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MDCT Evaluation of Congenital Coronary Anomalies: Pictorial Essay

Pictorial Essay

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Abstract

Objective: The purpose of this pictorial essay is to review the multi detector computed tomography (MDCT) coronary angiography appearance of congenital coronary anomalies [CCA]. CCA might also be classified as hemodynamically significant or insignificant. The clinical symptoms may include chest pain, dyspnoea, palpitations, syncope, cardiomyopathy, arrhythmia, infarction and sudden cardiac death. Although CCA are relatively uncommon, they're the second most typical reason for sudden cardiac death among young athletes and so warrant detailed review. Familiarity with atypical anatomy and their clinical presentation may facilitate appropriate diagnosis and management. This will be of immense help to the clinician planning interventional procedures like stenting, balloon dilatation, or graft surgery particularly when there are secondary changes of calcification, plaque formation and stenosis.

Conclusion; Increasing use of MDCT for cardiac imaging has helped within the detection of the many benign congenital coronary anomalies (CCA), but a little number is related to myocardial ischemia and sudden death. Increasing the employment of MDCT in cardiac imaging may yield diagnostic information on CCA not obtained with invasive coronary angiography. Axial sections, multiplanar reconstructions, virtual angioscopy, and 3D volume-rendered images should aid within the detection and improve the interpretation of such anomalies, which might be of immense help to the clinician planning interventional procedures.

Keywords: Computed tomography; Coronary angiography; Congenital coronary anomalies; Malignant coronary artery

Introduction

Congenital coronary anomalies (CCA) are uncommon and most of them are diagnosed incidentally during conventional coronary angiography or MDCT angiography. In step with the literature, CCAs affect around 1% of the general population, starting from 0.3%-5.6% in studies on patients undergoing coronary angiography, and in approximately 1% of routine autopsy [1]. Based on the functional relevance of every abnormality, coronary artery anomalies may be classified as anomalies with obligatory ischemia, without ischemia or with exceptional ischemia. The clinical symptoms may include chest pain, dyspnea, palpitations, syncope, cardiomyopathy, arrhythmia, myocardial infarct and sudden cardiac death. Although congenital coronary artery anomalies are relatively uncommon, they're the second most typical reason behind sudden cardiac death (SCD) among young athletes. The chance of SCD in time of life or elderly individual with an incidentally discovered coronary anomaly is unclear, but is perhaps negligible. The anomaly most often related to SCD is that the anomalous origin of a coronary artery, particularly with a course between the aorta and the PA [1]. For several decades, premorbid diagnosis of CCA has been made with conventional angiography. Although catheter angiography is an efficient tool, it's invasive and related to procedural morbidity (1.5%) and mortality (0.15%) [2]. Because of its two dimensional nature, catheter angiography has projectional limitations and it cannot show the link of aberrant vessels with the underlying cardiac structures [3]. Recent development of ECG gated MDCT coronary angiography allows accurate and noninvasive depiction of coronary artery anomalies of

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origin, course, and termination. CT coronary angiography (CTCA) is superior to standard catheter angiography in delineating the ostial origin and proximal path of an anomalous coronary artery [4].

Illustrative Cases

Case 1: 56 years old male with dominant Right coronary arterial (RCA) system (Figure 1).

Case 2: 49year's old male with dominant Left coronary arterial (LCA) system (Figure 2).

Case 3: 36 year old male with Co-dominant coronary arterial system (Figure 3).

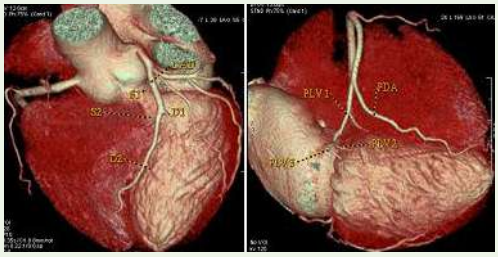

Figure 1: (A,B) Computed tomography coronary angiography (CTCA) 3D volume rendered images shows dominant right coronary arterial system, posterior descending artery (PDA) and posterior left ventricular (PLV) branches arising from RCA.

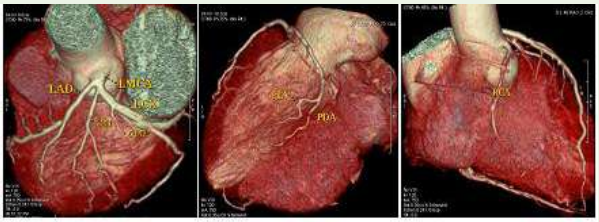

Figure 2: (A-C) CTCA 3D volume rendered images shows dominant left coronary arterial system, posterior descending artery (PDA) and posterior left ventricular (PLV) branches arising from LCA.

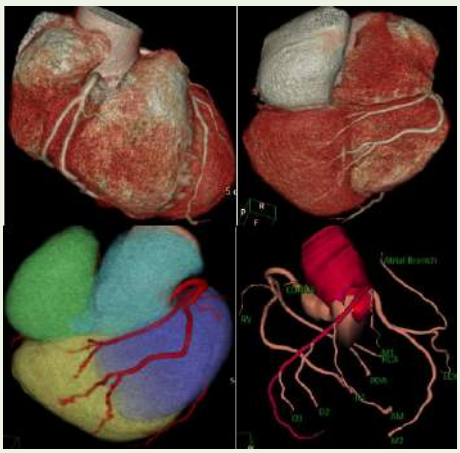

Figure 3: (A-D) CTCA 3D volume rendered (A-C) and coronary tree images show co-dominant coronary arterial system, PDA and PLV branches arising from both LCA & RCA.

Case 4: 38 year old male with trifurcation of LMCA into LAD, LCX & Ramus intermedius (Figure 4).

Anomalies of origin; A. Number of Ostia: single, Multiple (>2)

Case 5: Single Left coronary artery in a 39 year-old man (Figure 5).

Case 6: Single Right coronary artery in a 46 year old man (Figure 6).

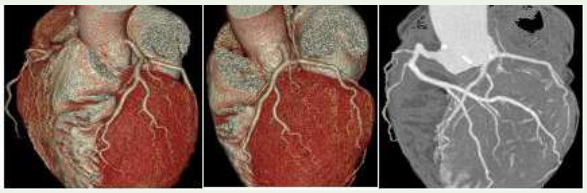

Figure 4: (A-C) CTCA 3D volume rendered (A,B) and MIP coronary tree (C) images shows trifurcation of LMCA into LAD, LCX & Ramus Intermedius (RI).

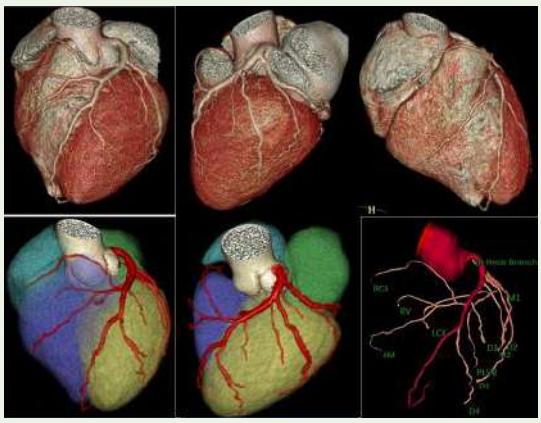

Figure 5: (A-F) Computed tomography coronary angiography (CTCA) 3D volume rendered (A-E) and coronary tree (F) views shows single left coronary artery arising from left coronary sinus (LCS) with RCA arising as a large branching septal branch from the mid LAD artery and supplying RCA territory.

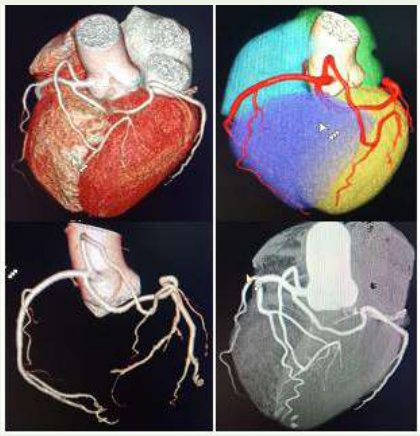

Figure 6: (A-D) CTCA 3D volume rendered (A,B), coronary tree (C), MIP Coronary tree (D) views shows single right coronary artery arising from right coronary sinus (RCS) with large conus branch and large right ventricular (RV) branch from RCA supplying LAD and LCX territory respectively.

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Case 7: Absent left main coronary artery (LMCA) in a 40year-old woman with normal origin of RCA (Figure 7).

B. Anomalous location of ostium in the appropriate coronary sinus

Case 8: 40 year old male with high origin of LMCA from Left coronary sinus (Figure 8).

C. Origin from opposite coronary sinus

LCX or LAD artery arising from the right coronary sinus (RCS)

Case 9: 70 year-old women with absent LMCA and anomalous origin of LAD & LCX arteries from RCS (Figure 9).

Case 10: 40 years old man with anomalous origin of LCX from RCS (Figure 10).

Case 11: 58 years old man with anomalous origin of LCX from RCS (Figure 11).

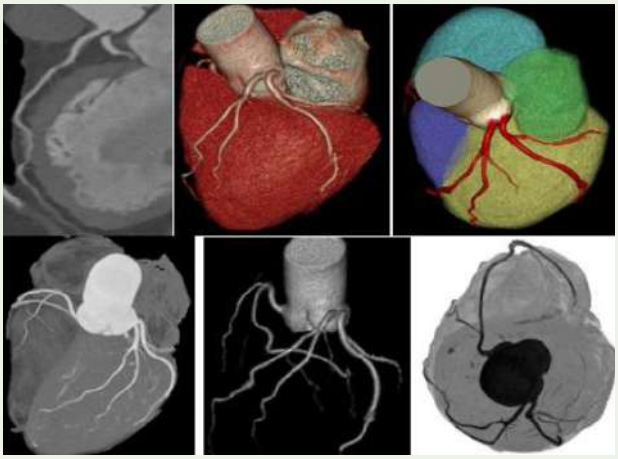

Figure 7: (A-F) CTCA 2D curved view(A), 3D volume rendered images (B-D), coronary tree (E) and positive coronary tree (F) images reveal absent LMCA with separate ostia of the LAD and LCX artery from LCS.

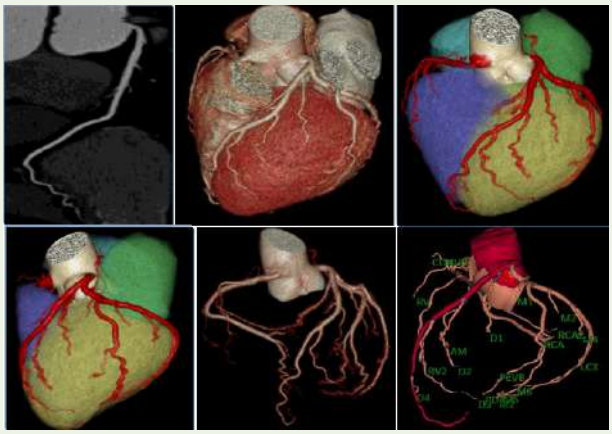

Figure 8: (A-F) CTCA 2D curved view (A), 3D volume rendered (B-D) and coronary tree (E,F) images reveal anomalous high origin of LMCA from LCS and anomalous low origin of RCA from aorta above the sino-tubular junction.

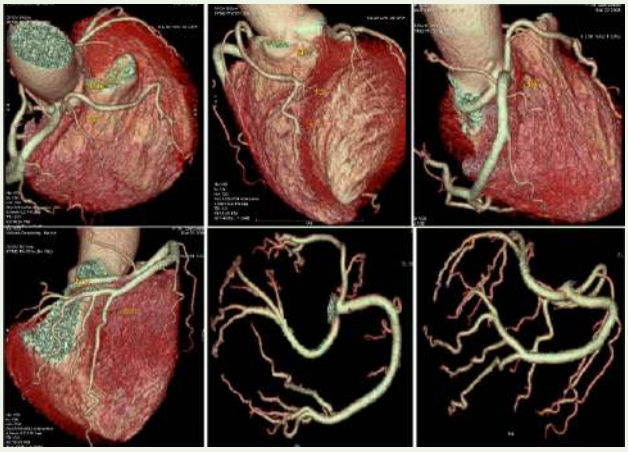

Figure 9: (A-F) CTCA 3D volume rendered images (A-D) and coronary tree view (E,F) images reveal absent LMCA with separate ostia of LAD & LCX from RCS, LCX having posterior (retro-aortic) course, and LAD having anterior (pre-pulmonic) course.

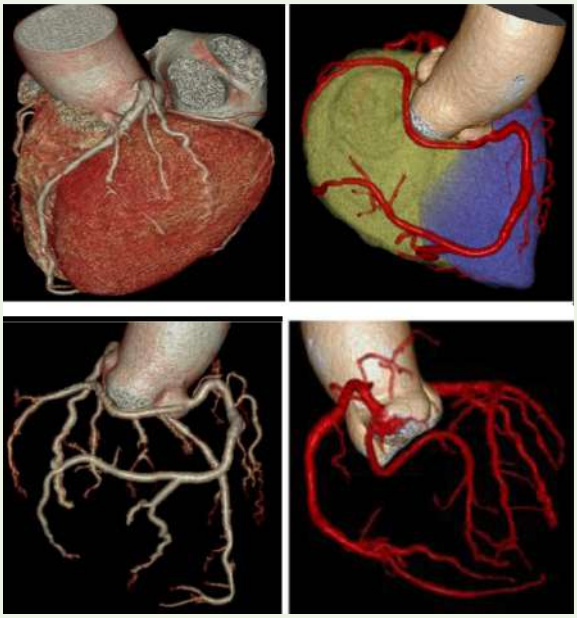

Figure 10: (A-D) CTCA 3D volume rendered (A,B) and coronary tree view (C,D) images reveal anomalous origin of LCX from RCS having retro-aortic course.

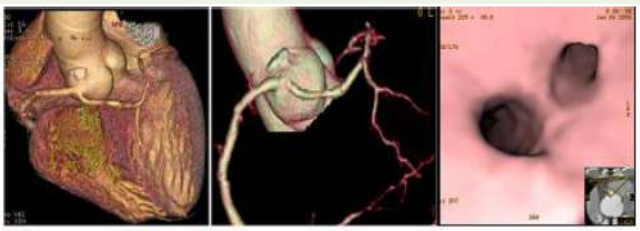

Figure 11: (A,B) CTCA 3D volume rendered (A), coronary tree (B) and virtual angioscopy images reveal anomalous origin of LCX from RCS having pre-pulmonic course.

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Case 12: 45 years old woman with anomalous origin of LAD branching off from proximal RCA (Figure 12).

RCA arising from the LCS

Case 13: 35 years old man with nonspecific chest pain on exertion (Figure 13).

Case 14: 35 years old man with recurrent chest pain on exertion (Figure 14).

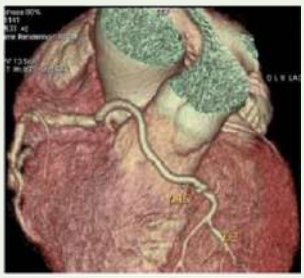

Figure 12: CTCA 3D volume rendered images reveal anomalous origin of LAD from proximal RCA having pre-pulmonic course

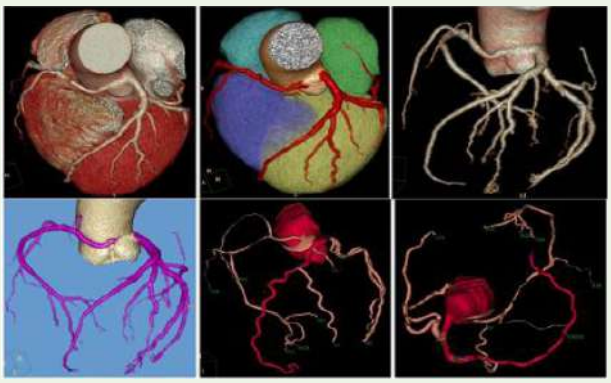

Figure 13: (A-F) CTCA 3D volume rendered (A,B) and coronary tree view (C-F) images reveal anomalous high origin of RCA from LCS having inter-arterial (malignant) course.

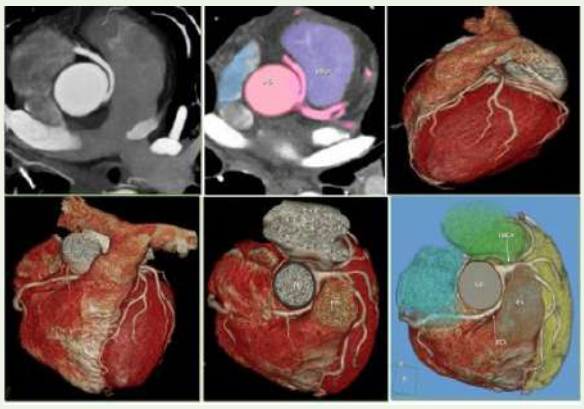

Figure 14: CTCA 2D axial (A,B) and 3D volume rendered images (C-F) images shows anomalous origin of RCA from LCS having inter-arterial (malignant) course.

Origin from Non-coronary sinus (NCS)-LCA or RCA (or branch of either artery)

Case 15: 63 years old woman with anomalous origin of RCA from NCS (Figure 15).

Abnormalities of angle of origin

Case 16: 48 year old male with recurrent chest pain on exertion (Figure 16).

Case 17: 45 year old woman with lowaortic origin of LMCA and high aortic of RCA (Figure 17).

Case 18: 65 year old female with recurrent chest pain on exertion (Figure 18).

Anomalies of course (normal origin); Myocardial bridging, Duplication

Case 19: 38 yerars old man with atypical chest pain on exertion (Figure 19).

Case 20: 49 years old man with atypical chest pain (Figure 20).

Case 21: 64 year old male with atypical chest pain (Figure 21).

Case 22: 45 year old male with duplication of LAD (Figure 22).

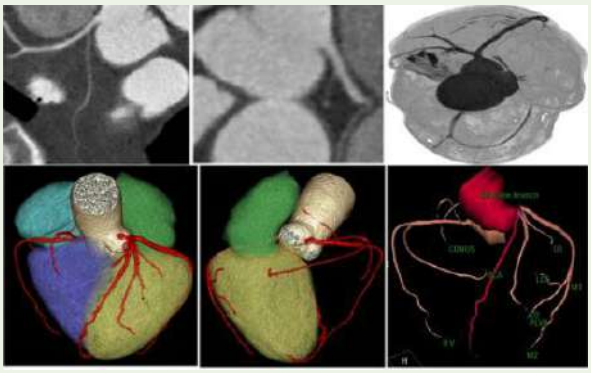

Figure 15: (A-F) CTCA 2D axial (A,B), Coronary tree positive image (C), 3D volume rendered (D,E) and coronary tree (F)images shows anomalous origin of RCA from NCS having retro-aortic course.

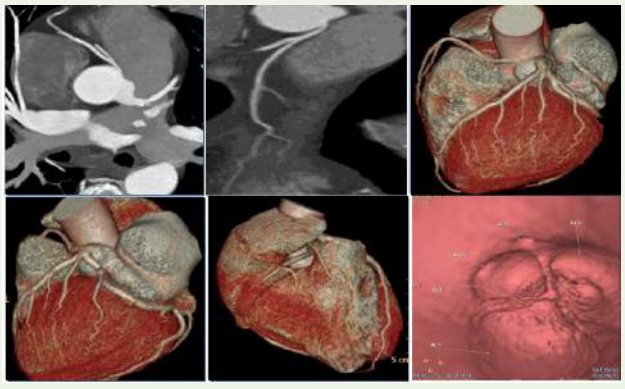

Figure 16: (A-F) CTCA 2D axial (A,B), 3D volume rendered (C-E) and virtual aortic angioscopy image (F), shows anomalous low aortic origin of RCA above the right sino-tubular junction with abnormal angulation and having inter-arterial (malignant) course.

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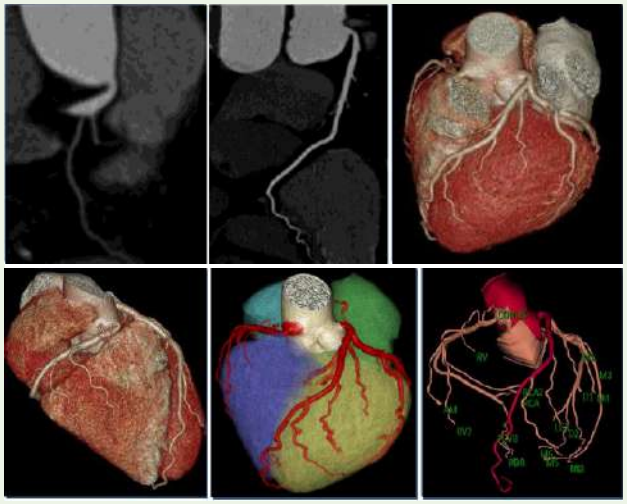

Figure 17: (A-F) CTCA 2D curved view (A,B), 3D volume rendered images (C-E) coronary tree (F) reveal anomalous low aortic originof LMCA & high aotic origin of RCA, above the sino-tubular junction with abnormal angulation.

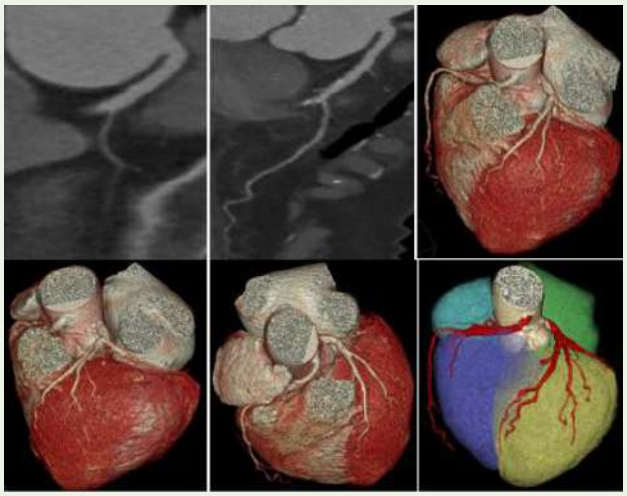

Figure 18: (A-F).CTCA 2D curved analysis (A,B) &3D volume rendered images (C-F) reveal anomalous high aortic originof LMCA & RCA, above the sino-tubular junction with abnormal angulation and interarterial (malignant) course of RCA.

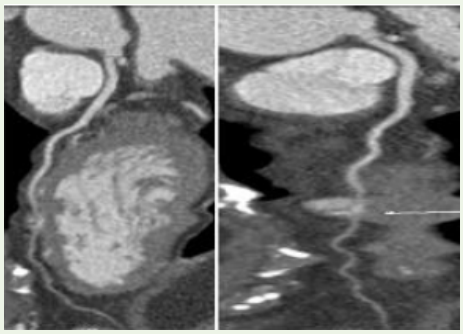

Figure 19: (A,B) CTCA 2D Curved analysis images reveal focal myocardial bridging of mid LAD.

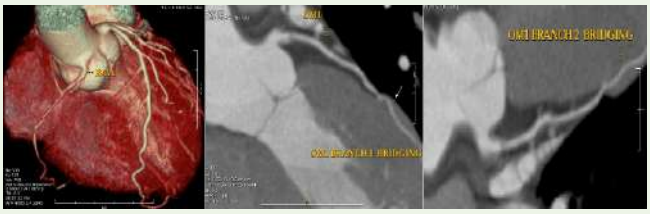

Figure 20: (A-C) CTCA 3D volume rendered (A) and 2D Curved analysis (B,C) images reveal focal myocardial bridging of first obtuse marginal (OM1) branches.

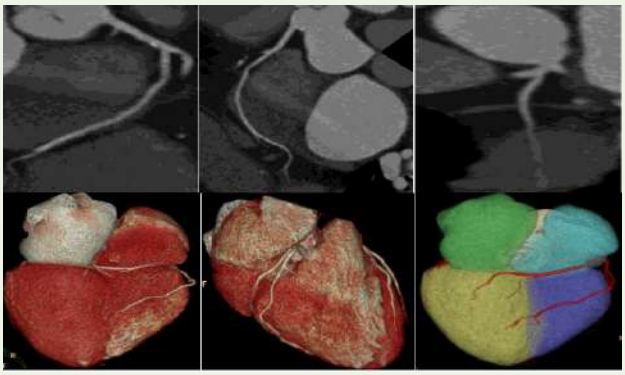

Figure 21: (A-F) CTCA 2D curved analysis (A-C) & 3D volume rendered images (D-F) reveal duplication of proximal RCAinto RCA1 and RCA2 with focal myocrdial bridging of mid RCA1.

Figure 22: (A-F) CTCA 3D volume rendered images (A-E) and coronary tree (F) reveal duplication of proximal LAD into LAD1 & LAD2 in the anterior interventricular groove.

Case 23: 48 year old female with RCA duplication (Figure 23).

Intrinsic coronary arterial abnormality; Coronary stenosis, Atresia, Ectasia / Aneurysm

Case 24: 59 year old man with unsuccesful attempts at catheterisation of LMCA,who developed arrythmias while trying to enter the ostia (Figure 24).

Case 25: 71 year old male with LAD & first obtuse marginal (OM1) ectasia (Figure 25).

Case 26: 45 old female with RCA ectasia (Figure 26).

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Case 27: 3 year old female child with Kawasaki disease (Figure 27).

Case 28: 35 year old male with LMCA bifurcation aneurysm (Figure 28).

Case 29: 65 year old male with LAD aneurysm (Figure 29).

Case 30: 65 year man with LAD & LCX aneurysm (Figure 30).

Case 31: 71year old manwith multiple atherosclerotic RCA aneurysm (Figure 31).

Case 32: 62 year old man with LAD hypoplasia (Figure 32).

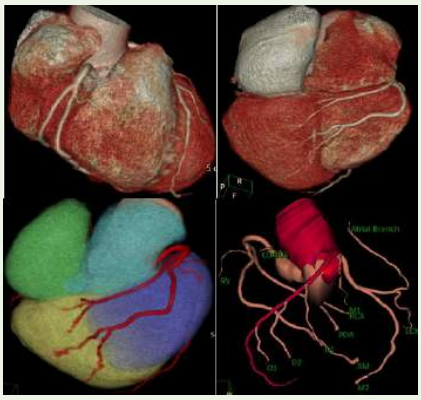

Figure 23: (A-D) CTCA 3D volume rendered images (A-C) and coronary tree view (D) reveal duplication of mid RCA into RCA1&RCA2.

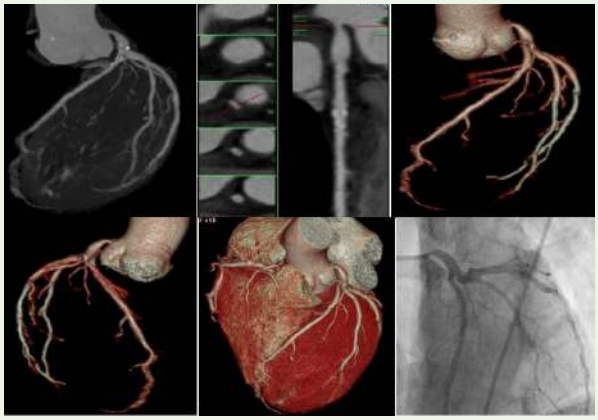

Figure 24: (A-F) CTCA MIP coronary tree (A), Straight vessel analysis (B) and 3D volume rendered coronary tree (C,D), cardia (E) and conventional cath angio (F) images show Ostio-proximal LMCA stenosis.

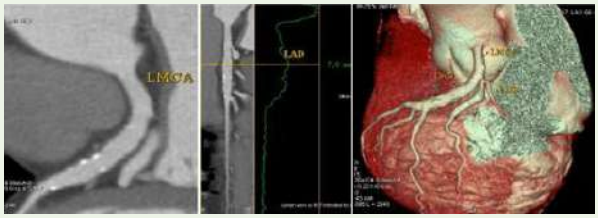

Figure 25: (A-C) CTCA 2D curved (A) & straight vessel analysis (B) views and 3D volume rendered images (C) and coronary tree view (C) reveal ectasia of proximal LAD & OM1 of LCX.

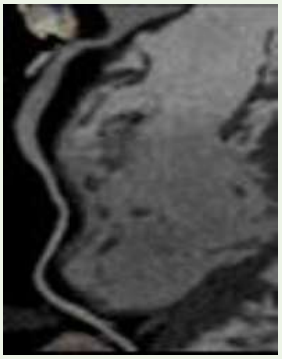

Figure 26: CTCA 2D Curved vessel analysis show ectasia of proximal RCA.

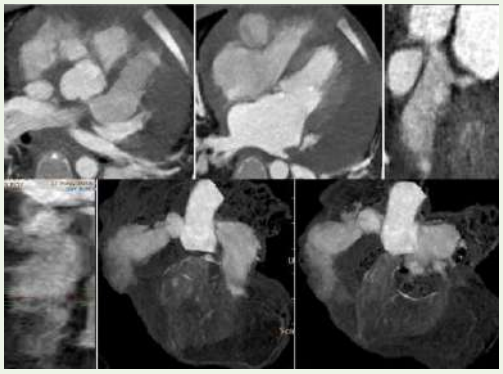

Figure 27: (A-F) CTCA 2D axial (A,B), Straight vessel analysis (C,D) and 3D volume rendered coronary tree images(E,F) show diffuse Ectasia of LMCA, LAD & RCA.

Figure 28: (A-C) 2D axial (A), 3D volume rendered (B) and coronary tree view(C) images show LMCA bifurcation saccular aneurysm.

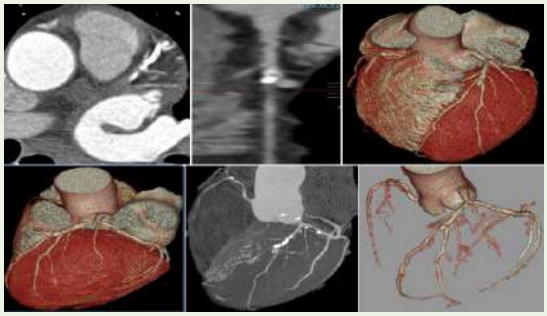

Figure 29: (A-F) CTCA 2D axial (A), Straight vessel analysis(B),3D volume rendered (C-E) and coronary tree (F) images show diagonal 3 (D3) bifurcation saccular aneurysm with coronary atherosclerotic disease.

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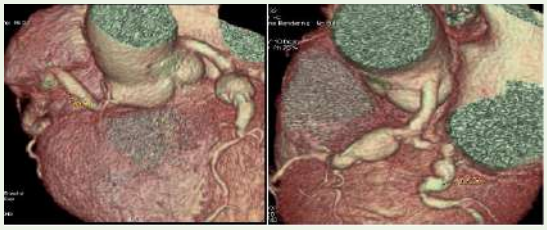

Figure 30: (A,B) 3D volume rendered images show multiple fusiform aneurysm of LAD & LCX.

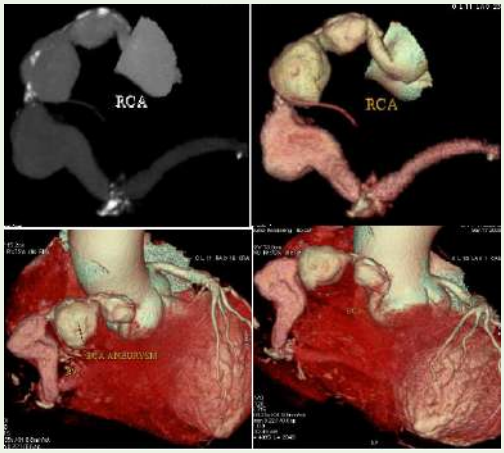

Figure 31: (A-D) CTCA MIP 2D curved (A), coronary tree (B) and 3D volume rendered images show multiple saccular and fusiform aneurysm of RCA with calcific hard plaques along the walls.

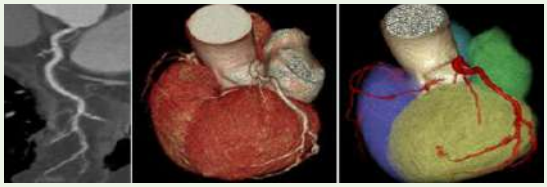

Figure 32: (A-C) CTCA 2D curved coronary vessel analysis(A) and 3D volume rendered images (B,C) show LAD hypoplasia.

Table 1: Classification of Congenital coronary anomalies [10].

Anomalies of origin	A. Number of ostia: Single, Multiple (>2) , B. Anomalous location of ostium in the appropriate coronary sinus C. Origin from opposite coronary sinus - (a) RCA arising from the LCS, (b) LMCA arising from the RCS, (c) LCX or LAD artery arising from the RCS D. Origin from non-coronary sinus-LCA or RCA (or branch of either artery) E. Originfrom Pulmonary artery– (a)Anomalous left coronary artery(ALCAPA) (b) Anomalous right coronary artery (ARCAPA) F. Abnormalities of angle of origin
Anomalies of course (normal origin);	Myocardial bridging, Duplication
Anomaliesoftermination;	Coronaryartery fistula, Coronary arcade, Extra cardiac termination
Intrinsic coronary arterial abnormality	Coronary stenosis,Atresia, Ectasia / aneurysm

Discussion

The coronary arteries arise from the aortic sinuses, converging towards the apex of the heart. Normally, there are three main coronary arteries, the right coronary artery (RCA) which typically arises from the right sinus of Valsalva (RSV) of the ascending aorta and supplies the right side of the heart, left circumflex artery (LCX) and left anterior descending (LAD), artery arising from a common stem, the left main coronary artery (LMCA) which arises from left sinus of Valsalva (LSV). Among these, the origin of the posterior descending coronary artery (PDA) from either the right (70%) (Figure 1) or the left (10%) (Figure 2) coronary artery defines the coronary dominance, co-dominance (Figure 3) in 20% of cases, with the dominant artery usually providing blood supply to the sino-atrial (SA) and atrio-ventricular (AV) nodes, albeit with some exceptions. Other common possible findings include trifurcation of the LMCA, with a Ramus intermedius (in ≈20% of the cases) (Figure 4), distributing across a variable portion of the lateral wall of the left ventricle [7].

Congenital coronary anomalies (CCA) may be defined as a coronary pattern or feature that's encountered in less than 1% of the general population.

In summary, we are able to divide the coronary feature in two groups:

(1) Normal coronary anatomy, defined as any morphological characteristics seen in > 1% of unselected sample. This group also includes normal anatomical variants, defined as alternative and comparatively unusual morphological feature observed in > 1% of the population; and

(2) Anomalous coronary anatomy, defined as morphological features found in < 1% of the population [5-7].

Most of the coronary anomalies remain asymptomatic and are incidental to investigations by coronary angiography. Coronary artery anomalies are classified as benign (80.6%) but potentially serious anomalies (19.4%) [2].

Anatomic characteristics that make a CCA malignant include-

(1) Single coronary artery (Figures 5 & 6),

(2) Origin from the pulmonary artery,

(3) Origin from the opposite aortic sinus (Figures 9-11, 13 & 14),

(4) Passing between the aorta and pulmonary artery (Figures 13, 14, 16 & 18),

(5) Acute-angle take off resulting in a slitlike orifice (Figures 16 & 17),

(6) Passing intramurally (Figures 19-21), and

(7) Small artery due to ostial stenosis or atresia [2].

For several decades, these anomalous coronary arteries were identified by conventional catheter coronary angiography. MDCT coronary angiography has been accepted as the ideal method for evaluation of patients with atypical chest pain due to its excellent temporal and spatial resolution [2,8].

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Among patients with CCA identified with MDCT, conventional angiography alone allowed correct identification of the anomalies in precisely 53% of cases [2].

Magnetic resonance coronary angiography could be a non-invasive method that doesn't require the utilization of contrast agents or ionizing radiation, and thus is superior compared to cardiac CT angiography and conventional coronary angiography. Its disadvantages are lengthy acquisition time and lower spatial resolution [9].

Congenital coronary anomalies are classified according to origin, course, termination and intrinsic arterial abnormality [10] (Table 1).

Anomalies of origin;

A. Number of Ostia: Single, Multiple (>2);

Single coronary artery is an extremely rare congenital abnormality seen in 0.024%-0.044% of the population (Figures 5 & 6) [11]. In most of the cases, aberrant RCA originates from the left main coronary artery and traverses anterior to the right ventricle or between the pulmonary trunk and ascending aorta [12]. The presence of a single coronary artery with an inter-arterial course may increase the danger of major adverse cardiac events [13]. A proximal obstruction within the main trunk can be devastating, due to the unfeasibility of collateral circulation development.

The RCA originating as a branch from the midportion of the LAD may be a very rare anomaly (Figure 5). Six cases have been reported within the literature up to now, and no patient had underlying congenital heart disease [14].

The absence of the LMCA with a separate origin of the LAD and LCX arteries is found in up to 0.67% of subjects (Figure 7) [7]. They may cause difficulties in catheterization during angiography, but they allow for the development of collateral circulation in the event of proximal obstruction in one of those vessels [8].

B. Anomalous location of ostium in the appropriate coronary sinus (Figure 8)

Ectopic RCA from right sinus of valsalva has a frequency of 1.13% and therefore by some classifications can be considered a variant.

C. Origin from opposite coronary sinus

According to historical reports, both the right and left coronary arteries originating from the opposite sinus could also be related to an increased risk of SCD [15].

(a) LCX or LAD artery arising from the RCS

The LCX artery most ordinarily arises from a separate ostium within the right sinus or as a proximal branch of the RCA in approximately 0.32%–0.67% of the population (Figures 9-11) [16]. Retro-aortic pathway is its commonest course, and there's no association with sudden death [8] (Figure 10).

An aberrant origin of the LMCA or LAD from the right sinus of Valsalva could be a rare anomaly that has been related to myocardial ischemia and sudden cardiac death [17] (Figure 9). Depending on the anatomic relationship of the anomalous vessel to the aorta and the pulmonary trunk, the anomaly is classified into 4 common courses: posterior (retro-aortic), interarterial (preaortic), anterior (prepulmonic), and septal (subpulmonic) course.

LAD artery branching off the RCA called the Type IV "Dual LAD" artery whose incidence ranges from 0.01 to 0.03% [18] (Figure 12).

(b) RCA arising from the LCS

RCA originating from the left coronary sinus or as a branch of a single coronary artery is found in 0.03% to 0.17% of the individuals submitted to angiography (Figures 13-15) [19]. The most common course of anomalous RCA from LCSis inter-arterial (malignant) (Figures 13,14). This variant will be related to sudden cardiac death in up to 30% of patients [20]. According to the finding on CT angiography, the interarterial course of the RCA is classified as either "high" or "low". A high course/outflow tract predisposes the patient to more adverse effects, like angina and sudden death, and requires more attention on the part of the radiologist. This is due to the fact that, during systole, both vessels adjacent to the coronary artery (the aorta and pulmonary artery) dilate, narrowing the channel through which the anomalous coronary artery passes, a phenomenon that's aggravated during physical work out. Conversely, when the interarterial course is low, the right ventricular outflow tract contracts during systole, counterbalancing the systolic expansion of the aorta and creating less narrowing within the coronary arterial course between the right ventricular outflow tract and the aorta [21].

(c) LMCA arising from the RCS

LMCA originating from the right coronary sinus (RCS) or as a branch of a single coronary artery occurs in 0.09% to 0.11% of the individuals submitted to angiography [19]. Proximal inter-arterial course occurs in 75% of such patients [19]. The origin of the LMCA from the RCS will be classified into 4 types- the LMCA passes between the aorta and pulmonary trunk (interarterial), anteriorly over the right ventricular outflow tract (pre-pulmonic), right of the RCA and pass posteriorly to the aortic root (retroaortic) and along the crista supraventricularis intramyocardially or subendocardially, surfacing within the proximal interventricular sulcus.

Sudden death can result from transient compression of the anomalous left coronary artery course, caused by dilation of the aorta and pulmonary artery, which is successively caused by the rise in blood flow during intense exercise, thus creating torsion or compression of the coronary artery between the aorta and also the right ventricular outflow tract [22].

D. Origin from Non-coronary sinus-LCA or RCA (or branch of either artery)

RCA from posterior sinus or Non-coronary sinus is a very rare anomaly (0.003%) in hearts without other congenital anomalies (Figure 15). The anomaly is sometimes not related to symptoms or complications so should be considered benign [23].

The origin of the LMCA from posterior sinus of Valsalva is sometimes seen in patients with other anomalies of heart and great vessels. This situation is extremely rare (0.0008%) and even

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more rarely associated with sudden death. This anomaly should be considered benign [23].

E. Origin from Pulmonary artery (PA)

(a) Anomalous left coronary artery from PA (ALCAPA)

It is also referred as Bland-white-Garland syndrome from the eponym of the authors who described it for the primary time in 1956. ALCAPA is one in all the foremost serious congenital coronary artery anomalies with prevalence of 1 in 300,000 live births [24]. Most affected patients show symptoms in infancy and early childhood. Approximately 90% of untreated infants die within the 1st year of life, and only some patients survive to adulthood [25]. In a study by Alexander. et al, the patients with ALCAPA had the extra finding of aortic coarctation with a patent ductus arteriosus [26].

(b) Anomalous right coronary artery from PA (ARCAPA)

ARCAPA has an incidence of 0.002% within the general population. Only 25-30% cases are related to structural heart defects [27]. Patients with associated cardiac anomalies are diagnosed early in life compared to patients with isolated ARCAPA. Those with associated cardiac defects may present with cardiac murmur, congestive symptoms, and sudden cardiac death or may remain asymptomatic and detected incidentally during evaluation of other problem [28]. The associated cardiac defects found in these patients were aorto-pulmonary window, tetralogy of Fallot, VSD, PDAAQ3, and aortic stenosis [28].

Left anterior descending artery from PA- Pulmonary origin of the LAD could be a very rare with a frequency of 0.0008%. This anomaly leads to myocardial ischaemia and sudden cardiac death [2].

All coronary arteries from PA - In this anomaly all the coronary arteries arise from PA, therefore the entire coronary circulation is supplied by the pulmonary artery. The prognosis of this CAA is poor and these patients usually die during the primary month of life. This CAA is additionally related to patent ductus arteriosus and with other major anomalies of the heart or great arteries [26].

F. Abnormalities of angle of origin

Acute take-off of LCX- Angelini et al defined this coronary anomaly in patients with an angle ≤ 45° between LMCA and LCX, within the left anterior oblique/caudal and/or right anterior oblique/caudal angiography Xray projections [29]. This anatomic variant has an incidence around 2% and should be relevant in reference to the technical difficulties that can complicate angiographic procedures on the LCX [29].

Anomalous location of the coronary ostium in the aortic root (Figures 16-18)

High origin of a coronary artery or left coronary trunk -Is defined as origin over 1 cm above the sino-tubular junction [30]. Preoperative identification of this anomaly is very important just in case of ascending aorta surgery and should cause difficulties in catheterization during angiography. Cross clamping of the aorta below a high-origin coronary artery may lead to unsuccessful induction of cardioplegia (Figure 18). Moreover, the higher the position of a coronary ostium, the higher the risk of coronary hypoperfusion, because the sinuses of Valsalva facilitate maximal coronary diastolic perfusion [30].

Anomalies of course (normal origin); Myocardial bridging, Duplication.

Myocardial bridging- A muscular or myocardial bridge is defined as an atypical course of artery during which it dips intramyocardially with resulting compression of the vessel during systole (Figures 19-21). The prevalence of this anomaly incorporates a wide selection from 0.15%-25% angiographically, to between 5% and 86% at autopsy. However, many reports from angiography may underestimate the prevalence of this anomaly, as recent studies with computed axial tomography (CT) have shown that myocardial bridging may be found in up to 25% of patients [31]. In some cases, myocardial bridging is responsible for angina pectoris, myocardial infarction, life-threatening arrhythmias, or perhaps death [32].

Duplication- Duplication of RCA (60%) &duplication of the LAD artery (40%) and their incidence in normal hearts is about 0.13%-1% of the overall population [33].

A split RCA is defined as an RCA that features a split PDA with the anterior subdivision of the RCA resulting in the distal portion of the PDA in the anterior free wall of the RV (Figures 21,23). The posterior bifurcation of the RCA maintains a course within the atrio-ventricular groove. Split RCA is typically called "double RCA", whether or not truly there don't seems to be two RCAs, but split portions of the posterior descending branch of the RCA with separate proximal courses and forms the uppermost portion of the posterior descending branch [1].

Duplication of LAD isn't intrinsically haemodynamically significant, but its presence may complicate surgical intervention when aorto-coronary bypass or other coronary artery surgery is performed [30] (Figure 22).

Anomalies of termination; Coronary artery fistula, Coronary arcade, Extracardiac termination

Coronary artery fistula(CAF)- Could be a condition within which a communication exists between one or two coronary arteries and either a cardiac chamber, the coronary sinus, the superior vena cava, or the pulmonary artery. In CAF, the involved coronary artery is dilated because of increased blood flow and is usually tortuous to an extent determined by the shunt volume [34]. The drainage site of the fistula encompasses a greater clinical and physiologic importance than does the artery of origin. The foremost common site of drainage is that the RV (45%), RA (25%),& PA (15%) [35]. The fistula drains into the LA or LV in less than 10% of cases [36].

When the shunt leads into a right-sided cardiac chamber, the hemodynamics resembles those of an extra cardiac left-to-right shunt; when the connection is to a left-sided cardiac chamber, the hemodynamics mimic those of aortic insufficiency. Myocardial perfusion is also diminished for that portion of the myocardium supplied by the abnormally connecting coronary artery. This situation represents a hemodynamic steal phenomenon and will result in myocardial ischemia [37].

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Coronary arcade- Could be a rare instance of communication that's large enough to be identified angiographically between the RCA and therefore the LCA within the absence of coronary artery stenosis [38].

Extracardiac termination- Connections may exist between the coronary arteries and extracardiac vessels (ie, the bronchial, internal mammary, pericardial, anterior mediastinal, superior and inferior phrenic, and intercostal arteries and also the esophageal branch of the aorta [9].

Intrinsic coronary arterial abnormality; Coronary stenosis, Atresia, Ectasia / aneurysm

Atresia- Coronary ostial stenosis or atresia may be a spectrum of rare developmental conditions with different patho-physiologic mechanisms and clinical implications. Coronary ostial stenosis or atresia affects the LMCA (Figure 24)**,** more frequently than it does the RCA. In adult patients with coronary ostial or proximal coronary stenosis it can be difficult to rule out acquired causes, like arteritis, which might produce conditions morphologically same as COSA; for instance, atherosclerotic, syphilitic, Kawasaki, and Takayasu aortitis have all been cited as causes of acquired ostial stenosis or occlusion [39]. LMCA atresia ia a rare congenital malformation, which is charecterised by absence of left coronary ostium and left main trunk within the left coronary arterial system. Patients is also asymptomatic or present with syncope,angina pectoris, myocardial infarction or sudden death [40].

Coronary artery aneurysms and ectasia are characterized by an abnormal dilatation of a coronary artery.

Ectasia- Ectasia is diffuse dilatation (>1.5 times the normal diameter) of the coronary arteries that involves 50% or more of the length of the artery [46]. Coronary artery ectasia is more common than coronary artery aneurysm (Figures 25-27) [42].

In Western countries, atherosclerotic aneurysms are most typical (50%), followed by congenital (17%) and infectious causes (10%); and in Japan, Kawasaki disease represents the predominant cause of coronary artery aneurysm [41,44]. Most of the patients are asymptomatic, and dilatation is an incidental finding. There's the chance of thrombosis, embolization and rupture.

Aneurysm - Is a focal dilatation of the vessel [43]. In true aneurysm the vessel wall consists of three layers: adventitia, media, and intima and in False aneurysm, the Vessel wall consists of one or two layers.Definition; coronary artery segments that have a diameter that exceeds the diameter of normal adjacent coronary segments or the diameter of the patient's largest coronary vessel by 1.5 times and involve less than 50% of the overall length of the vessel [41].

The reported frequency of coronary artery aneurysms varies widely from 0.3% to 5% [43]. The incidence is higher in men than in women (2.2% vs 0.5%)[38]. Atherosclerotic aneurysms are usually multiple & involve more than one coronary artery (Figures 29-31), as compared with congenital, traumatic, or dissecting aneurysms, which are mainly solitary [43,44]. The RCA is that the most often involved vessel(40%-61%), followed by LAD (15%-32%) & the LCX (15%–23%) [45] (Figures 29-31), left main trunk involvement is rare (0.1%-3.5%), & its presence is sometimes related to significant underlying two- or three-vessel artery disease [45] (Figure 28).

According to the American Heart Association statement, aneurysms are also classified in keeping with internal diameter as, small (<5-mm), medium (5–8-mm), or giant (>8-mm) [46]. In children, according to Japanese Ministry of Health in 1984, a coronary artery aneurysm is present when the diameter of the lumen is > 3 mm in children younger than 5 years old or > 4 mm in those 5 years old or older [46].

Kawasaki disease(mucocutaneous lymph node syndrome); It is an acute self-limited multisystemic panarteritis which will occur worldwide; nevertheless, it's markedly more prevalent in Japan and in children of Japanese ancestry [47]. The etiology of mucocutaneous lymph node syndrome remains unclear, although several epidemiologic and clinical features are strongly suggestive of an infectious cause because the initiating factor in a genetically susceptible host, but autoimmunity can also play a role in the pathogenesis [48].

Cardiac sequelae are the foremost important manifestations of Kawasaki disease and include coronary artery dilatation (Figure 27), Premature atherosclerosis and stenosis (4.7%), thrombosis, or occlusion with myocardial infarct (1.9%) [45].

Coronary artery aneurysm or coronary artery ectasia (Figure 27), develops in 15%-25% of untreated children with Kawasaki disease, in most cases within 3-6 months of the acute illness [49], but since the introduction of γ-globulin therapy, coronary artery aneurysm or ectasia occurs in exactly 5% of the cases[50]. Moreover, by 2 years after the onset of mucocutaneous lymph node syndrome, 49% of the patients have spontaneous regression of the aneurysms [51]. The LMCAis involved in 12% of the cases, the RCA in 3%,and both arteries in 8% (Figure 27) [52].

Hypoplasia of coronary arteries - Congenital hypoplasia of coronary arteries presents as a narrowed luminal diameter (less than 1.5 mm) in one or two of the three main coronary arteries with no compensatory branches. Limitations to blood flow caused by the narrow lumen result in symptoms of myocardial ischemia and sudden cardiac death. Most frequent variants in reported cases are hypoplasia of both LCX and RCA and hypoplasia of the LAD (Figure 32) [53].

Conclusion

Increasing use of MDCT for cardiac imaging has helped in the detection of many benign congenital coronary anomalies, but a little number is related to myocardial ischemia and sudden death. Increasing the employment of MDCT in cardiac imaging may yield diagnostic information on congenital coronary anomalies not obtained with invasive coronary angiography. Axial sections, multi planar reconstructions, virtual angioscopy, and 3D volume-rendered images should aid in the detection and improve the interpretation of such anomalies, which can be of immense help to the clinician planning interventional procedures.

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