

Our experience regarding congenital coronary arteries anomalies at King Hussien Hospital

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Abstract

Aim: To evaluate the frequency and different types of congenital coronary arteries anomalies and their imaging features.

Methods: Our retrospective study will include 5250 patients, aged 18-75 years, of both sexes, with history of chest pain at King Hussein Hospital during the 2020 period. There are many types of congenital coronary arteries anomalies for example: when the right coronary artery (RCA) arising from the left coronary sinus instead of the right coronary sinus this anomaly is the most common anomaly, the other anomalies include left main coronary artery (LM) arising from the right coronary sinus instead of the left coronary sinus, Anomalous origin of the left coronary artery from the pulmonary artery and absent of any of the coronary arteries.

Conclusions: Multiple types of congenital coronary arteries are present, some of them are asymptomatic, the others are predisposing for myocardial ischemia and could be lethal.

The coronary arteries congenital anomalies are not common but are very vital to detect early, to avoid complications.

It can be occur in isolation or associated with other congenital anomalies.

Keywords: coronary artery disease, coronary CT angiography, congenital coronary arteries anomalies

Introduction

The word congenital is defined as the disease that present since birth, congenital coronary arteries anomalies refers to the abnormalities regarding the structure origin and courses of the coronary arteries. It is seen in 1% of whole population, with severity ranging from asymptomatic in mild cases to sudden death in young patients (1).

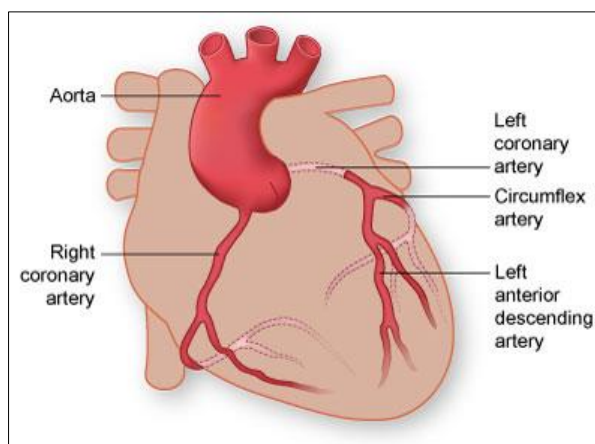


Fig 1: Normal Coronary arteries anatomy.

Materials and Methods

Retrospective study performed at King hussien hospital of Royal Medical services at Radiology department for 1250 patients of both genders, aged between 18-75 years, presented with chest symptoms such as chest pain and shortness of breath (SOB).

Coronary CT angio study was performed to rule out any evidence of coronary artery diseases (CAD). The patients were scanned by SIEMENS CT (Somatom Force 256-slice) during the 2020. There was no exclusive criteria except for low quality studies that showed severe breathing artifacts. Patient's preparation included kidney function test to assess the creatinine level and measurement of the heart rate. During the exam the patients were asked to hold their breath and while IV contrast was given images were obtain. The images were viewed via Syngo-via software station using Multiplanar reconstruction MPR, curved multiplanar reformations cMPRs and 3-Dimensional reconstruction techniques. All the patient's images were examined and reviewed by two radiology specialists in two separate sessions with all results being analyzed by simple statistical methods.

Discussion

The coronaries arises from the aortic sinuses distally to the aortic valve (also known as Valsalva sinuses), the function of the coronary artery is to supply oxygen to the heart myocardium, there are 3 aortic sinuses: two of them considered coronary sinuses, the anterior sinus which gives the RCA and the left posterior sinus which gives the left main coronary artery and then divided into the LAD and the CLX, finally the right posterior sinus is the non-coronary sinus, no coronary artery arises typically from this sinus.

From histology point of view: each coronary artery wall is consist of three layers (from outside to inside layers are): tunica adventitia, tunica media and the innermost layer which is tunica

intima (2).

The wall of the coronary artery is consist from three layers: 1-Tunica intima which is the innermost layer, 2-Tunica media which is the middle layer, 3- Tunica adventitia which is the outmost layer (2).

during the embryonic period (at the first 8 weeks of gestation) major coronaries start to develop and for the following weeks it get increased in diameter with increase thickness of tunica media layer, at the 18th week the coronary arteries increase in number and gets more thickness of tunica media and showed expansion of tunica adventitia due to addition of elastic fibers (3)

Congenital coronary arteries anomalies are very rare which is seen in 1-2 % of population, however there is two types of congenital coronary arteries anomalies 1)hemodynamic significant, 2)Non hemodynamic significant. The former include absent of coronary artery (atresia), arising of coronary artery from pulmonary artery, Myocardial bridging (this occur when the coronary passes within the myocardium instead of epicardial fat) and congenital fistulas

The non-hemodynamic significant: they are usually incidental finding and include many types like high aortic origin of coronary arteries or from anomalous position like arising from other coronary artery or cusp, shepherd crook course anomaly of the RCA(4).

Single coronary artery or atresia of the coronary artery is extremely rare congenital anomalies, it occur when there is a single coronary which follows the normal pattern of right coronary artery or left coronary artery, the most common form of this anomaly is atresia of the left main coronary artery, also a few cases of other coronary arteries atresia has been reported, there is high risk of sudden cardiac death if the single coronary passes between ascending thoracic aorta and pulmonary trunk the origin of the coronary artery from pulmonary artery is also very rare seen in 0.2-0.3% of all congenital coronary anomalies cases, although the pulmonary artery can gives any coronary artery the most common coronary to do this is the left coronary artery hence the name ALCAPA which referred to (Anomalous left coronary artery from pulmonary artery), There are two forms of ALCAPA 1)infantile: the most severe form, most of the infants die within first year of life. 2) Adult: discovered and manifest in adulthood, lead to serious cardiac events such as multiple cardiac ischemia, heart failure and eventually sudden death.

Anomalies of the coronary artery origin and courses this occur when any coronary artery arise from another contralateral coronary or sinus, which mean when the RCA arises from the left coronary or left coronary cusp or when the LM coronary arises from the right coronary or cusp, there are two types 1)benign course seen in majority of cases and not associated with stenosis or ischemia, types of benign courses are retroaortic, transeptal and prepulmonic courses 2)malignant course (interarterial) the coronary passes and compressed between the pulmonary trunk and ascending thoracic aorta, this compression is usually significant and can lead to sudden death hence the name malignant.

Myocardial bridging is not uncommon congenital anomaly of coronary arteries and defined as when the coronary artery courses within the myocardium muscle instead of epicardial fat, in most of cases it consider benign harmless condition but in small percentage of patients it may lead to myocardial infarction and

chest pain, there are two types of bridging 1)superficial 2)deep. LAD coronary artery is the most common coronary to manifest bridging specially at the mid segment, but it still can occur in all other coronaries.

Congenital fistulas are rare congenital coronary arteries anomaly and defined as when there is abnormal connection between coronary artery and a cardiac chamber (most commonly the right ventricle) or to a vascular structure (most commonly the pulmonary artery) with enlargement and tortuosity of the artery. Fistulas could be acquired due to many causes such as post coronary artery bypass grafting (CABG), Post catheterization, post cardiac transplant and chest trauma. Congenital shunt usually lead to right to left shunt and this leads to less blood supply to the myocardium leading to (steal phenomena)

Non-hemodynamically significant congenital coronary arteries anomalies

Duplication: duplication or split of the coronary artery is rare congenital disorder, both RCA and LAD coronaries can be duplicated, most commonly occur in the LAD coronary, in even more rare cases the duplicated LAD coronary arise from the RCA and make a malignant course, nine subtypes of LAD duplication has been described in the literature.

High origin this occur when the coronary artery arise above the sino tubular junction by 1 cm or more, it consider hemodynamic stable but makes catheterization difficult, so it is important to recognize high origin coronary artery before any intervention, high origin RCA occur more commonly than LAD, and studies reveals close relationship between high origin of the RCA and bicuspid aortic valve.

Shepherd crook deformity: it occur exclusively to the RCA and characterize by being torturous and high course but from a normal origin from the right coronary sinus (unlike the other coronary anomalies), it is also hemodynamic stable but makes catheterization difficult and increase risk of complication.

Systemic termination: the is occur when there is termination of the coronary artery into systemic artery, it is important to differentiate this entity from coronary fistula, in fistula cases there is enlargement and torturous course of the coronary artery which is not the case in systemic termination

Benign courses: prepulmonic course this occur when any coronary artery passes anterior to the pulmonary artery or posterior to the ascending thoracic aorta, for example when the RCA or LM or LAD or CLX coronary arteries arises from the contralateral coronary artery or cusp, and unlike the malignant course when coronary passes behind the pulmonary artery and anterior to the ascending thoracic aorta in the prepulmonic course they passes anterior to the pulmonary artery, no compression will occur so no increased risk of sudden death, this course is highly associated with tetralogy of fallot

Transseptal course is occur when the coronary artery arise from the contralateral coronary or cusp and passes via the myocardial septa, it looks like interarterial artery so it is important to differ between two of them, in the transseptal course it is surrounded by myocardium and takes downward course and does not have slitlike or oblong orifice while the interarterial variant it is surrounded by epicardial fat and does not have downward course and have oblong or slitlike orifice

Retroaortic course occur when the coronary artery arise from the contralateral coronary artery or cusp and passes behind the

ascending thoracic aorta and interatrial septum, it is consider benign course with no compression and no increased risk of death. Multiple ostia: it typically occur when the coronary artery and one of its branches arise directly from the aortic sinus (for example the RCA and the conus artery) or when the LAD coronary artery and the CLX artery arises separately from the left coronary sinus without LM coronary artery, presence of multiple ostia increase the difficulty of catheterization and increase risk of its complications.

Results

From the 5250 patients underwent Coronary CT angio in this study only fifty one patients had coronary congenital anomaly the incidence is 0.97%.

Our study revealed that forty nine patients (96%) had anomalies of origin with either benign or malignant course, two patients had coronary artery fistula (4%) of these fifty one patients forty patients were males (78.4%) and eleven patients were females (21.5%).

Table 1: incidence of congenital coronary artery anomalies

	Number of patients	% of total CT angios	% of total anomalies
Total coronary CT angios	5250		
Total anomalies	51	0.97%	
Origin and course anomalies	49	0.93%	96%
Coronary artery fistulas	2	0.03%	4%

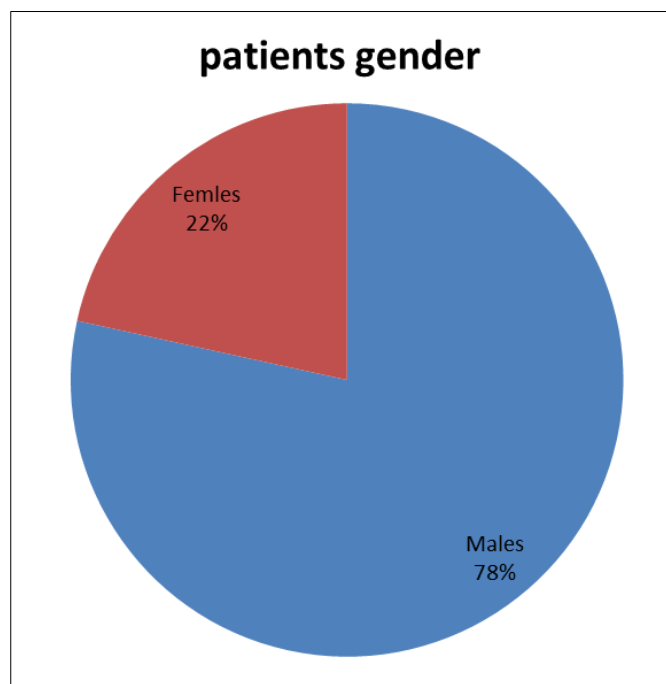


Fig 2

Table 2: incidence of congenital coronary artery anomalies according to gender

Types of origin anomalies	Number of patients	Angiographic %	Anomaly %
RCA arise from LCS or LAD	15	0.28	30.7%
LAD arise from RCS or RCA	13	0.24	26.5%
CLX arise from RCS or RCA	7	0.13	14.2%
LAD and CLX ariseseparately	8	0.15	16.3%
High RCA origin or shepherd crook deformity	3	0.057	6.1%
Absent Coronary artery	2	0.038	4.1%
ALCAPA	1	0.019	2.1%

Table 3: types of origin anomalies of coronary arteries.

Type of course anomalies	Number of patients	Angiographic %	Anomaly %
Malignant course	10	0.19%	28.6
Benign (Prepulmonic)	9	0.17%	25.7
Benign (Transseptal)	5	0.09%	14.3
Benign (Retroaortic)	11	0.20%	31.4

Table 4: types of course anomalies of coronary arteries.

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Fig 3: Images of separate origin of LAD coronary artery and CLX coronary artery without LM coronary artery.

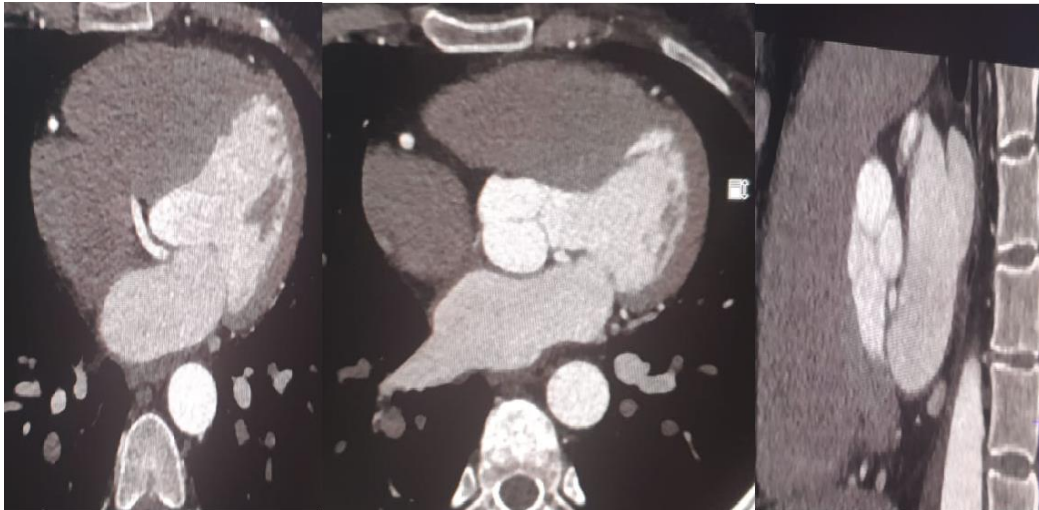


Fig 4: benign course of the CLX coronary artery from the right coronary sinus

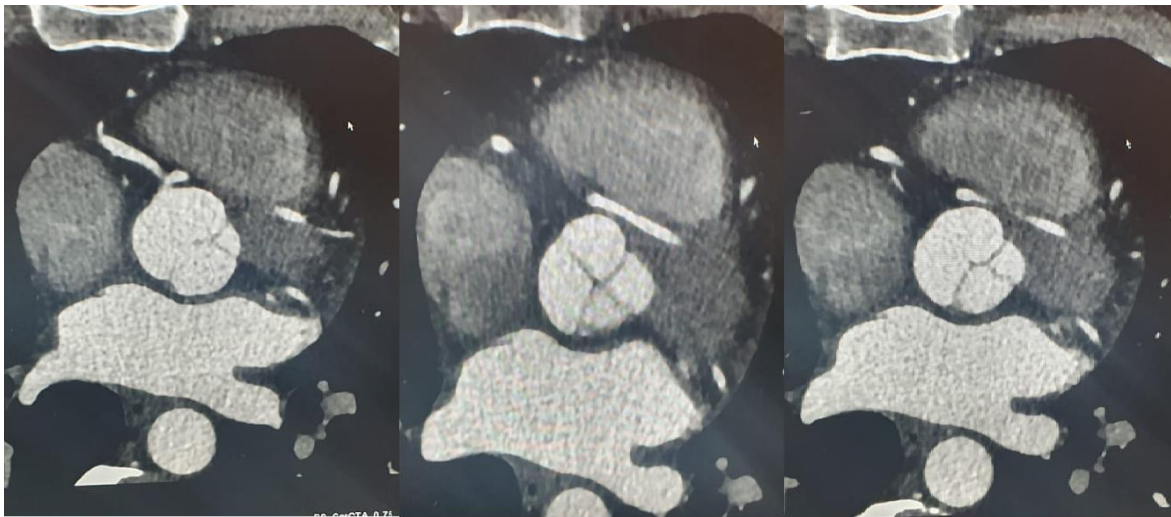


Fig 5: anomalous course of LAD coronary artery from the RCA with septal course.

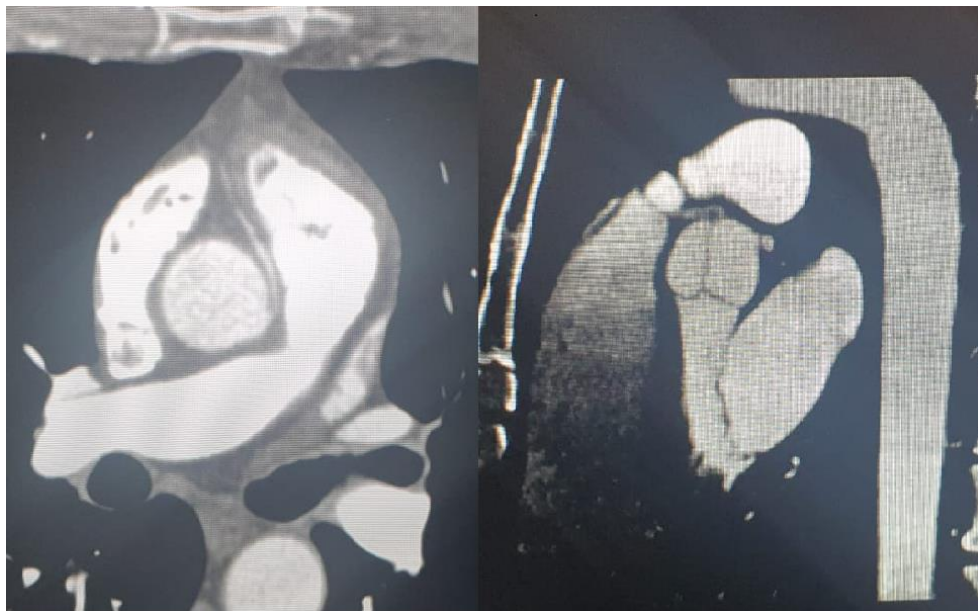


Fig 6: anomalous course of the RCA from the left coronary sinus with malignant course.



Fig 7: superficial bridging of LAD coronary artery

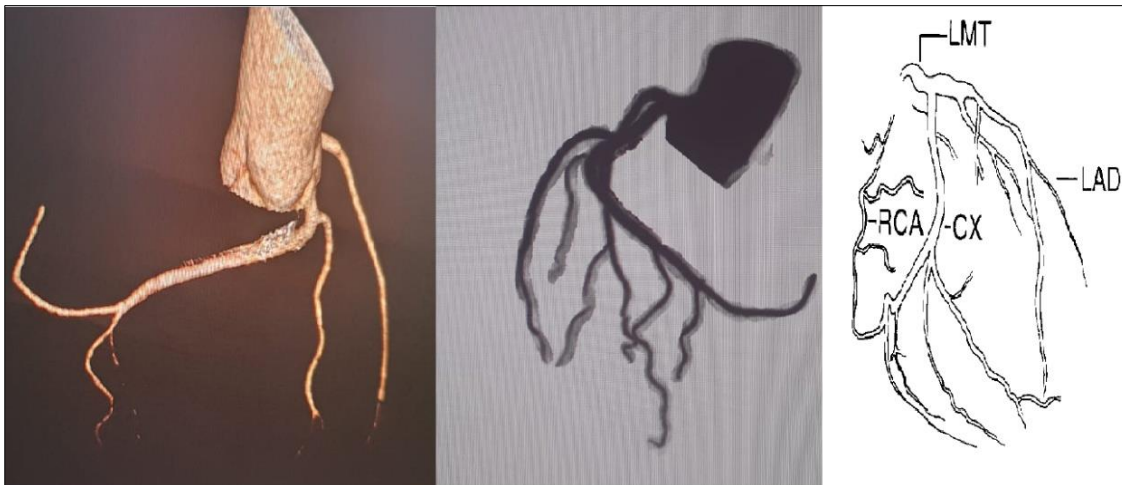


Fig 8: absent RCA with super dominant CLX coronary artery

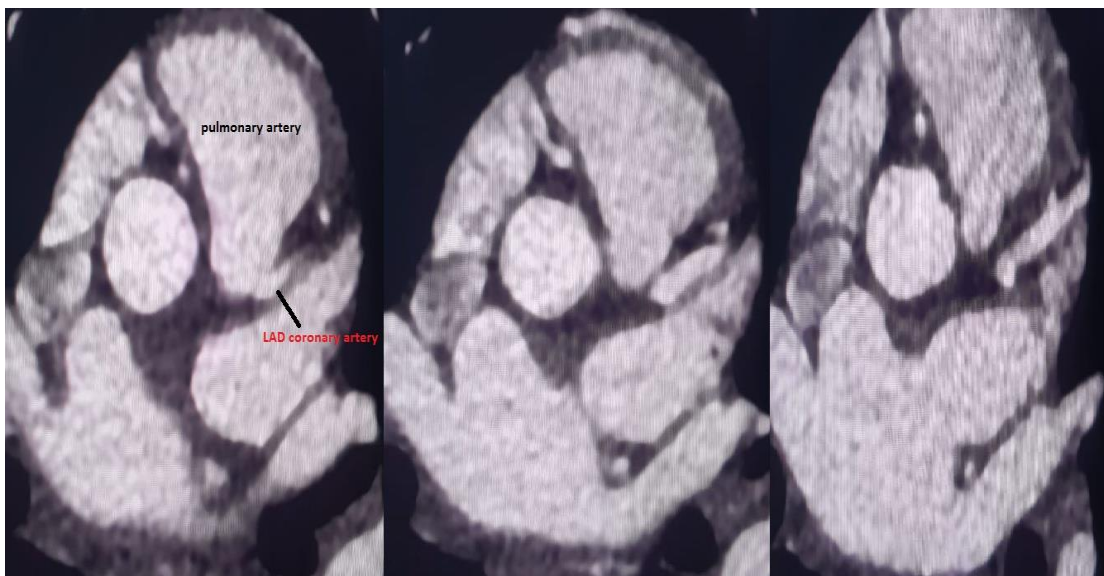


Fig 9: LAD coronary artery anomalies origin from the pulmonary artery ALCAPA

Conclusion

Our study revealed that the most common congenital coronary anomalies are anomalies of origin in which the most common is the RCA arising from the contralateral coronary or cusp. The most common anomalies of course is the benign retroaortic course.

The incidence of congenital coronary arteries is more common in males than in females

Detection of congenital coronary arteries anomalies is vital prior to percutaneous catheterization or cardiac surgery intervention.

Conflict of interest

None.

Funding source

None.

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