationship of the ectopic vessel with the adjacent structures, mainly the aorta and the pulmonary artery [5,6,14]. In our patient, these abnormalities concern the ectopic connection of the common trunk (CT) in the right anterior sinus, and several paths of the CT are possible (Fig. 3), whose relationship with the arterial trunks defines four anatomical types [7]:

- Type A with the trunk passing in front of the pulmonary artery (pre-pulmonary path)
- Type B with the trunk passing between the aorta and the pulmonary artery (inter-arterial path)
- Type C with the trunk passing in the infundibular septum (intra septal path);
- Type D with the trunk passing behind the aorta (retro aortic path).

The recommendations of the European Society of Cardiology have well defined the anatomical forms at high risk of myocardial ischemia

evaluated by The coronary CT scan, which are represented by an intramural pathway, an interarterial pathway, hypoplasia of the proximal coronary artery, orifice abnormalities (cleft deformity, acute angle of departure of the coronary artery, high orifice located > 1 cm above the sinotubular junction) [15]. Recommended non invasive ischemia tests are

stress echocardiography, stress cardiac MRI, and stress scintigraphy [15].

The surgical indications for Coronary artery connection abnormalities according to the European Society of Cardiology recommendations published in 2020 are represented in Table 1.

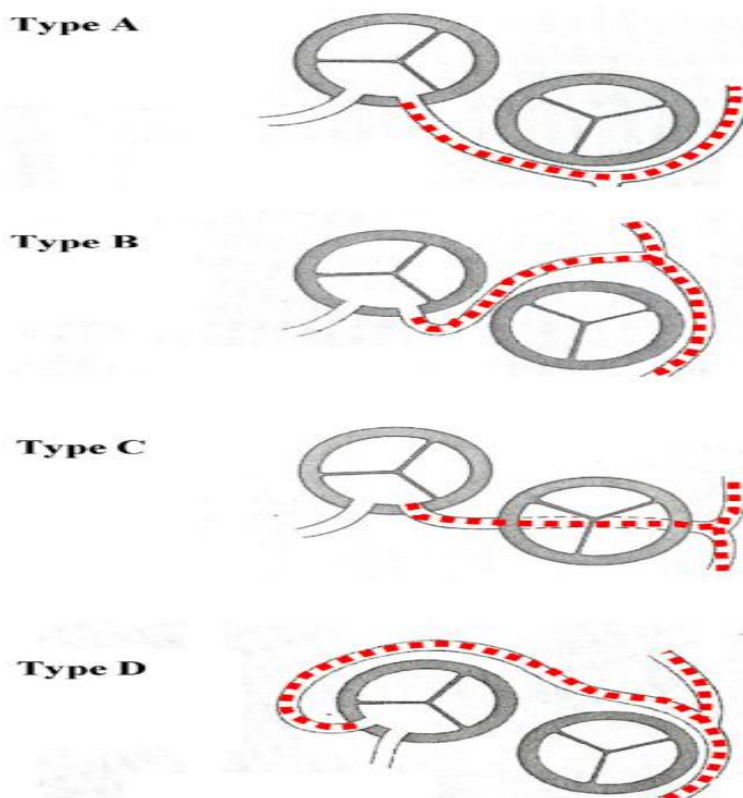


Fig. 3. Diagram of the 4 anatomical types of common trunk connection in the right sinus

Table 1. Surgical indications for Coronary artery connection abnormalities according to the recommendations of the European Society of Cardiology [11]

Anomalous aortic origin of the coronary artery		
Surgery is recommended for AAOCA in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischaemia in a matching territory or high-risk anatomy. <sup>c</sup>	I	C
Surgery should be considered in <i>asymptomatic</i> patients with AAOCA (right or left) and evidence of myocardial ischaemia.	IIa	C
Surgery should be considered in <i>asymptomatic</i> patients with AAOLCA and no evidence of myocardial ischaemia but a high-risk anatomy. <sup>c</sup>	IIa	C
Surgery may be considered for symptomatic patients with AAOCA even if there is no evidence of myocardial ischaemia or high-risk anatomy. <sup>c</sup>	IIb	C
Surgery may be considered for <i>asymptomatic</i> patients with AAOLCA without myocardial ischaemia and without high-risk anatomy <sup>c</sup> when they present at young age (<35 years).	IIb	C
Surgery is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy. <sup>c</sup>	III	C

#### 4. CONCLUSION

Coronary artery connection abnormalities are a rare entity. Their symptomatology is very varied, from incidental discovery to sudden death by myocardial ischemia. According to the recommendations of the European Society of Cardiology published in 2020, the coronary CT scan is currently the recommended imaging technique for the evaluation of high-risk anatomical forms of myocardial ischemia with the aim of codifying surgical management.

Indeed, the pooling in the form of a national registry of these abnormalities with their anatomical, clinical, morphological, and therapeutic particularities could eventually provide some answers to the questions still raised.

#### CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

#### ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

#### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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