Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Cardiology

Coronary Artery Connection Abnormalities: About Four Cases

Benlafkih Otmane[1*], Abderazzaq Chachi[1], Mohcine Naim[1], El Jamili Mohammed[1], El Karimi Saloua[1], Benzarouel Dounia[1], El Hattaoui Mustapha[1]

[1]Cardiology Department, University hospital, Marrakech, Morocco

DOI: 10.36347/sjmcr.2022.v10i04.018 | **Received:** 28.02.2022 | **Accepted:** 04.04.2022 | **Published:** 19.04.2022

***Corresponding author:** Benlafkih Otmane
Cardiology Department, University hospital, Marrakech, Morocco

Abstract **Case Report**

Coronary artery connection abnormalities are a diverse group of congenital disorders whose manifestations and pathological mechanisms are highly variable. This abnormalities are undergoing significant changes in terms of description, morphogenesis, clinical presentation, diagnostic assessment, prognosis and treatment. It is an important entity because of its clinical impact, the risk of sudden death, its prevalence and its practical management. The rapid development of imaging techniques such as computed tomography, magnetic resonance imaging, intravascular ultrasound and optical coherence tomography has brought a lot of new information on the issue. It is most often associated with sudden cardiac mortality if the abnormal connection of a coronary artery from the controlateral sinus, especially if it passes between the aorta and the pulmonary artery. Patients are often asymptomatic and coronary abnormalities are incidentally identified during coronary angiography or autopsy after sudden cardiac death in the majority of cases. Symptoms such as angina pectoris, syncope, heart failure and myocardial infarction can occur under certain circumstances. The purpose of this article is to provide a brief overview of the various variants of this disease, focusing on some clinical manifestations, the risk of sudden cardiac death and the physiopathological mechanism of symptoms, as well as valuable information on diagnostic assessment and treatment options.

Keywords: Abnormal coronary artery connection; inter-arterial route; sudden death; myocardial ischemia.

ABBREVIATIONS
LCX: Left circumflex artery
LAD: Left anterior descending artery
MRI: Magnetic resonance imaging

INTRODUCTION

Congenital coronary artery abnormal connections are an important issue in cardiology and cardiovascular surgery. It is an important entity because of its clinical impact, the risk of sudden death, its prevalence and its complexity of practical management. They concern 0.3 to 1.5% of the general population. The abnormal connection of the coronary artery is a common type of congenital anomaly of the coronary artery. Several classification schemes have been proposed for this congenital disease. Based on their clinical significance, some authors classify them as major and minor. From the point of view of their functional impact, they can be classified as follows: 1- abnormalities associated with a defined ischemia 2- abnormalities not associated with ischemia and 3- abnormalities with exceptional ischemia. However, the most detailed and accurate classification was proposed by Angelini [1] and is based on anatomical features. Based on this classification, it can be characterized as: 1- coronary connection and pathway abnormalities; 2- abnormalities of the intrinsic coronary anatomy; 3- coronary termination abnormalities; and 4- abnormal collateral vessels [2]. The abnormal origin of the coronary artery has important clinical implications due to its association with myocardial ischemia [3], lethal arrhythmia [4] and even sudden cardiac death [5]. In some patients, atherosclerotic coronary disease or valvular heart disease may be associated with abnormal coronary artery connection [6]. As a result, this lesion is often referred to as a "malignant" coronary artery abnormality [7]. The management of these coronary abnormalities requires a multidisciplinary approach, which includes the clinical cardiologist, interventional cardiologist, radiologist and the cardiovascular surgeon. In this report, we describe our experiences with 4

patients in whom an abnormal coronary artery origin was found during coronary angiography.

Case 1

A 15-year-old patient with no particular pathological history and no cardiovascular risk factor admitted for cardiac arrest recovered in the school after 3 minutes of cardiopulmonary resuscitation. Upon admission, after the patient was discharged, the clinical examination and electrocardiogram were normal. Cardiac ultrasound a preserved systolic function and size of the cardiac chambers. Coronary angiography revealed connection defect of the left coronary artery from the antero-right sinus. The coronary CT scan confirmed the diagnosis showing the birth of the left coronary artery of a common stem with the right coronary artery from the right antero-lateral sinus and describes an inter-aortopulmonary malignant path without atheroscholic coronary infiltration (Figure 1). Magnetic resonance imaging (MRI) showed the same angiographic and CT findings, and it did not find an argument in favour of interstitial fibrosis, or myocardial infarction (Figure 2). The case is presented in multidisciplinary session, it is decided to proceed to surgery with creation of a neo-ostium. In surgery, we repositioned the left coronary artery, creating a new ostium in the appropriate aortic sinus with good evolution.

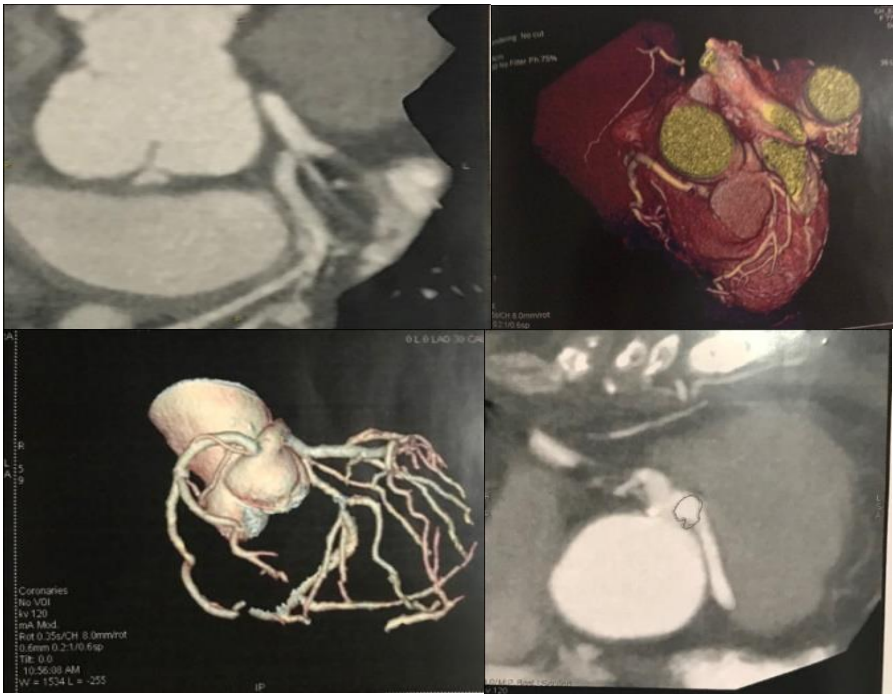
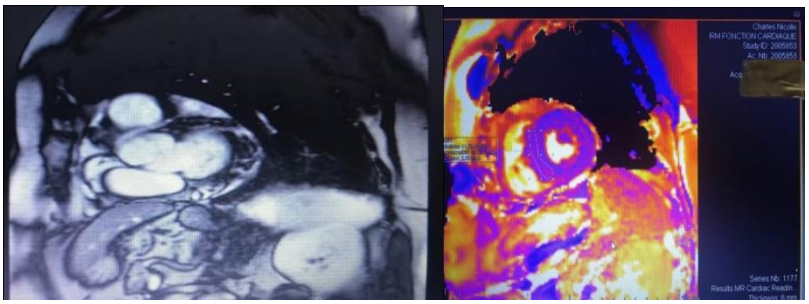


Figure 1: The coronary CT Scan showing the abnormal connection of the left coronary artery of a common stem with the right coronary artery from the right antero-lateral sinus and describes an inter-aortopulmonary malignant path



Figures 2: The MRI showing the common connection by a right ostium of both coronaries

Case 2

80-year-old patient followed for arterial hypertension under treatment, no other specific pathological history or cardiovascular risk factor, admitted for acute coronary syndrome with ST segment elevation. Cardiac ultrasound a preserved systolic function and size of the cardiac chambers with disorders of segmental kinetics. Coronary angiography revealed an abnormal artery of the left coronary artery born from the antero-right sinus; with significant distal right coronary artery stenosis (Figure 3). 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.

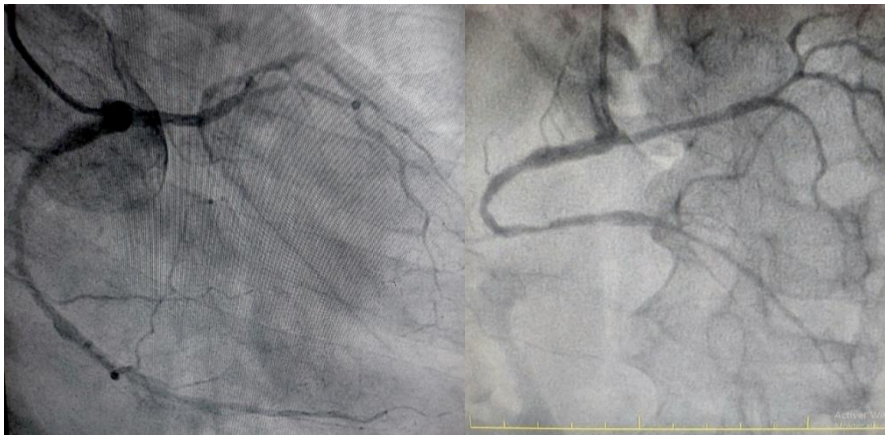
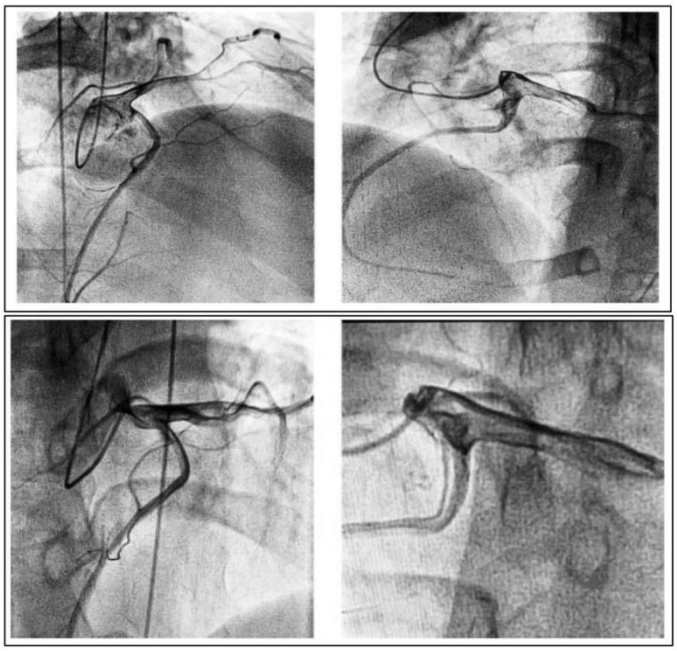


Figure 3: Abnormal connection of the left coronary birth at the right sinus

Case 3

65-years-old patient, with chronic smoking, followed for right ventricular arrhythmogenic dysplasia. The patient was admitted for chest pain. He had a good hemodynamic and respiratory condition with a physical examination without abnormalities. An electrocardiogram was performed recording a regular sinus rhythm, a right branch block with secondary repolarization disorders. The ultrasensitive troponins was high. Other biological tests, including serum electrolytes, were normal, as was chest radiography. As a result, the diagnosis of acute coronary syndrome without ST segment elevation was retained. A transthoracic echocardiogram was performed detecting a preserved systolic function and size of the cardiac chambers, correct global and segmental kinetics, the right ventricle was not dilated, hypertrophied with an akinesia of its side wall suggestive of right ventricular dysplasia. Coronary angiography was performed showing coronary arteries without significant stenosis (Figure 4). However, the right coronary artery had an abnormal connection in the left sinus more precisely in the left trunk. Acute aortic syndromes and pulmonary embolisms were excluded. Thus, the possibility of an abnormal path of this artery especially in inter-aorto-pulmonary was suspected. A thoracic coronary scanner was requested which unfortunately was not performed due to patient refusal.



Figures 4: Abnormal connection of the right coronary birth at the left sinus in the left trunk

Case 4

48-years-old patient, followed for diabetes and high blood pressure. The patient was admitted for chest pain radiating to both shoulders. He had a good hemodynamic and respiratory condition with a physical examination without abnormalities. An electrocardiogram was performed recording a regular sinus rhythm, negative T-waves in the inferior territory. A dose of ultrasensitive troponins was carried out with a high result rate. Cardiac ultrasound a preserved systolic function and size of the cardiac chambers without disorders of segmental kinetics. Coronary

angiography was performed showing the right coronary artery had an abnormal connection in the left sinus and a significant stenosis of the distal right coronary artery confirmed by fractional flow reserve study. An angioplasty of the distal right coronary artery was perfomed with stent placement. A coronary CT was later planned confirming the diagnosis.

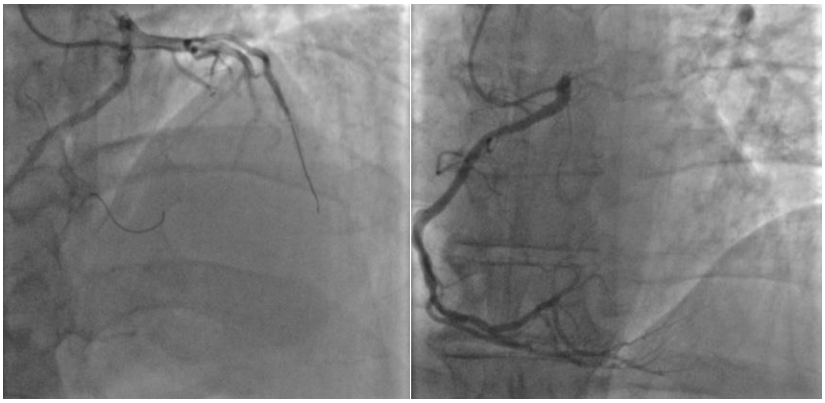


Figure 5: Abnormal connection of the right coronary birth at the left sinus

DISCUSSION

Defining which anatomy of the coronary arteries is normal can be difficult. Some normal characteristics may be defined in numerical terms (for example, the number of coronary ostia), while in other cases a more qualitative description is required. Angelini and colleagues proposed a classification dividing the coronary characteristic into two groups: 1- normal coronary anatomy, defined as any morphological characteristic observed in > 1% of the non-selected sample. This group also includes normal anatomical variants, defined as alternative and relatively unusual morphological features observed in > 1% of the population; and 2- abnormal coronary anatomy, defined as morphological features found in < 1% of the population [1, 8-10]. About 26% of coronary abnormalities involve some kind of aortic root abnormality such as a bicuspid aortic valve [1, 8-11]. The most common coronary abnormality is a distinct origin of LAD and LCX, with an incidence of 0.41%, followed by RCA with an incidence of 0.37% [9, 11-14]. Although congenital coronary artery connection abnormalities are relatively rare but they are the second most common cause of sudden cardiac death in young athletes. The most common abnormality associated with sudden cardiac death is the abnormal connection of a coronary artery, especially with a path from the aorta to the pulmonary artery[10]. The ectopic connection of the coronary artery can be from: 1- the aorta, either from a false sinus, or beyond the sinuses; 2- the pulmonary artery; 3- as a branch of another coronary artery; 4- other arteries; and 5- the ventricular chamber [15;16]. There are some complex anatomical variants regarding the categories of ectopic connection of the coronary artery: the "multiple" type, involving more than one coronary artery or branch, which can be subdivided into two sub-types: 1- more than one coronary artery or branch from one location; and 2- two coronary arteries/branches from separate ectopic sites; and "complex" type, associated with acquired heart disease or congenital heart defects [16, 17]. The abnormal origin of a coronary artery from Valsalva's contralateral sinus has a particular clinical interest, because these abnormalities may be associated with sudden cardiac death, especially when the abnormal coronary artery intersects inter-arterially between the aorta and the pulmonary artery [2, 18-20]. They represent 12-19% of the causes of sudden death among young athletes [20-22]. Several types of aberrant pathways are described: inter-arterial, retro-aortic or retro cardiac, pre-pulmonary or pre-cardiac, intra-septal or sub-pulmonary. Apart from the inter-aortopulmonary path that can be the cause of sudden death, these abnormalities in the path of the coronary branches have no clinical consequence. The clinical symptoms of a patient with coronary artery abnormality vary depending on the group to which the abnormality belongs. In addition to ischemia, other clinical consequences may occur. Fistulas can cause volume overload. Aortic root distortion may be observed in patients with very large coronary fistula or aneurysms. Complications from aortic valve surgery or coronary angioplasty are described [8-11, 13, 23, 24]. The origin of the coronary artery from the innominate artery can be the cause of syncope [25] or chest pain in adults [26]. The origin of the right coronary artery from the descending thoracic aorta may be associated with atypical and striking elastotonic changes and a thickening of the coronary artery wall as an underlying pathogenesis with serious consequences [27]. Circulatory symptoms may also stem from the ectopic coronary artery pathway between the pulmonary trunk and the aorta despite the absence of atherosclerotic plaques in the coronary artery [28]. The abnormal connection of the coronary artery may be associated with a common congenital cardiac malformation [29] or a rare congenital cardiac malformation such as the cervical aortic arch [30]. Robiseck [31] reported one case in a 4-year-old boy associated with a Fallot tetralogy that was successfully repaired without any

post-operative complications. The abnormal origin of the coronary artery from other arteries other than coronary arteries is often associated with more complex congenital cardiac malformations and treatment is more difficult and the prognosis is darker [32]. As for myocardial ischemia, the mechanism most often mentioned is an increase of pressure and dilation at the level of the aorta and the pulmonary artery during exertion, responsible for coronary compression [33]. The second hypothesis put forward to explain myocardial ischemia is the circuitous path of these coronary arteries running between the aorta and the pulmonary artery [34] which is potentially responsible for a coronary artery fold. This plicature may be responsible for myocardial hypoperfusion in the territory of the affected coronary artery. Finally, some pathologists suggest the presence of fibrous folding in the ostium of ectopic coronary arteries whose implantation is tangentially to the aortic wall [4, 34]. This fibrous folding is potentially responsible for a stenosis of the ostium of the coronary artery during an increase in pressure at the root of the aorta, resulting in myocardial ischemia, especially stress. The abnormal origin of the coronary artery can sometimes be associated with acquired heart disease, including coronary artery disease, heart valve disorders and cardiomyopathy. Acute myocardial infarctions are reported in patients with abnormal coronary artery origin with diffuse coronary stenosis lesions [35]. The diagnostic assessment should also include electrocardiography and holter monitoring. If at least two normally located coronary ostia can be identified by echocardiography, no further investigation is required. However, if the echocardiographic results are inconclusive, additional imaging is recommended [2, 9, 36]. Coronarography allows the diagnosis of this type of coronary abnormality in the majority of cases. Nevertheless, a detailed analysis by the multi-slice coronary scanner with electrocardiogram synchronization has many advantages in the detection and exploration of coronary artery travel abnormalities [37]. MRI appears to be a promising examination for the exploration of these abnormalities as it is a non-irradiating examination, requiring no iodine contrast injection. Some authors propose the realization of a 3D angio-MRI with respiratory synchronisation for the assessment of these ectopic arteries [37]. But MRI, in this indication, remains, despite these advantages, a longer and less reproducible imaging technique than the multi-slice scanner. The inter-arterial path known as the coronary killer presents risks to note an arterial compression between the 2 large vessels, a stress paroxysmal rhythm disorders, a chronic ischemic heart disease. Thus an accurate and early diagnosis of this risky path is essential in order to assess the need for surgical treatment which is heavy and aggressive, alone can prevent the risk of sudden death. CT scan is therefore valuable for selecting potential candidates for cardiac surgery in patients with such malformations. A particular case that must also be mentioned before an arterial path is the one that comes after the surgical repair of a congenital heart disease. This is the case of patients who have a coronary artery transfer after an arterial switch for transposition of large vessels or patients in whom a tube has been interposed between the right ventricle and the pulmonary artery for a right obstacle. Several surgical techniques have been described [38]. Some are aimed at removing the pulmonary artery from the abnormal coronary artery. Others aim to open the intramural path. Others aim to create a coronary neo-ostium at the appropriate coronary sinus.

CONCLUSION

Abnormal connections of the coronary arteries are rare. Among these anatomical variations, those with the greatest risk of complication are those whose aberrant path runs between the trunk of the pulmonary artery and the aorta. They can be responsible of myocardial ischemia and sudden death in the young patients. It is therefore necessary to systematically investigate this anomaly, at least in all young adults who engage in intense sports activities.

These anomalies are easily detected and characterized by the scanner's spatial resolution. Knowledge of the clinical impact and prognosis of coronary artery travel abnormalities, including the most dangerous arterial, allows for better preventive surgical management.

Conflicts of Interest: The authors declare no financial interest nor any other conflict of interest.

Informed Conscent: The patients conscented.

REFERENCES

1. Angelini, P. (2007). Coronary artery anomalies: an entity in search of an identity. *Circulation*, *115*(10), 1296-1305.
2. Kastellanos, S., Aznaouridis, K., Vlachopoulos, C., Tsiamis, E., Oikonomou, E., & Tousoulis, D. (2018). Overview of coronary artery variants, aberrations and anomalies. *World journal of cardiology*, *10*(10), 127-140.
3. Aydin, M., Ozeren, A., Peksoy, I., Cabuk, M., Bilge, M., Dursun, A., & Elbey, M. A. (2004). Myocardial ischemia caused by a coronary anomaly: left circumflex coronary artery arising from right sinus of valsalva. *Texas Heart Institute Journal*, *31*(3), 273-275.
4. Cheitlin, M. D., De Castro, C. M., & MCALLISTER, H. A. (1974). Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva: a not-so-minor congenital anomaly. *Circulation*, *50*(4), 780-787.
5. Greet, B., Quinones, A., Srichai, M., Bangalore, S., & Roswell, R. O. (2012). Anomalous right coronary artery and sudden cardiac

death. *Circulation: Arrhythmia and Electrophysiology*, *5*(6), e111-e112.
6. Kimbiris, D. E. M. E. T. R. I. O. S., Iskandrian, A. S., Segal, B. L., & Bemis, C. E. (1978). Anomalous aortic origin of coronary arteries. *Circulation*, *58*(4), 606-615.
7. Golubickas, D., Motiejūnaitė, J., Jankauskas, A., Šlapikas, R., & Basevičius, A. (2013). Incidentally diagnosed malignant coronary artery anomaly: a clinical case. *Medicina*, *49*(10), 462-465.
8. Angilini, P. (2002). Coronary artery anomalies-current clinical issues. *Tex Heart Inst J*, *29*, 271-278.
9. Angelini, P., Velasco, J. A., & Flamm, S. (2002). Coronary anomalies: incidence, pathophysiology, and clinical relevance. *Circulation*, *105*(20), 2449-2454.
10. 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.
11. Young, P. M., Gerber, T. C., Williamson, E. E., Julsrud, P. R., & Herfkens, R. J. (2011). Cardiac imaging: Part 2, normal, variant, and anomalous configurations of the coronary vasculature. *American Journal of Roentgenology*, *197*(4), 816-826.
12. Yamanaka, O., & Hobbs, R. E. (1990). Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Catheterization and cardiovascular diagnosis*, *21*(1), 28-40.
13. Yurtdas, M., & Gülen, O. (2012). Anomalous origin of the right coronary artery from the left anterior descending artery: review of the literature. *Cardiology Journal*, *19*(2), 122-129.
14. Alexander, R. W., & Griffith, G. C. (1956). Anomalies of the coronary arteries and their clinical significance. *Circulation*, *14*(5), 800-805.
15. Duncan, J. R., & Byard, R. W. (2022). Sudden Infant Death Syndrome: An Overview. In: Duncan, J. R., Byard, R. W., éditeurs. SIDS Sudden Infant and Early Childhood Death: The Past, the Present and the Future [Internet]. Adelaide (AU): University of Adelaide Press; 2018 [cité 3 mars 2022]. Disponible sur: http://www.ncbi.nlm.nih.gov/books/NBK513399/
16. Yuan, S. M. (2014). Anomalous origin of coronary artery: taxonomy and clinical implication. *Brazilian Journal of Cardiovascular Surgery*, *29*, 622-629.
17. Chaitman, B. R., Bourassa, M. G., Lesperance, J., Dominguez, J. L., & Saltiel, J. (1975). Aberrant course of the left anterior descending coronary artery assoicated with anomalous left circumflex origin from the pulmonary artery. *Circulation*, *52*(5), 955-958.
18. Corrado, D., Thiene, G., Nava, A., Rossi, L., & Pennelli, N. (1990). Sudden death in young competitive athletes: clinicopathologic correlations in 22 cases. *The American journal of medicine*, *89*(5), 588-596.
19. Right ventricular cardiomyopathy and sudden death in young people - PubMed [Internet]. [cité 3 mars 2022]. Disponible sur: https://pubmed.ncbi.nlm.nih.gov/3336399/
20. Burke, A. P., Farb, A., Virmani, R., Goodin, J., & Smialek, J. E. (1991). Sports-related and non-sports-related sudden cardiac death in young adults. *American heart journal*, *121*(2), 568-575.
21. Taylor, A. J., Rogan, K. M., & Virmani, R. (1992). Sudden cardiac death associated with isolated congenital coronary artery anomalies. *Journal of the American College of Cardiology*, *20*(3), 640-647.
22. Maron, B. J., Poliac, L. C., & Roberts, W. O. (1996). Risk for sudden cardiac death associated with marathon running. *Journal of the American college of cardiology*, *28*(2), 428-431.
23. Flessas, D., Mamarelis, I., Maniatis, V., Souretis, G., Laschos, N., Kotoulas, C., & Lazaridis, K. (2013). An unusual pattern of three major components of the cardiovascular system: multimodality imaging and review of the literature. *Journal of Cardiothoracic Surgery*, *8*(1), 1-7.
24. Camarda, J., & Berger, S. (2012). Coronary artery abnormalities and sudden cardiac death. *Pediatric cardiology*, *33*(3), 434-438.
25. Santucci, P. A., Bredikis, A. J., Kavinsky, C. J., & Klein, L. W. (2001). Congenital origin of the left main coronary artery from the innominate artery in a 37-year-old man with syncope and right ventricular dysplasia. *Catheterization and cardiovascular interventions*, *52*(3), 378-381.
26. Duran, N. E., Duran, I., & Aykan, A. Ç. (2008). Congenital anomalous origin of the left main coronary artery from the innominate artery in a 73-year-old woman. *The Canadian Journal of Cardiology*, *24*(12), e108.
27. Cheatham, J. P., Ruyle, N. A., McManus, B. M., & Gammel, G. E. (1987). Origin of the right coronary artery from the descending thoracic aorta: angiographic diagnosis and unique coronary artery anatomy at autopsy. *Catheterization and cardiovascular diagnosis*, *13*(5), 321-324.
28. Mahajan, D., Agnihotri, G., & Brar, R. (2012). Anomalous origin of right coronary artery: an anatomico-clinical perspective of 2 cases. *Acta Informatica Medica*, *20*(1), 56-57.
29. McMahon, C. J., DiBardino, D. J., Ündar, A., & Fraser, C. D. (2002). Anomalous origin of left coronary artery from the right pulmonary artery in association with type III aortopulmonary window and interrupted aortic arch. *The Annals of thoracic surgery*, *74*(3), 919-921.
30. Charrot, F., Tarmiz, A., Glock, Y., & Léobon, B. (2010). Diagnosis and surgical treatment of an aneurysm on a cervical aortic arch associated with an anomalous origin of the left main coronary

artery. *Interactive cardiovascular and thoracic surgery*, *10*(2), 346-347.

31. Robicsek, F. (1984). Origin of the left anterior descending coronary artery from the left mammary artery. *American Heart Journal*, *108*(5), 1377-1378.
32. Liu, F., Huang, G., & Zhang, J. (2010). Anomalous origin of a coronary artery from the right branchiocephalic trunk associated with complex congenital heart disease. *Pediatric cardiology*, *31*(1), 163-165.
33. Trivellato, M., Angelini, P., & Leachman, R. D. (1980). Variations in coronary artery anatomy: Normal versus abnormal. *Cardiovascular diseases*, *7*(4), 357-370.
34. Virmani, R., Chun, P. K., Goldstein, R. E., Robinowitz, M., & Mcallister, H. A. (1984). Acute takeoffs of the coronary arteries along the aortic wall and congenital coronary ostial valve-like ridges: association with sudden death. *Journal of the American College of Cardiology*, *3*(3), 766-771.
35. Cho, H. O., Cho, K. H., Jeong, Y. S., Ahn, S. G., Choi, S. J., Yoo, J. Y., & Kim, E. J. (2006). Anomalous origin of the left coronary artery from the right sinus of Valsalva, which presented as acute myocardial infarction. *Korean Circulation Journal*, *36*(12), 817-819.
36. Post, J. C., van Rossum, A. C., Bronzwaer, J. G., de Cock, C. C., Hofman, M. B., Valk, J., & Visser, C. A. (1995). Magnetic resonance angiography of anomalous coronary arteries: a new gold standard for delineating the proximal course?. *Circulation*, *92*(11), 3163-3171.
37. McConnell, M. V., Ganz, P., Selwyn, A. P., Li, W., Edelman, R. R., & Manning, W. J. (1995). Identification of anomalous coronary arteries and their anatomic course by magnetic resonance coronary angiography. *Circulation*, *92*(11), 3158-3162.
38. Aydinlar, A., Çiçek, D., Sentürk, T., Gemici, K., Serdar, O. A., Kazazoglu, A. R., ... & Cordan, J. (2005). Primary Congenital Anomalies of the Coronary Arteries A Coronary Arteriographic Study in Western Turkey. *International heart journal*, *46*(1), 97-103.