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CLINICAL & BASIC RESEARCH

Prevalence and Spectrum of Coronary Anomalies Detected on Coronary Computed Tomography Angiography

A single centre experience in Oman

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معدل انتشار وأنواع الشذوذ الخلقية للشرايين التاجية المكتشفة باستخدام التصوير المقطعي المحوسب للاوعية الدموية

تجربة مركز واحد في سلطنة عمان

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abstract: ***Objectives:*** Coronary artery anomalies (CAAs) are uncommon congenital abnormalities with a prevalence ranging from 0.2–2%. CAAs can be asymptomatic or less commonly present with life-threatening symptoms. This study aimed to investigate the prevalence and spectrum of CAAs in patients who underwent coronary computed tomography angiography (CCTA) in Oman. ***Methods:*** This retrospective study was conducted at the National Heart Centre, Muscat, Oman between September 2012 and August 2018. All consecutive patients who had undergone CCTA were included. ***Results:*** A total of 4,445 patients were included in this study. Of these, 59 patients (1.3%) were diagnosed with CAAs with a mean age of 52.6 years (range: 12–80 years) and an equal gender distribution. Among the patients with CAAs, the majority (69.5%) had anomalous origins from the opposite or non-coronary sinus. Right coronary artery arising from the left coronary sinus was the most common type (33.9%). Fewer patients (18.6%) had left circumflex arising from the right coronary sinus (RCS). Seven patients (11.9%) had left main arising from the RCS. Other CAAs were in the dual left anterior descending artery (8.5%), high coronary artery take-off (6.8%), single coronary *ostia* (6.8%) and coronary artery *fistula* (6.8%). ***Conclusion:*** The prevalence of CAAs was 1.3% which is similar to the literature.

Keywords: Coronary Vessel Anomalies; Computed Tomography Angiography; Prevalence; Oman.

الملخص: الهدف: الشذوذ الخلقية للشرايين التاجية هي تشوهات خلقية نادرة ويتراوح معدل انتشارها من 0.2-2%. في كثير من الأحيان تكون الشذوذ الخلقية للشرايين التاجية بدون اعراض ولكن في حالات قليلة تكون الاعراض مهددة للحياة بالخطر. هدفت هذه الدراسة الى معرفة معدل انتشار وأنواع الشذوذ الخلقية للشرايين التاجية عند المرضى الذين خضعوا للتصوير المقطعي المحوسب للشرايين التاجية في سلطنة عمان. **الطريقة:** اجريت هذه الدراسة الاستعادية في المركز الوطني لطب وجراحة القلب، مسقط، سلطنة عمان. وشملت هذه الدراسة جميع المرضى الذين خضعوا للتصوير المقطعي المحوسب للشرايين التاجية ما بين شهر سبتمبر لعام 2012 وشهر أغسطس لعام 2018. **النتائج:** تضمنت هذه الدراسة 4,445 مريضا ومن بين هؤلاء وجد ان 59 مريضا لديهم شذوذ خلقية في الشرايين التاجية (1.3%) وكان متوسط اعمارهم 52.6 سنة (المدى: 12-80 عاما) وبتوزيع متساو بين كلا الجنسين. وكانت الغالبية العظمى من مرضى الشذوذ الخلقية للشرايين التاجية لديهم منشأ شاذ من الجيب التاجي المقابل اوالجيب غير التاجي (69.5%). وقد مثل منشأ الشريان التاجي الأيمن من الجيب التاجي الايسر (33.9%). وجد عند عدد قليل من المرضى ان الشريان التاجي المنعطف الايسر ينشا من الجيب التاجي الأيمن (6.81%). ووجد عند سبعة من المرضى (11.9%) ان الشريان التاجي الايسر لديهم ينشأ من الجيب التاجي الأيمن. توزعت بقية حالات الشذوذ الخلقية للشرايين التاجية بين ازدواجية الشريان النازل الامامي (8.5%) وارتفاع فتحة الشريان التاجي (6.8%) ووجود فوهة تاجية وحيدة (6.8%) وناسور الشريان التاجي (6.8%). **الخلاصة:** معدل انتشارالشذوذ الخلقية للشرايين التاجية هو 1.3% وهو مماثل لما هو موجود في الادبيات الطبية.

الكلمات المفتاحية: تشوهات الأوعية التاجية؛ التصوير المقطعي المحوسب للاوعية الدموية؛ معدل انتشار؛ عمان.

Advances in Knowledge

- *The findings of this study demonstrate that there is no significant difference in the prevalence and spectrum of coronary artery anomalies (CAAs) between this study's population and the rest of the world.*

Application to Patient Care

- *This study highlights the importance of utilising coronary computed tomography angiography as a robust non-invasive test for evaluating CAAs.*

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Coronary artery anomalies (CAAs) are uncommon congenital abnormalities with a prevalence of 0.2–2% in the general population.[1–5] CAAs encompass a spectrum of anomalies including abnormalities of origin, course, number, structure and termination of the coronary arteries.[6] Most patients with CAAs are asymptomatic; however, some CAAs have been linked to ischaemic heart disease, syncope and sudden cardiac death.[7–9] CAAs are generally detected incidentally on autopsy, conventional coronary angiography (CCA) or through coronary computed tomography angiography (CCTA). Although CCA is the gold standard for assessing contrary arteries, it is an invasive procedure and has limited ability to demonstrate the anatomy of complex CAAs. In contrast, CCTA is non-invasive and can show complex anatomy.[1,2,10] The National Heart Centre, Muscat, Oman, is a tertiary cardiac centre which receives referrals from all of Oman. This study aimed to assess the prevalence and spectrum of CAAs in Oman's general population.

Methods

A total of 4,445 consecutive patients who underwent CCTA at the National Heart Centre, from September 2012 to August 2018 were included. The indications for performing CCTA were chest pain to exclude coronary artery disease (CAD), assessment of coronary artery grafts or stents, cardiomyopathy, evaluation for congenital heart disease, evaluation of syncope and pre-cardiac surgery assessment. Patients were excluding from CCTA if they had a history of allergy to iodinated contrast material, impaired renal function defined as an estimated glomerular filtration rate of <45 mL/min/1.73m^2, an inability to follow breathing instructions, an uncontrolled heart rate, cardiac arrhythmia and severe coronary calcification on calcium score scan. Patients with myocardial bridging (MB) were excluded as some authors considered MB to be a normal variant, whereas others considered it to be an anomaly. In addition, there is a lack of clarity regarding its definition in the literature.[11–13]

Prior to the CCTA, participants received an electrocardiogram (ECG) and heart rate and blood pressure were checked. Procedure preparation included the oral administration of 25–100 mg of atenolol with a baseline heart rate >70 beats/min in order to lower the heart rate. Sublingual nitroglycerine (0.8 mg) was administered to all patients one minute prior to injecting 60–75 mL of contrast followed by 30 mL of saline solution at a rate of 6 mL/second, unless it was contraindicated. The CCTA was performed using a dual source 256 slice (2 × 128) scanner (SOMATOM Definition Flash, Siemens AG), a rotation time of 280 minutes and dual source 384 (2 × 192; SOMATOM Force, Siemens AG, Berlin and Munich, Germany) with a rotation time of 250 minutes. All scans started with a topogram followed by a prospective ECG-gated non-enhanced computed tomography (CT) scan at a 75% R-R interval. Subsequently, a contrast-enhanced scan was performed where an ECG-gated prospective or retrospective scan with a slice thickness of 0.6 mm was acquired during breath holding after inspiration. The scan parameters were adjusted automatically or manually to acquire the best quality image with the lowest radiation dose.

Data from the scanners were sent to separate image processing work stations (Syngo.via, Siemens AG) and analysed by three cardiothoracic radiologists with three, six and nine years of experience. Coronary arteries were assessed for atherosclerotic disease, anomalous origins, courses and terminations. Patients with CAAs were selected and images were again reviewed by a single radiologist before inclusion in the study.

The Scientific Research Committee of the Royal Hospital, Muscat, Oman approved this study (SRC#53/2018) and waived informed consent.

Results

In the current study, the prevalence rate of patients with CAAs was 1.3% [Table 1]. The baseline clinical characteristics of these patients are shown in Table 2.

Anomalous origin of the coronary artery from the opposite or non-coronary sinus (ACAOS) was the most common CAA. Right coronary artery (RCA) arising from the left coronary sinus (LCS) was the most common anomaly (33.9%) [Figures 1A and B]. The RCA arose from the non-coronary sinus (NCS) in two patients (3.4%) [Figures 1C and D]. In all patients, the anomalous RCA arose from the LCS and then passed between the aortic root and the pulmonary artery taking an interarterial course. One patient underwent deroofing surgery and another patient refused to undergo surgery. The second most common type (18.6%) of ACAOS was a left circumflex artery (LCX) originating from the right coronary sinus (RCS) or RCA with a retro-aortic course.

Five patients (8.5%) had anomalous origin of the left main coronary artery (LMCA) from the RCS and two patients had an interarterial course [Figures 2A and B]. One patient underwent a coronary artery bypass surgery and one patient refused surgery. Three patients with anomalous LMCA from the RCS had a transseptal course [Figures 2C and D].

Table 1: Prevalence and spectrum of coronary artery anomalies in Omani patients undergoing coronary computed tomography angiography (N = 59)

Coronary artery anomaly	n (%)	Percentage among total sample (n = 4,445)
Anomalous origin from opposite or NCS with anomalous course		
RCA arising from the LCS with interarterial course	20 (33.9)	0.450
LCX arising from the RCS or RCA with retroaortic course	11 (18.6)	0.250
LMCA arising from the RCS with interarterial course	2 (3.4)	0.045
LMCA arising from the RCS with transseptal course	3 (5.1)	0.076
LMCA arising from the RCS with retroaortic course	2 (3.4)	0.045
RCA arising from the NCS	2 (3.4)	0.045
LMCA from the NCS	1 (1.7)	0.022
Multiple *ostia*		
Absent left main trunk with LAD and LCX originating separately from the aorta	1 (1.7)	0.022
Single coronary *ostium*	4 (6.8)	0.089
High take-off		
LMCA	1 (1.7)	0.045
RCA	3 (5.1)	0.076
Dual LAD anomaly	5 (8.5)	0.110
CAF	4 (6.8)	0.089

NCS = non-coronary sinus; RCA = right coronary artery; LCS = left coronary sinus; LCX = left circumflex artery; RCS = right coronary sinus; LMCA = left main coronary artery; LAD = left anterior descending artery; CAF = coronary artery fistula.

Table 2: Baseline characteristics of Omani patients with coronary artery anomalies who underwent coronary computed tomography angiography (N = 59)

Characteristic	n (%)
Mean age in years (range)	52.6 (12–80)
Male:female ratio	30:29
Presenting symptoms	
Typical chest pain	13 (22)
Atypical chest pain	28 (47.5)
Syncope	2 (3.4)
Dyspnoea	12 (20.3)
Palpitation	9 (15.3)
Risk factors	
Diabetes	18 (30.5)
Hypertension	24 (40.7)
Dyslipidaemia	7 (11.9)
Underlying cardiac problem	
Dilated cardiomyopathy	5 (8.5)
Previous myocardial infarction	1 (1.7)

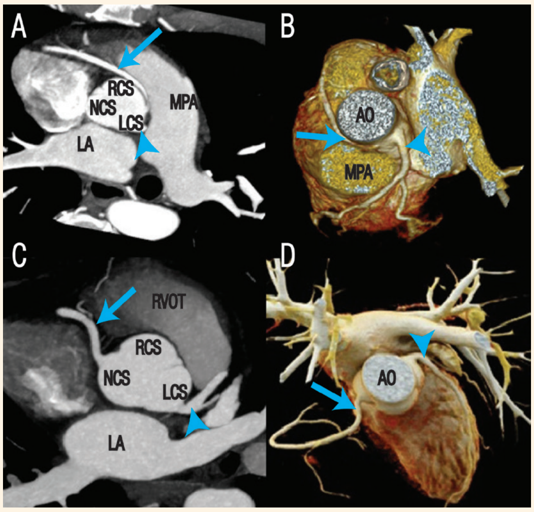

Figure 1: Axial maximal intensity projection via coronary computed tomography angiography imaging (**A & C**) and three-dimensional volume rendering (**B & D**) showing (**A & B**) the right coronary artery (arrow) arising from the left coronary sinus before it courses to the right between the main pulmonary artery and the aorta and showing (**C & D**) the right coronary artery (arrowhead) arising from the non-coronary sinus.

RCS = right coronary sinus; NCS = non-coronary sinus; MPA = main pulmonary artery; LCS = left coronary sinus; LA = left atrium; AO = aorta; RVOT = right ventricular outflow tract.

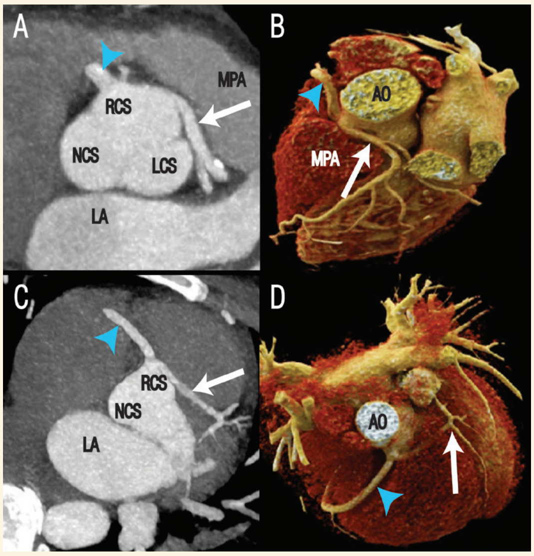

Figure 2: Axial maximal intensity projection via coronary computed tomography angiography imaging (**A & C**) and three-dimensional volume rendering (**B & D**) showing (**A & B**) the left main coronary artery (arrow) arising from the right coronary sinus before it courses to the left between the main pulmonary artery and the aorta and showing (**C & D**) the left main coronary artery (arrow) arising from the right coronary sinus before it courses to the left through the interventricular *septum*. The right coronary artery is indicated by the arrowhead.

RCS = right coronary sinus; MPA = main pulmonary artery; NCS = non-coronary sinus; LCS = left coronary sinus; LA = left atrium; AO = aorta.

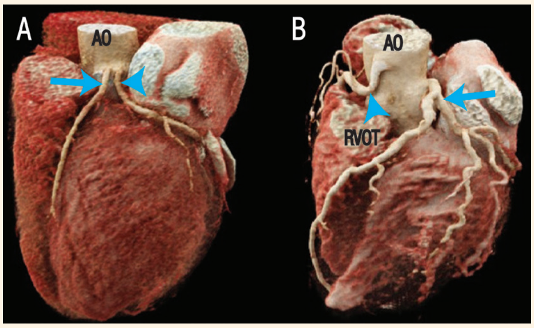

Figure 3: Three-dimensional volume-rendered coronary image showing (**A**) the separate origin of the left anterior descending artery (arrow) and the left circumflex artery (arrowhead) from the left coronary sinus and (**B**) the anomalous origin from the right coronary artery (arrowhead) from the aorta above the sinotubular junction.
AO = aorta; RVOT = right ventricular outflow tract.

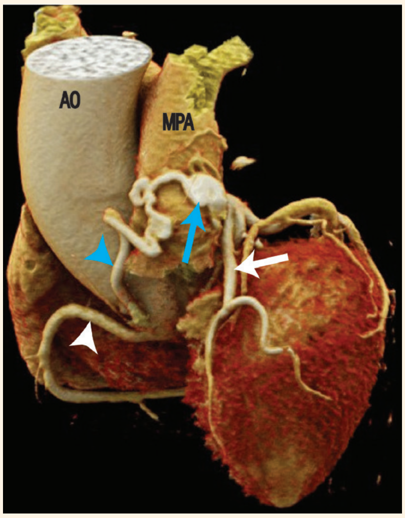

Figure 4: Three-dimensional volume rendered coronary computed tomography angiography image showing a tortuous vessel (blue arrowhead) arising from the right coronary artery (white arrowhead). The tortuous vessel passes anterior to the main pulmonary artery and forms a small aneurysm (blue arrow) before it enters the main pulmonary artery distal to the valve. The left anterior descending artery is marked by the white arrow.
AO = aorta; MPA = main pulmonary artery.

LMCA was absent in one patient (1.7%) with the left anterior descending (LAD) coronary artery and LCX arising separately from the LCS [Figure 3A]. Three patients (5.1%) had high take-off RCA [Figure 3B]. Four patients (6.8%) had coronary artery *fistulas* (CAFs) in the forms of a *fistula* connecting the RCA to the main pulmonary artery (MPA) [Figure 4], a *fistula* between the left anterior descending artery (LAD) or the MPA, which is a complex *fistula* that connects the RCA, the LAD and a bronchial artery. One patient had a complex *fistula* between the RCA, LMCA, LAD and MPA.

Discussion

CAAs are part of a spectrum of uncommon coronary artery congenital anomalies.[13] In the literature, discrepancies in the prevalence of CAAs are largely due to the inclusion or exclusion of MB due to the fact that some authors consider MB as a normal variant, whereas others consider it an anomaly.[12,14,15] When MB is excluded, the prevalence of CAAs ranges between 0.5–3.1%; however, when MB is included, prevalence increases to 7.9–18.4%.[3] In the current study, the prevalence of CAAs, excluding MB, was 1.3% which falls within the reported range in the literature.[3,5,11,16] To the best of the authors' knowledge, this study is the first to estimate the prevalence and spectrum of CAAs in the general Omani population.

CCA can be used to detect CAAs, although it is an invasive procedure and has a risk of complications.[17] However, CCTA is a non-invasive test that has recently become the gold standard for detecting and characterising CAAs. CCTA can precisely delineate the course of the anomalous artery and provide three-dimensional information about the relation of anomalous to other cardiovascular structures, namely cardiac chambers and major arteries.[18–20]

CAAs can be categorised based on the origin, course, number and termination of the coronary arteries.[6] In the current study, ACAOS was the most common CAA (69.5%) which was similar to reports from previous studies.[7,11,20,21] ACAOS can be classified into four groups: 1) RCA arising from the LCS, 2) LCA arising from the RCS, 3) LCX or LAD arising from the RCS and 4) LCA or RCA arising from the NCS.[6,22] The course of the anomalous artery can be interarterial between the aorta and the MPA, anterior to the MPA, retroaortic or transseptal through the interventricular *septum*.[5,9,13,16] Of these courses, the most clinically significant anomaly is the interarterial course because it is strongly associated with sudden death, particularly in young competitive athletes.[23] Although not fully understood, the assumed mechanism of ischaemia in an anomalous artery with an interarterial course is that the anomalous artery is squeezed between the aorta and the MPA.[11] Therefore, patients with an interarterial course should be treated; however, many patients with these anomalies are asymptomatic. Moreover, there are difficulties implicit in routine examinations and clinical testing for these anomalies. In such cases CCTA is an important diagnostic tool as it can differentiate between interarterial and transseptal courses with high spatial and temporal resolution.[9,16] However, a disadvantage of using CCTA is the radiation that accompanies it; however, the new generation of multidetector scanners has much lower radiation compared to older generations.[24]

There are three different treatment options for patients with interarterial anomalous arteries: 1) medical treatment, 2) angioplasty and 3) surgical repair.[25] In the current study, all 20 patients with anomalous RCA from the LCS had an interarterial course.

Multiple coronary artery *ostia* is either due to the separate origins of the RCA and *conus* branch from the aorta or the absent LMCA with the LCX and LAD arising separately from the aorta. The prevalence of the absence of LMCA ranges from 0.5–8%.[5] In this study's sample, the prevalence was lower (0.03%) and only one patient had no LMCA and separate *ostia* of the LAD and LCX. Multiple *ostia* allow alternative collateral supplies in patients with proximal CAD; however, these can cause technical difficulties in cannulation during invasive coronary angiography.[11,13,26]

A single coronary *ostium* is an extremely rare congenital anomaly characterised by a single artery arising with a single *ostium* from the aorta. It is seen in only 0.0024–0.044% of the population.[6] Patients with a single coronary artery are at increased risk for sudden death if a major coronary branch has an interarterial course between the pulmonary artery and the aorta.[27] Four of the current patients (6.8%) had single coronary *ostia* which originated from the RCS.

A high take-off CAA is a coronary artery which arises from the aortic wall 0.5 cm above the sinotubular junction.[6] Although this anomaly is benign, it can cause technical difficulties of coronary artery cannulation during conventional angiography. Moreover, cardiac surgeons should be aware of this anomaly prior to cardiac surgery to avoid damaging the anomalous artery while cross-clamping the aorta.[5,16] In the current study, four patients (6.8%) had high take-off coronary arteries, one patient (1.7%) had high take-off of the LMCA and three (5.1%) had high take-off RCA.

Dual LAD is a rare congenital anomaly which is defined by the presence of short and long LADs which supply the course of the LAD.[28] It is traditionally classified into four types based on the origin, course and termination of the short and long LADs; however, CCA has led to recognition of other variants. To date, eleven types of dual LAD variants have been identified.[29] Awareness of dual LAD variants is crucial for interpreting cardiac CTs in patients with dual LAD anomalies and for planning percutaneous and surgical reperfusion strategies. Five of the patients (8.5%) in the current study had dual LADs.

CAFs are defined by abnormal communication between a coronary artery and another structure such as a heart chamber, the MPA or the coronary sinus.[16,30] Small CAFs are usually asymptomatic and detected incidentally on echocardiography, coronary arteriography or CCTA performed for other reasons. However, large CAFs can lead to the 'steal phenomenon' which can lead to ischaemia due to diminished blood supply to the portion of the myocardium supplied by the involved coronary artery. CAFs are seen in 0.05–0.25% of patients undergoing conventional angiography.[30] In this study, the prevalence of CAFs was 0.089%.

This study had some limitations. This was a single centre experience and some important clinical characteristics of the patients included in this study were not documented due to its retrospective nature. In addition, minor CAAs, including coronary artery ectasia, aneurysm and variant origins of the *conus* artery were excluded. MB was also excluded due to uncertainty about diagnostic and classification criteria.

Conclusion

The prevalence of CAAs in Oman was similar to reported figures from earlier studies performed worldwide. In Oman, anomalous origin of the RCA arising from the LCS was the most common type.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

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