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Anomalous Aortic Origin of Coronary Artery: A Cases Series

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Abstract **Case Report**

Congenital coronary artery anomalies (CCAA) are not that common, they are usually found in 0.3 to 5.6% of people undergoing coronary angiography [1], these numbers are still to be widely confirmed since they can be biased by the fact that angiography or autopsy are usually preformed in patients with suspected cardiac disease. However, they are particularly important as they occupy the second place of most common cause of sudden death in young athlete according to a series reported by Basso *et al.,* [2]. Definition, clinical presentation, physiopathology of symptoms, diagnostic workup and prognosis, can be challenging and varies according to different subgroups established by classifications, this last one can falls into two paths, anatomical based on the description of origin, course, termination and functional based on the degree of ischemia that can be caused by it. The management and treatment of these anomalies is still not firmly entrenched, it depends mostly on the patient, circumstances of diagnosis and proof of hemodynamic significance. In this case series of 3 patients, we will focus mainly of anomalous aortic origin of a coronary artery, encountered in our cathlab.

Keywords: Congenital coronary artery anomalies, non-ST segment myocardial infarction, angina, inter-arterial course, malignant course.

INTRODUCTION

Although the prevalence of anomalous aortic origin of coronary artery (ANOCOR) in the general population is low, more frequent use of invasive and non-invasive imaging to rule out coronary artery disease has seen an increase in absolute numbers of ANOCOR.

Clinical manifestations of these anomalies can range from sudden cardiac death to fortuitous discovery during an invasive or non-invasive imaging. Conversely, it seems that older people with ANOCOR are less predisposed to adverse cardiac events.

Non-invasive anatomic imaging (coronary CT for example) is complementary to invasive imaging and helps to further identify high-risk anatomic features. Using functional non-invasive perfusion imaging can assess potential ischaemia induced by dynamic compression of malignant ANOCOR. Information gained from clinical imaging guides the management of these patients.

ANOCOR are traditionally classified as malignant (with an interarterial course) and benign variants. Malignant variants have been recognised in autopsy studies to be an underlying cause of sudden cardiac death in young athletes [2].

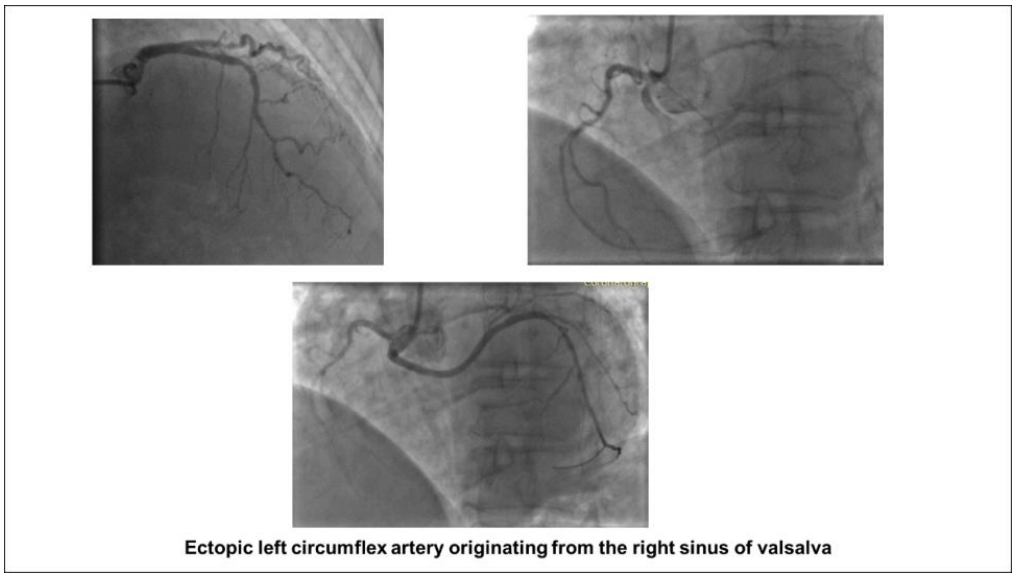
We report 3 cases of patients with an anomalous aortic origin of coronary artery.

1st CASE

56 years old woman, with hypertension and dyslipidemia and no other medical or surgical history, was admitted in our cathlab for the management of a non-ST segment myocardial infarction (NSTEMI)

Coronary angiography (CA) showed a particular coronary anatomy with the left circumflex artery (LCX) taking origin from the right sinus of valsalva (RSV), while the left anterior descending artery (LAD) took origin from the left one. The LCX had a significant thrombotic lesion in the mid-section, with no other significant lesion in the rest of the coronary system.

Knowing that ANOCORs with LCX originating from the right sinus of valsalva, has typically a retro aortic course, considered as benign, an Ad hoc angioplasty was successfully performed via a direct stenting using a drug eluting stent. cf. Figure 1.



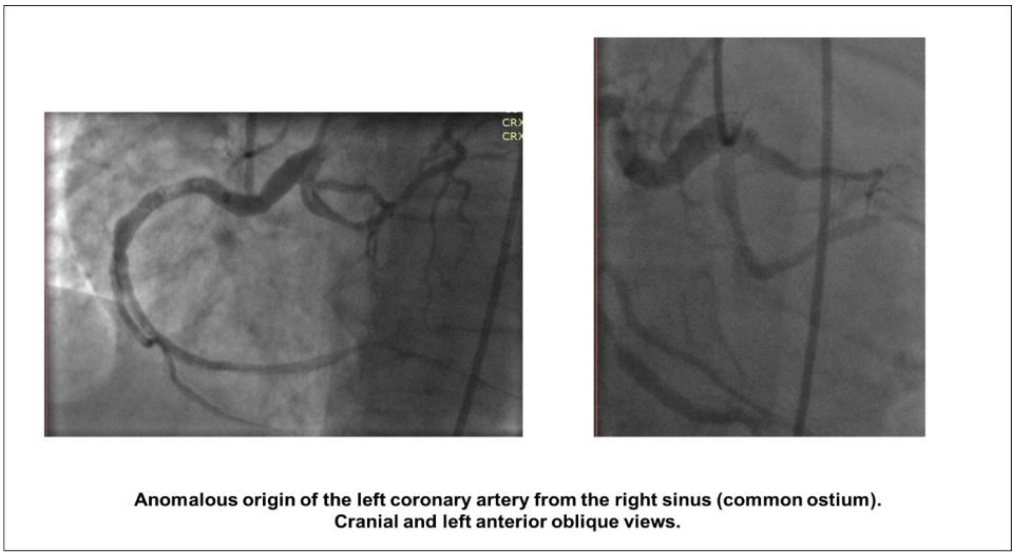
Ectopic left circumflex artery originating from the right sinus of valsalva

Figure 1

2nd CASE

A 69 years old woman with hypertension and a history of an inferior STEMI, was admitted for coronarography on the basis of a typical angina (class II of CCS) associated to a positive stress test.

Coronarography found an entire left coronary circulation arising from the right sinus of valsalva (RSV). The right coronary artery (RCA) had a normal origin and course with a significant thrombotic lesion in the proximal section of the third section. The left coronary circulation took origin from the same ostium as the right, after a common left main stem the coronary divided into a Cx and LAD. cf. Figure 2.



Anomalous origin of the left coronary artery from the right sinus (common ostium).
Cranial and left anterior oblique views.

Figure 2

3rd CASE

54 years old woman with diabetes, with the history chronic coronary syndrome that beneficiated from PCI of the LAD 10 years ago, was admitted for the management NSTEMI with a long history of angina, even after the PCI.

Coronary angiography showed a patent LAD stent, an anomalous origin of the RCA from the left sinus of valsalva, an inter-arterial course was confirmed with a coronary CT.

Surgical reparation was later indicated (malignant course), since then the patient is totally asymptomatic. cf. Figure 3.

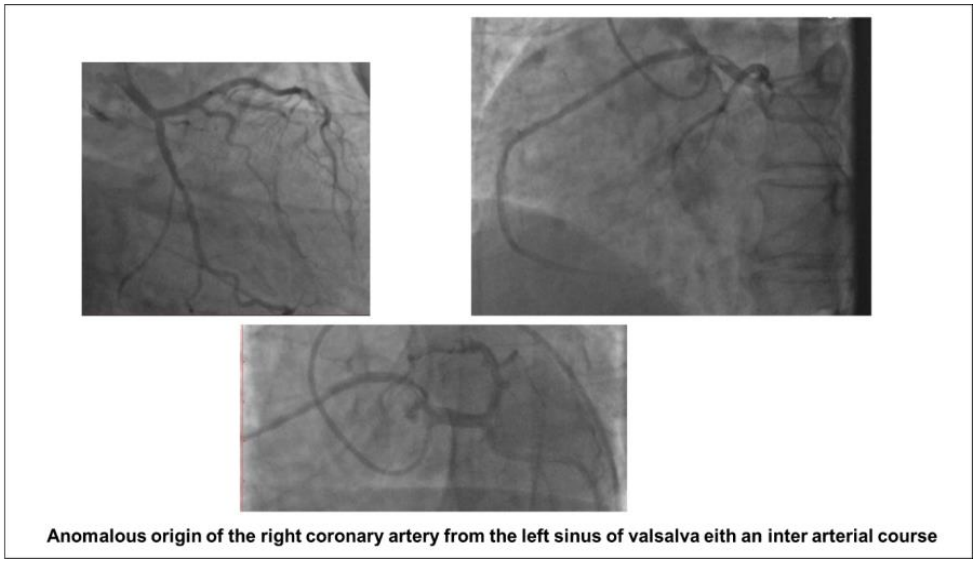


Figure 3

DISCUSSION

When it comes to defining CCAAs, the main issue is establishing what the normal spectrum of variation is. In this matter, Angelini and coworkers [3] proposed to classify "normal" as every feature with > 1% of frequency in an unselected general population, this leave us with a definition that require a frequency of less than 1%. Normal variants can be represented by a separate origin of the conus branch [4], myocardial bridging, sheperd's crook RCA and many other forms seen every day via imaging modalities.

Anomalous aortic origin of a coronary artery is rare with a prevalence of 0.3% to 3% according to series [5, 6] 5/1000 coronarographies and 13/1000 CT, but it affects the left main artery (LMA) in less than 1 per 1000.

The commonest anomaly is a separate origin of the left anterior descending artery (LAD) and left circumflex artery (LCX), with an incidence of 0.41%, followed by LCX arising from the right coronary artery (RCA), with an incidence of 0.37%.

The diagnosis via coronary angiography is usually easy, but it's difficult to define the course of the ectopic artery, this is where different noninvasive imaging modalities find place, especially Coronary CT that can provide information of the exact origin, course and relationship with other cardiac structure, particularly the aorta and pulmonary artery.

Classification of different anomalies is built on 2 main features, anatomical and functional.

Based on the functional relevance of each abnormality, CCAAs can be classified as: (1) anomalies with obligatory ischemia, as seen in anomalous origin of the left main coronary artery (LMCA) from the pulmonary artery (2) anomalies without ischemia; this group contains the majority of CCAAs encountered; and (3) anomalies with exceptional ischemia: This is a group of CCAAs that only occasionally cause critically severe clinical events, but are otherwise compatible with leading a normal life, including athletic training.

The risk correlated with a CCAA usually depends on the location and course of the anomalous origin of coronary artery. A coronary artery that arises from the contralateral sinus of Valsalva has five potential paths it may take to its perfusion territory:

1. Pre-pulmonic (type A): In this case usually there are no hemodynamic consequences, although in a small proportion of cases there may be an association with angina.
2. Inter-arterial (type B): Between the aorta and pulmonary artery. This course is associated with more severe prognosis and increased risk of sudden cardiac death for reasons, which remain unclear. One of them is based on the fact that exercise leads to expansion of the aortic root and pulmonary trunk and this may increase the existing angulation of the coronary artery, decreasing the luminal diameter. Another hypothesis is based on the fact that the vessel has an aberrant course within the aortic wall and is often hypoplastic and exposed to a lateral compression over the entire proximal intramural tract (intussusception into the aortic wall). However, in these patients, resting electrocardiograms are usually normal and stress

tests are not always positive for inducible ischaemia [2].

3. Trans-septal (type C): Sometimes it can be difficult to differentiate a trans-septal coronary artery from an inter-arterial one.
4. Retro-aortic (Type D): This variant does not seem haemodynamically significant, but may complicate valve surgery.
5. Retro-cardiac: In this case the path is behind mitral and tricuspid valves, in the posterior AV groove.

It has been proposed that the abnormal origin and course of anomalous coronary arteries could make them more prone to atherosclerosis. A study by Jiang *et al.*, aimed to characterize coronary artery disease (CAD) among adults diagnosed with an anomalous aortic origin of a coronary artery and found that it didn't appear to increase the severity [7].

These course anomalies can originate from:

- Anomalies of origin of left coronary artery from the right sinus of valsalva (RSV): The left coronary artery can arise from a separate or a commun ostium with the RCA, or from the proximal portion of the RCA. Usually it is easy to identify using a catheter designed for the RCA.
- Anomalies of origin of left circumflex artery (LCX) from the right sinus of valsalva (RSV) or right coronary artery (RCA): LCX and RCA can arise from a common ostium or from separate ostia. It may also arise as a proximal branch of the RCA. Usually LCX courses posterior to the aorta and provides branches to the LV lateral wall. In the absence of coronary atherosclerosis, the anomaly may be considered benign (3).
- Anomalies of origin of right coronary artery from the left sinus of valsalva or left coronary artery (LCA): The selective opacification of RCA in this case is usually laborious, the use of different imaging modality is necessary.

On the subject of clinical presentation, it can vary from patients dying suddenly at young age after extreme exertion to totally asymptomatic for a large portion of their lives, and an atypical chest-pain syndrome is the most common reason they are referred for coronary angiography, which is when the diagnosis is typically made.

The management of CCAAs and its different subgroups is still not clarified, for both indications and revascularization techniques.

For anomalous aortic origin of a coronary artery classified as benign, therapeutic abstention is the rule.

For ANOCORs categorized as major or hemodynamically significant, surgical treatment should be discussed, especially for a left main with an inter arterial course [8].

Surgical correction, may consist of osteoplasty, which creates a new ostium at the end of the ectopic artery's intramural segment (technique of choice), of direct reimplantation of the ectopic artery at the aortic root (a technically difficult and unreliable approach, it may cause a restenosis); unroofing of the intramural coronary segment, from the ostium to the exit point, off the aortic wall.

Surgical correction can be proposed for right ANOCORs with a documented ischemia, even though percutaneous angioplasty is also doable [8].

Age, symptomatology, documented ischemia by a stress test, myocardial territory, association with atherosclerosis and the need for an intense activity (profession) should also play a role in the decision making of, wether to correct or not an ANOCOR at high risk.

Series published by Mirchandani S *et al.*, and Van Son *et al.*, have few patients, but the results are in favor of surgical treatment, with a net decrease in ischemia related symptoms and sudden cardiac death risk [9, 10].

CONCLUSION

Anomalous aortic origin of a coronary artery is quite infrequent and for most of the time asymptomatic, but it's crucial to bring it to light as it can be related to sudden death, even if for the majority, it remains a simple anatomical curiosity. Surgical treatment should be proposed every time an ischemic proof was made.

REFERENCES

1. Villa, A. D., Sammut, E., Nair, A., Rajani, R., Bonamini, R., & Chiribiri, A. (2016). Coronary artery anomalies overview: The normal and the abnormal. *World journal of radiology*, *8*(6), 537-555. doi: 10.4329/wjr.v8.i6.537
2. Basso, C., Maron, B. J., Corrado, D., & Thiene, G. (2000). Clinical profile of congenital coronary artery anomalies with origin from the wrong aortic sinus leading to sudden death in young competitive athletes. *Journal of the American College of Cardiology*, *35*(6), 1493-1501.
3. Angelini, P. (1989). Normal and anomalous coronary arteries: definitions and classification. *American heart journal*, *117*(2), 418-434.
4. Boffano, C., Chiribiri, A., & Cesarani, F. (2009). Native whole-heart coronary imaging for the identification of anomalous origin of the coronary arteries. *International Journal of Cardiology*, *137*(1), e27-e28.

5. Click, R. L., Holmes, D. R., Vlietstra, R. E., Kosinski, A. S., & Kronmal, R. A. (1989). Anomalous coronary arteries: location, degree of atherosclerosis and effect on survival—a report from the Coronary Artery Surgery Study. *Journal of the American College of Cardiology*, *13*(3), 531-537.
6. Sloan, K., Majdalany, D., Connolly, H., & Schaff, H. (2008). Inferior wall myocardial infarction caused by anomalous right coronary artery. *Canadian Journal of Cardiology*, *24*(12), S102-S103.
7. Michael X.JiangMD, MEng[a]Ellen K.BrinzaMS[b] Joanna GhobrialMD[c] Dominique L.TuckerBS[d] Sohini GuptaBA[d] Jeevanantham RajeswaranPhD[e] Tara KaramlouMD, MSc
8. Warnes, C. A., Williams, R. G., Bashore, T. M., Child, J. S., Connolly, H. M., Dearani, J. A., ... & Webb, G. D. (2008). ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the american college of cardiology/american heart association task force on practice guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease) developed in collaboration with the american society of echocardiography, heart rhythm society, international society for adult congenital heart disease, society for cardiovascular angiography and interventions, and .... *Journal of the American College of Cardiology*, *52*(23), e143-e263.
9. Mirchandani, S., & Phoon, C. K. (2005). Management of anomalous coronary arteries from the contralateral sinus. *International journal of cardiology*, *102*(3), 383-389.
10. Van Son, J. A., & Mohr, F. W. (1997). Modified unroofing procedure in anomalous aortic origin of left or right coronary artery. *The Annals of thoracic surgery*, *64*(2), 568-569.