



## COMPREHENSIVE MDCT ANALYSIS OF RARE CORONARY AND NON-CORONARY ANOMALIES IN PEDIATRIC PATIENTS

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### ABSTRACT

**Aims and Objectives:** to assess the spectrum of coronary and non-coronary artery anatomy and its various anomalies as depicted on CT angiography.

**Material and Method:** A prospective observational study was conducted in department of Radiodiagnosis, KGMU, for one year duration. CT Angiography (CTA) was done in 64 SLICE MDCT SCANNER (PHILIPS BRILLIANCE, Netherland). Study included pediatric population (age 1 - 20 years) having complaints of acute chest pain and suspected coronary artery disease with or without equivocal findings in 2D ECHO. Patients with Allergy to iodinated contrast agents, Cardiac arrhythmias, Renal insufficiency (GFR <30 mL/min), history of asthma were excluded from the study.

**Results:** Among total 20 enrolled patients 13 (65%) had coronary anomaly and 7 (35%) had extra-coronary anomaly. CT coronary angiography has 100% PPV for diagnosis of coronary and extra coronary anomalies.

**Conclusion:** ECG-gated multidetector CTA is an essential imaging tool for evaluating the coronary arteries and extra coronary anomalies. Knowledge of the imaging anatomy of various cardiac anomalies is essential for accurate diagnosis and proper treatment. Pre procedure CTA will help in better planning and understanding the hemodynamic changes and better prognostication of disease.

**Keywords:** CT Coronary Angiography, Congenital Heart Disease, Coronary Cameral Fistula, Malignant RCA, Hemi Truncus Arteriosus.

### INTRODUCTION

Congenital heart disease (CHD) encompasses a large group of cardiac and extra cardiac anomalies with variability of symptoms and severity of disease. In India, the prevalence of CHD is reported to be 8.5-13.6 per 1000 children (1). CHD contributes significant percentage to infant mortality (10%) in India. CHD even though present since birth but even could remain asymptomatic for a long time. CHD may result in intrauterine/childhood/adulthood mortality. A significant proportion of children born with CHD may lead a normal, productive life if diagnosed early and appropriate medical/surgical intervention is provided. Commonly used imaging for CHD evaluation includes echocardiogram, electrocardiogram, chest X-ray, CT angiography, and MRI. CT angiography is important in the evaluation of pediatric heart and associated extra cardiac anomaly thereby helping in taking the appropriate and timely decision for treatment.

With the help of Multiplanar reconstruction software cardiac anatomy and vascular course could be better appreciated. With the immense utility of CTA, there are certain inherent limitations with CT like risk of radiation. Pediatric thoracic CTA doses quoted in the literature vary greatly from 1–2 mSv to more than 50 mSv. Multiple advancement and modification were made in CT machines to reduce the radiation dose. CTA should be judiciously used with the thought of benefit risk ratio.

### MATERIALS AND METHODS

This prospective observational study was conducted in the Department of Radiodiagnosis using a 64-slice multidetector computed tomography (MDCT) scanner (Philips Brilliance). Institutional Ethics Committee approval was obtained prior to study commencement, and written informed consent were secured from all participants or their legal guardians. The study was carried out over a one-year period, from July 2023 to June 2024.

**Inclusion criteria:** a total of 20 patients (12 males and 8 females) aged 1–20 years were included. Patients were recruited based on clinical suspicion



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of congenital cardiovascular disease, presenting with symptoms such as chest pain, growth retardation, or abnormal cardiovascular findings on clinical examination. Additionally, patients with definitive or equivocal findings on two-dimensional echocardiography (2D echocardiography) suggestive of congenital cardiovascular anomalies were enrolled.

**Exclusion criteria:** included cardiac arrhythmias that could compromise image quality, impaired renal function (glomerular filtration rate  $<30$  mL/min/1.73 m<sup>2</sup>), and a documented history of hypersensitivity to iodinated contrast agents.

As part of the pre-imaging assessment, all patients underwent routine laboratory investigations, including complete blood count, renal function tests, and serum electrolyte analysis. Resting 12-lead electrocardiography and comprehensive transthoracic echocardiographic evaluation were performed in all cases prior to MDCT examination.

**Imaging Protocols:** CT Angio was done in 64 SLICE MDCT SCANNER (PHILIPS BRILLIANCE, Netherland) with all patients lying in a supine position involving the anesthetist to sedate the patients wherever needed. After initial frontal dual scout images (at 90 degree and 180 degree), a preliminary non-enhanced data set was acquired to calculate the calcium score and to optimize the field of view for the CTA. CT parameters were collimation -40 x 0.625 mm, tube voltage-120 kV and tube current-55 mAs. The data in the contrast enhanced studies, both in sequential and spiral modes, were collected by using the following parameters: collimation- 64 x 0.625 mm, gantry rotation time - 350 msec, tube voltage -100-120 kV and tube current -600-750 mAs depending on patient size. In the sequential mode, the pitch was 1.0 with 5 mm overlap between the slice stacks. Prospective ECG triggering determined the centre of the acquisition (end diastole, 75% of the cycle) while the width of this window was set between 100 and 200 ms according to HR. Contrast agent used in our study was iodixanol (Visipaque 320 mg/ml; GE Healthcare, Little Chalfont, UK) or iomeprol (Iomeron 400 mg/ml; Bracco, Milan, Italy).

**Premedication:** The image quality of coronary CTA is substantially improved in patients with a heart rate of less than 65 beats per minute because of reduced motion artifacts. The most common approach used for reducing the motion artifacts in patient with HR $>75$  was administration of an oral beta-blocker (50–100 mg of oral metoprolol is administered 1 h before the scan) or an intravenous beta blocker (5–20 mg of intravenous metoprolol is administered immediately before the scan) with a short half-life. Alternatively, calcium channel blockers were also used to reduce the heart rate in patients with a contraindication to beta-blockers. In addition, sublingual administration of short-acting nitroglycerin (2 tablets, equal to 0.8 mg)

immediately before a scan has been used to improve the visualization of the coronary artery lumen

**Statistical analysis:** The sensitivity, specificity, and accuracy of CT Angio in detecting coronary and extracoronary anomalies were calculated. Chi-square test was used to calculate the significance of CTA findings.

## RESULTS

The study included 20 patients, consisting of 12 males and 8 females, with an age distribution ranging from one to 16 years (Table 1). Among the patients, 11 out of 20 (55%) had coronary anomaly and rest 9 (45%) had extra coronary anomalies (table 2). Among 11 patients who had coronary artery anomaly, four of them had anomalous origin and course of coronary arteries, while seven had anomaly of coronary termination in the form of coronary cameral fistula. Among four patients who had anomaly of coronary origin and course, three had anomalous origin of RCA, which was arising from left coronary sinus (LCS) with retro aortic course, while in one patient LCX was arising from right coronary sinus (RCS).

Among seven patients who had coronary termination anomaly, two had anomaly of RCA to RV fistula, two had RCA aneurysm dissecting through interatrial septum, rest 3 had ruptured sinus of Valsalva aneurysm.

In extracoronary anomaly population, two had isolated pulmonary infundibulum stenosis with dilated right atrium due to severe TR and hypertrophy of right ventricular myocardium. Three had tetralogy of Fallot with hemi truncus arteriosus, three had sub mitral left ventricular aneurysm and one had left ventricular posterior wall big size aneurysm.

In our study, we identified 3 out of 20 patients having anomalous origin of RCA from left coronary sinus, the anomalous RCA has retro aortic course and was acutely angulated in proximal segment with significant luminal narrowing. Electively catheter angiography was done in the patients and we found the similar findings as was in CT Angio. (Fig 1 a, b &c).

We also identified 1 out of 20 patients having anomalous origin of LCX from right coronary sinus with inter-arterial course. (Fig 2 a &b)

In our study, we identified 2 out of 20 patients having coronary cameral fistula. This patient underwent echo study, which showed dilated RCA, but termination of RCA could not be identified properly. However, in CT Angio we clearly identified that RCA was terminating into right ventricle thereby creating left to right shunt in the patient. This patient had clinical feature of marfanoid habitus (arm span more than limb span), with feeble pulse and recurrent respiratory tract infection. Feeble pulse was due to left to right shunt due to coronary cameral fistula. (Fig 3)

In two patients terminal end of RCA was aneurysmally dilated, dissecting into interatrial septum (fig 4), with no obvious coronary cameral fistula. Second echo study was able to detect the aneurysm sac but the origin of sac and location of sac could not be properly commented.

Two patient found to have ruptured right sinus of Valsalva into left ventricle and was cause for continuous murmur in pre cordial region with bicuspid aortic valve. (5)

One patient had multiple sinus of Valsalva aneurysm but with no fistulous communication with any of the cardiac chamber (fig 6). Multiplicity of Valsalva aneurysm indicate some mycotic aneurysm the most common etiology therefore a meticulous work up to be done in the form of blood culture, urine

examination and CBC, sometimes bone marrow analysis if needed.

We also encountered two patients having isolated pulmonary infundibular stenosis with dilated right atria with TR with hypertrophy right ventricular myocardium. (Fig 7).

Three patients with diagnosed tetralogy of Fallot were found to have hemi truncus arteriosus in CT Angio with anomalous origin of LCA from RCS. (Fig 8).

Three patients had small sub mitral left ventricular aneurysm in CT Angio, which was missed in second echo study (fig 9).

One patient in the study was diagnosed with big size posterior wall left ventricular aneurysm (fig 10).

Table 1: Patient Distribution as Per Age Group in Study Population

Age Group	Number of Patients(N=20)
<5 years	2
5-10 years	6
10-15 years	4
15-20 years	8

Table 2: Coronary and Extracoronary Anomalies as Detected in CT Coronary Angio in Study Population

Diagnosis/Pathologies	Number of patients(N=20)
Anomalies of origin and course	
Anomalous origin of RCA from LCS	3
Anomalous origin of LCA from RCS	1
Anomalies of coronary termination with various coronary cameral fistula	
RCA to RV fistula	2
Ruptured sinus of Valsalva aneurysm	3
RCA aneurysm dissecting through atrioventricular septum	2
Extracoronary anomalies	
Isolated pulmonary artery stenosis	2
Tetralogy of Fallot with Hemitruncus arteriosus	3
Sub mitral left ventricular aneurysm	3
Left ventricular posterior wall large aneurysm	1

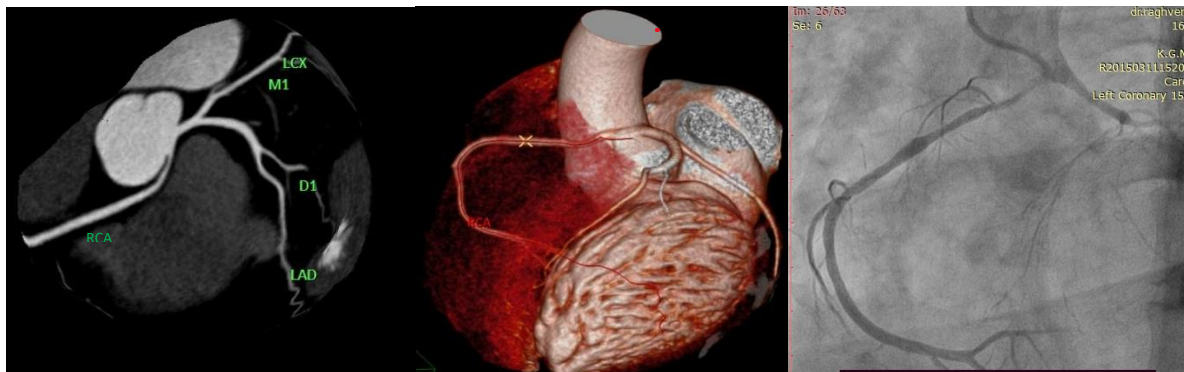


Fig 1: Coronary Ct Angio Mpr Images (A) And Vrt Images (B) Shows Origin of Rca from Left Coronary Sinus. Anomalous Rca is Taking Acute Course after Origin with Mild Narrowing at Its Proximal Segment, Similar Findings Noted in Catheter Angiography (C). Coronary Arteries and Its Branches are labelled with Green

Caption. Lad (Left Anterior Descending), M1 (Obtuse Marginal), D1 (First Diagonal), Lcx (Left Circumflex Artery), Rca (Right Coronary Artery)

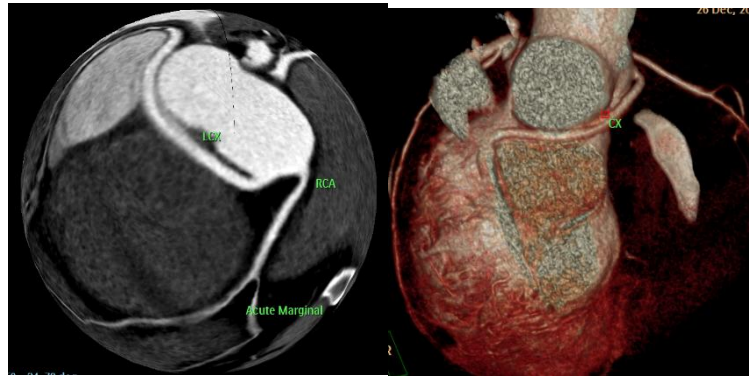


Fig 2 (a)

fig 2(b)

Fig2: Coronary CT Angio MPR Images (A) and VRT Images (B) Shows Origin of LCX from Right Coronary Sinus. Anomalous LCX Is Acutely Angulated at Its Origin and Taking Inter Arterial Course. Straight Red Arrow Shows Anomalous LCX. Coronary Arteries are labelled with Green Caption. LCX (Left Circumflex Artery), RCA (Right Coronary Artery) and Acute Marginal Branch

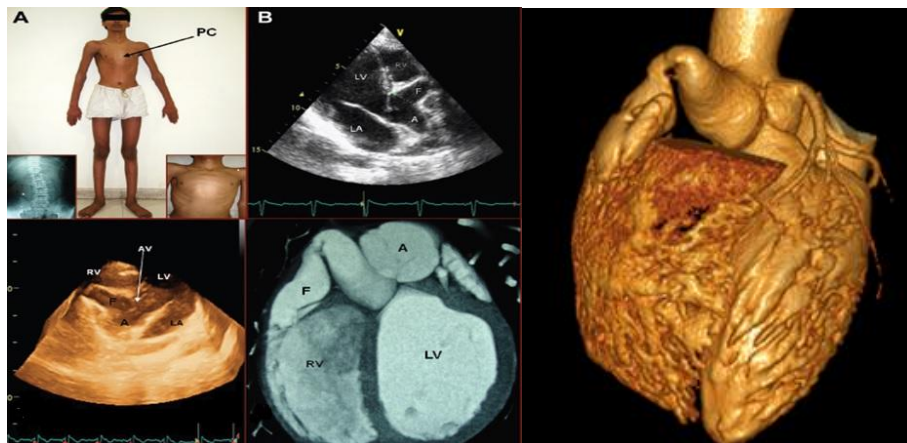


Fig 3 (a)

fig 3b)

FIG 3 (A) Shows Marfanoid Habitus of Patient with Precordial Bulge (Labelled PC) and Scoliotic Deformity in x ray Spine (Right Lower Insight Image). 2D Echo (Labelled B) Shows Dilated RCA (Labelled As A) and Fistula (Labelled As F). Left Ventricle Labelled as LV, Right Ventricle Labelled as RV, Left Atria Labelled as LA. CT Coronary Coronal Images Shows Dilated RCA with Coronary Cameral Fistula (Labelled As F). Aorta (A), Right Ventricle (RV), Left Ventricle (LV). Dilated RCA (Red Arrow). VRT Images (Fig 3b) Shows RCA TO RV Fistula (Red Arrow)

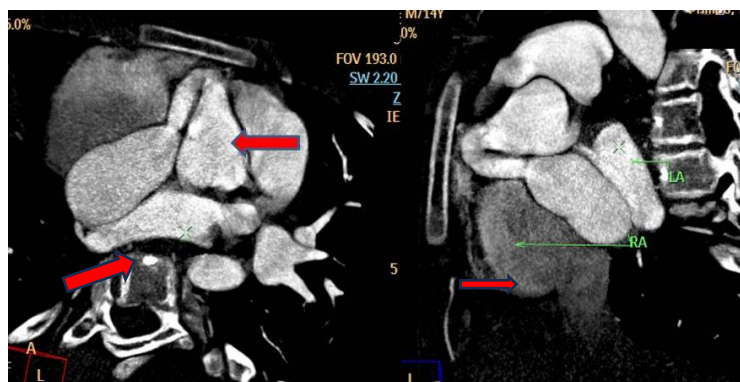


Fig 4(a)

fig 4(b)

Fig 4: Coronary CT Angio Axial (A) and MPR Images (B) Shows Dilated RCA with Terminal RCA Aneurysm Dissecting into Interatrial Septum. Thick Red Arrow Indicates Enlarged RCA, Thin Red Arrow Indicates Aneurysm. LA (Left Atria), RA (Right Atria)

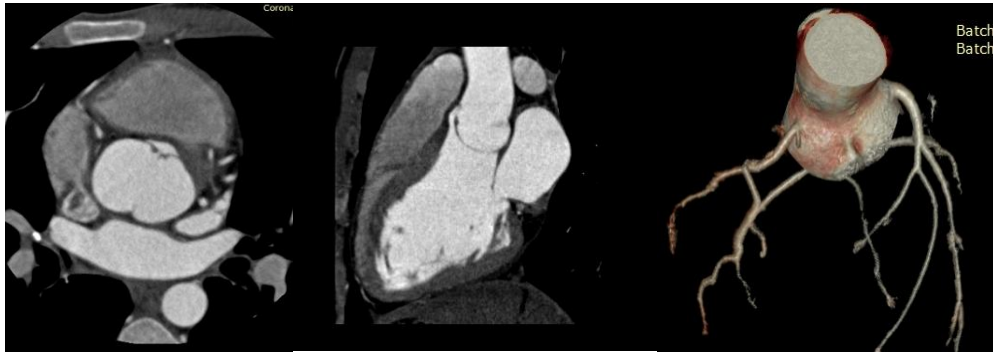


Fig 5(a)

fig 5(b)

fig 5(c)

Fig 5: Coronary CT Angio Axial (A) and MPR Images (B) Shows Ruptured Sinus of Valsalva (Shown In Red Arrow) from Right Coronary Sinus with Fistulous Communication To Left Ventricle (Shown In Curved Red Arrow). VRT Image Shows Ruptured Sinus of Valsalva from Right Coronary Sinus (Straight Red Arrow)

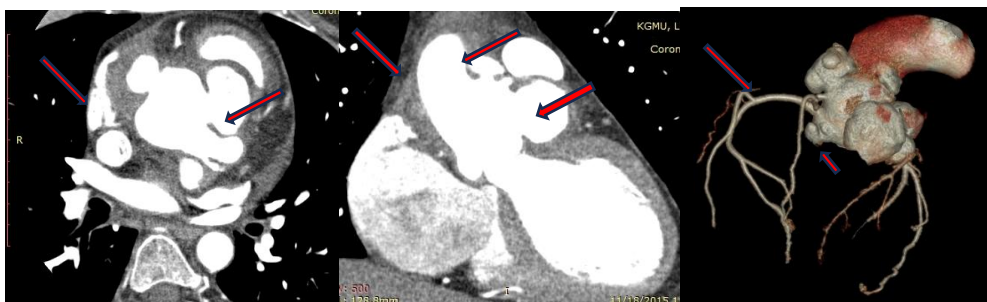


Fig 6(a)

fig 6(b)

fig 6(c)

FIG 6: Coronary CT Angio Axial (A) MPR Images (B) and VRT Images (C) Shows Multiple Sinus of Valsalva Aneurysm Arising From Right and Left Coronary Sinus (Depicted In Red Arrow)

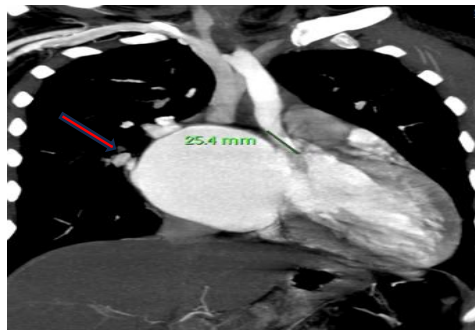


Fig 7

FIG 7: coronary CT Angio coronal images (a) shows infundibular stenosis of pulmonary artery with dilated right atria and increased right ventricular myocardium thickness. Length of stenosis is measured in green colour arrow

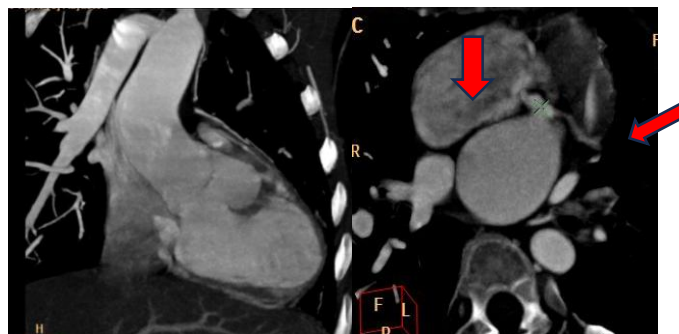


Fig 8 a

fig 8 b

FIG 8: Coronary CT Angio Coronal images (A) Shows Right Pulmonary Artery Arising from Ascending Aorta (Red Arrow). Axial Images (B) Show Common Origin of Left Coronary Artery (Straight Red Arrow) and Right Coronary Artery (Curved Arrow) from Right Coronary Sinus

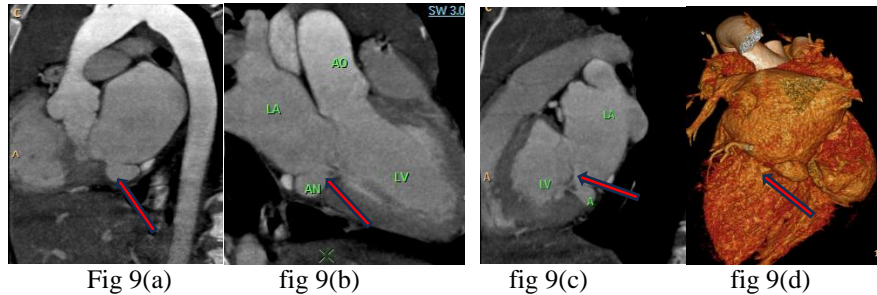


FIG 9: Coronary CT Angio Sagittal (A), Two Chambered Long Axis MPR (B), Two Chambered Short Axis MPR (C) and VRT Image (D) Shows Sub Mitral Aneurysm (Marked In Red Arrow). Aneurysm (AN), Left Atria (LA), Left Ventricle (LV), Aorta (AO)

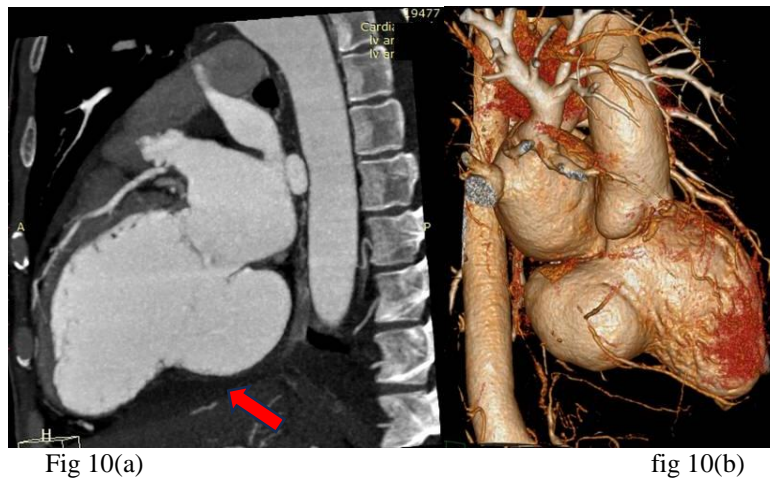


FIG 10: Coronary CT Angio Sagittal MPR (A) and VRT Images (B) Shows Giant Left Ventricle Posterior Wall Aneurysm (Shown In Red Arrow)

## DISCUSSION

The purpose of this study was to assess the range of cardiovascular anomaly in pediatric patients who presented with symptoms of recurrent RTI, poor growth, chest pain, recurrent transient ischemic attacks using CT coronary angiography. The findings indicated a male predominance, consistent with existing literature.

In our study, we identified three patients with anomalous RCA origin with malignant course. This finding correlates with a study conducted by Bae Young Lee [1]; anomalous RCA is labeled when RCA is arising from left coronary sinus or from non-coronary sinus.

RCA arising from left coronary sinus usually take acute retro-aortic course to supply the right ventricular myocardium. Retro aortic course is not always symptomatic but the acute angulation at the origin of anomalous RCA causes significant flow dynamics alteration with leading to occasional symptoms of chest pain. Catheter angiography was done and it showed the similar findings as detected in CT Angio, additionally it also proved that proximal RCA narrowing which was diagnosed in CT Angio was significant and was responsible for the symptoms in the patient (1).

We also identified one case in which left circumflex artery was arising from the right coronary sinus with inter-arterial course. Anomalous LCX origin is quite rare and whenever it is present, it is often symptomatic as seen in our case (2). The case report by Sotiris C Plastiras, et.al. also described the disease and its variable symptomatology according to left circumflex artery (LCX) course. Anomalous origin of the LCX is a well-read and identified coronary variant, and is considered most common coronary anomaly with reported incidence of approximately 0.37% to 0.7%.

Two of the patients in our study were diagnosed with coronary termination anomaly. In these cases RCA was terminating into the right ventricle and simulating the left to right shunt. 2D- Echo most often fails to detect the terminal end of the coronary artery and thereby could easily miss the diagnosis, CT angiography appears very useful in such cases for detailed analysis of cardiovascular anomaly due to high temporal and spatial resolution (3). Coronary cameral fistula is rare, and most of them are congenital. Because coronary cameral fistula can be asymptomatic and detected incidentally, the true incidence is difficult to evaluate. Coronary cameral fistula usually has various and complicated image

features, and the clinical symptoms mainly depend on the size, origin and drainage site of the fistulas. RCA is the most common origin site of this type, accounting for 50% of cases, followed by LAD (30%) and LCX (15%) (4). Study conducted by Ning li et al also found the similar results where RCA was the commonest involved artery in coronary cameral fistula.

Two patients in our study had interesting findings in which RCA was dilated with associated aneurysm in terminal end of RCA, which was dissecting in the interatrial septum. This is very rare entity where RCA end in aneurysm with high mortality if left untreated (5). Similar case reported by Hiroaki Osada where they found dissecting RCA aneurysm in old age patients, which was surgically corrected.

Two patients in our study have isolated pulmonary infundibular stenosis with dilated right atria with severe tricuspid regurgitation (TR) and hypertrophy of right ventricular myocardium due to high peripheral resistance to right ventricular outflow tract (RVOT). Pulmonary infundibular stenosis is commonly found in association with tetralogy of Fallot and associated Ventricular septal defect (VSD), but isolated pulmonary stenosis is rare. A Case report by Shyu KG et al. also had similar experience with the case and limited literature found regarding isolated pulmonary stenosis. Pulmonary stenosis can be valvular (the most common form), sub valvular (infundibular or sub infundibular) or supravulvular. On auscultation, systolic ejection murmur is found at left sternal border with right ventricular (RV) thrill. Surgical correction is the treatment of choice and is more successful when the obstruction is muscular with a short segment stenosis. (6)

Two patients with known teratology of Fallot in our study were found to have hemitruncus arteriosus in CT Angio. In hemi truncus arteriosus, either pulmonary artery takes origin from ascending aorta while the other pulmonary artery could have its normal origin from right ventricle. most often right pulmonary artery takes origin from ascending aorta. in few percentages of hemi truncus arteriosus left pulmonary artery may arise from ascending aorta, but in such case, arch is usually right side., however left pulmonary artery would have normal origin from right ventricular outlet (7). This anomaly has a high infant mortality of around 70% in first-year and 30% infant mortality within 3 months who do not undergo surgical correction(7). The anomaly created two separate blood circuits to each lung. The left lung received all of systemic blood volume from the right ventricle and the right lung received oxygenated blood from the left ventricle, which circulated back to left atrium. This lifelong systemic volume flow to the left lung caused progressive worsening of pulmonary hypertension and right ventricular hypertrophy. In contrast, the right lung received blood flow through the anomalous right

pulmonary artery; however, because of systemic pressure from the left ventricle, it too developed pulmonary hypertension. Surgical correction is best recommended therapy in such cases with good outcome (7).

Two patients in the study have left ventricular sub mitral aneurysm which was missed in second echo study, may be due to poor window or due to small size of aneurysm. Sub mitral aneurysm is a rare cardiac pathology of uncertain origin with varied clinical presentations (8). Some studies favor the congenital basis of this pathology, with suspected genetic link and racial predilection (in black population). Sub mitral aneurysm has stasis of blood leading to micro thrombi formation, which may be the cause of recurrent transient ischemic attack in patients. Usually, the submitral aneurysm patients had a history of previous myocardial infarction but in case no such history was found neither any ECG changes of old myocardial infarction found, so it was thought to be congenital. The other common associated cause of Sub mitral aneurysm are with Takayasu's arteritis and tubercular pericarditis. (8)

One patient in the study found to have bicuspid aortic valve with ruptured right sinus of Valsalva with fistula between left ventricles, and was responsible cause for continuous murmur in pre cordial region. Most aneurysms rupture into the right-sided heart chambers, and rarely into the left-sided heart chambers, pericardium, and pulmonary artery or superior vena cava (9). However, in our case the right sinus of Valsalva rupture in left ventricle. An aneurysm of the sinus of Valsalva ruptures in 35%, leading to acute symptoms in one fourth of the patients. Symptoms associated with rupture SOV are shortness of breath, chest pain, and fatigue (9).

Another patient had multiple sinus of Valsalva aneurysm (fig6) but with no fistulous communication with any of the cardiac chamber. Multiplicity of Valsalva aneurysm indicates some mycotic aneurysm as it was found in our case. Retrospective blood culture shows septicemia and positive bacterial growth Care report by Siddharthan Deepti a et al. also had similar findings and literature in support of our case. (10)

We also encountered one patient having large left ventricle posterior wall aneurysm. no other associated coronary anomaly seen in the case. Posterior left ventricular (LV) aneurysms are less common than anterior aneurysms. Large size aneurysm is mostly due to previous LCX territory infarct, but no ECG changes found in our case suggesting old mi changes. Idiopathic ventricular aneurysm is not very common, and whenever they occur, search for genetic mutation and family history needs to be done. (11)

## CONCLUSION

Multidetector CT coronary angiography is a tool to detect the cardiovascular anomalies in extensive way. It helps in detecting the things, which are not visible in second echo due to certain limitations. In cases of multiple sinus of Valsalva aneurysm, a meticulous search for infective cause should be sought for an extensive work up is needed in the form of blood culture, urine examination and CBC, sometimes bone marrow analysis if needed to reach the diagnosis and to offer best possible treatment. In pediatrics cases with congenital cardiovascular anomaly MDCT, coronary angiography is mandatory to know the disease status and its various anomalies.

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