ABSTRACT

Coronary Artery Anomalies (CAAs) are uncommon congenital cardiac anomalies, yet they frequently cause sudden cardiac death in the young population. Diagnosis is often challenging due to the diverse clinical presentation, which ranges from being asymptomatic and incidental to causing myocardial ischaemia or cardiac arrest. Imaging plays a vital role in detecting haemodynamically significant and non significant coronary anomalies, aiding in timely surgical or medical intervention to restore myocardial circulation and reduce morbidity and mortality. The present pictorial review aims to classify coronary anomalies and describe a few haemodynamically significant and non significant anomalies encountered in the department. Increasing awareness of these uncommon CAAs among radiologists can ensure early and accurate patient management with an exceptional prognosis.

INTRODUCTION

The CAAs are rare congenital cardiac anomalies with a documented incidence of only 1-2% of the population in general [1]. Despite being rare, CAAs stand as the second leading cause of sudden cardiac death in the younger population, especially athletes. The clinical presentation is often diverse, with a few anomalies being detected incidentally and others presenting with myocardial ischaemia. Chest pain, exertional dyspnoea, arrhythmia, myocardial infarction, and sudden cardiac arrest are a few of the clinical manifestations [2].

Several classification systems have been developed for CAAs. The widely accepted system is the one that classifies CAAs based on the origin, course, and termination of the coronaries. Another system is based on the haemodynamic impact of the anomaly, and CAAs are classified into haemodynamically significant categories [Atresia, Anomalous origin of Left Coronary Artery from Pulmonary Artery (ALCAPA), Anomalous origin of Right Coronary Artery from Pulmonary Artery (ARCAPA), interarterial course, and congenital fistula] and non haemodynamically significant anomalies [High take-off, duplication, aberrant conus artery, Shepherd’s crook RCA, systemic termination, prepulmonic/trans-septal course of the anomalously originated coronary vessel] [1]. Another system classifies CAAs based on the resultant myocardial ischaemia (with obligatory ischaemia or without ischaemia) [Table/Fig-1] [2,3].

Catheter angiography, although considered the reference standard for diagnosis of CAAs, is limited by its invasiveness and provides only a 2-dimensional projection of the anomalies [2]. Coronary Computed Tomographic Angiography (CCTA) is now the imaging investigation of choice due to its accuracy, quick acquisition, and superior spatial and temporal resolution [2]. The only setback for CCTA is the radiation hazard it poses. Cardiac Magnetic Resonance Imaging (CMRI) overcomes this limitation but is comparatively inferior to CCTA in imaging small-caliber vessels such as the coronaries [Table/Fig-2] [4].

Prompt diagnosis of haemodynamically significant CAAs is crucial as most of them require a timely surgical correction to restore myocardial circulation and prevent sudden cardiac death. However, detecting non haemodynamically significant anomalies is equally vital as they not only cause a dilemma during angiography but also result in difficult catheterisation and multiple other surgical complications [2].

The aim of the present pictorial review article is to provide an overview of a few of these rare congenital coronary and aortic sinus anomalies.
DISCUSSION

Anomalous Origin of Left Coronary Artery from Pulmonary Artery (ALCAPA)

Anomalous origin of Left Coronary Artery from Pulmonary Artery (ALCAPA), also known as Bland-White-Garland syndrome [5], is an exceedingly uncommon malignant coronary anomaly with an incidence of 1 in 300,000 population [4]. ALCAPA is often an isolated entity but can be associated with other congenital cardiac anomalies like interarticular or interventricular septal defects, coarctation of the aorta, etc., in 5% of cases [4].

The infant-type and adult-type presentations of the anomaly differ not only in the pathophysiology but also in the clinical presentation and prognosis. Most of the literature refers to the infant type ALCAPA, wherein the mortality rate is very high, with nearly 90% of untreated patients succumbing to death by one year of age due to myocardial ischaemia and congestive cardiac failure [6]. The adult counterpart of ALCAPA, which has been scarcely described, manifests as dysrhythmia, myocardial ischaemia/infarction, left ventricular dysfunction, and sudden cardiac death [1].

In the neonatal period, antegrade flow in the orthotopic RCA and anomalous LCA is maintained by the Ductus Arteriosus (DA). Hence, ALCAPA is clinically well-tolerated. Eight weeks after birth, closure of the DA occurs and leads to a fall in the pressures of the Main Pulmonary Artery (MPA) and ALCAPA.

In infant type ALCAPA, no collaterals develop. To compensate for the dropped pressures in MPA, retrograde flow occurs from LCA into MPA. This left-to-right shunt is termed the "coronary steal phenomenon" and leads to myocardial ischaemia [1]. If infant-type ALCAPA is promptly corrected by surgery, there is a good survival rate. However, if left untreated, death occurs by the end of the 1st year of life due to inevitable myocardial infarction and dysrhythmia [6]. In adult type ALCAPA, good collateralisation occurs between RCA and LCA (through intercoronary and systemic collaterals), compensating for the flow in ALCAPA. However, due to the left-to-right shunt, aneurysmal dilatation of the native vessels occurs. Despite flow compensation, chronic subendocardial ischaemia occurs, leading to myocardial infarction, dysrhythmia, and sudden cardiac death [1].

Adequate development of intercoronary and systemic collaterals, which compensate for the ischaemia due to the coronary steal phenomenon, and/or a dominant RCA, ostial stenosis of ALCAPA limit myocardial ischaemia and favour survival into adulthood [Table/Fig-3a-g].

2D and transthoracic echocardiography are the initial diagnostic tools in most cases, while invasive coronary angiography is the gold standard investigation [7]. However, coronary CT angiography and cardiac MRI have become reliable tools for diagnosis, essentially because they are non invasive, accurate, provide direct demonstration of the anomalous origin of the Left Coronary Artery (LCA), risk stratification, and easy follow-up [8]. CCTA is the imaging gold standard for ALCAPA [9].

On cross-sectional imaging, direct visualisation of the origin of the LCA from the pulmonary artery is the diagnostic hallmark [4]. The steal phenomenon can be demonstrated as a jet from the LCA to the Main Pulmonary Artery (MPA) and can be quantified by MRI [4]. Dilated and tortuous RCA, LCA, and other branches indicate compensated chronic left-to-right shunt. Intercoronary and systemic collaterals (along the epicardial surface, interventricular septum) indicate systemic compensation for the flow in the LCA. Left ventricular hypertrophy, wall motion abnormalities (hypokinesia, akinesia), Mitral valve dysfunction (prolapse, regurgitation), and delayed subendocardial gadolinium enhancement indicate chronic ischaemia [4].

The major differential diagnoses are atherosclerotic vessel dilatation (plaques can be visualised), Kawasaki disease (multiple coronary aneurysms in a young patient with a history of viral illness), coronary artery-coronary sinus fistula (dilatation of the affected vessel only), Takayasu arteritis (extensive involvement of the aorta and great vessels), endocardial fibroelastosis, and dilated cardiomyopathy in children [10].

Surgery (to restore the dual-coronary circulation) is the definitive treatment for ALCAPA [3], particularly in the infant type, because if left untreated, the mortality rate is very high. In asymptomatic adults with ALCAPA, medical management can be considered. One-coronary system restoration, with ligation of the LCA from the MPA, is now obsolete [11]. Coronary button transfer (reimplantation of the LCA into the aortic sinus) and the Takeuchi procedure (transpulmonary baffle from the MPA implanted into the aortic sinus) are two-coronary system surgeries performed in infants. Ligation of the LCA with Coronary Arterial Bypass Grafting (CABG) to the LCA is the surgery of choice in adults [3]. Patients with severe left ventricular dysfunction may need a cardiac transplantation [3].

Bleeding, kinking, graft stenosis/occlusion, supraavalvular pulmonary stenosis, aortic regurgitation, baffle obstruction, and leaks are common complications [3].

Shepherd’s Crook Right Coronary Artery (RCA)

A rare variant of the RCA with a normal origin but an abnormal course. After an orthotopic origin from the sinus of Valsalva, this variant of the RCA demonstrates a high and tortuous course with an acute or obtuse curvature and a kink, hence the name [Table/Fig-4a, b]. The documented incidence of Shepherd’s crook RCA is approximately 5% [12]. Although clinically indolent, reporting this variant is significant to plan percutaneous interventions. It poses a technical difficulty while negotiating the catheter beyond the kink and thus results in higher intra and perioperative complications [1]. The acuteness of the curvature and its distance from the coronary ostium can facilitate adequate preprocedural planning [1].
Separate Origin of Conus Artery

The conus artery is the first branch of the RCA and supplies the infundibulum of the pulmonary trunk [13]. In about 23 to 51% of patients, the conus artery may arise with a separate ostium from the right sinus of Valsalva [Table/Fig-5a, b] [3]. This aberrant conus artery is called a third coronary artery or accessory coronary artery by a few [14]. Although clinically inert, the aberrant conus artery poses a technical difficulty during angiography, may be missed on catheterisation, and is also at a risk for injury during cardiac surgeries [4]. Conversely, the separate origin of the conus artery has an advantage of providing a collateral source of blood supply in cases of Left Anterior Descending artery (LAD) obstruction by forming an anastomotic ring of Vieussen’s with a branch from the Left Main Coronary Artery (LMCA) or LAD [15].

Sinus of Valsalva Aneurysm (SOVA)

The three Sinuses of Valsalva (SOV) are dilatations in the aortic root complex, between the aortic annulus and sinotubular ridge. Functionally, they prevent the occlusion of coronary sinus ostia when the corresponding aortic valves open during systole [16]. By definition, SOVA are dilatations of one or more of the aortic sinususes or a saccular/tubular outpouching arising from the normal-sized SOV [16]. They are rare anomalies with a prevalence of 0.09% and constitute 0.1 to 3.5% of congenital heart defects [16], SOVAs are critical congenital anomalies, with catastrophic sequelae if left untreated. SOVA commonly involves the right sinus (65-85%), followed by the non coronary sinus (10-30%). The left coronary sinus is the least involved (<5%) [17]. Men are affected more than women, with a ratio of 3:1. The global prevalence is relatively higher in eastern and Asian countries [17].

The SOVA may be congenital or acquired in origin and hence can present at any age [16]. Congenital SOVAs may arise due to defects in the elastic lamina (as in Marfan’s or Ehler-Danlos syndromes) or due to the incomplete fusion between the aortopulmonary septum and distal bulbus cordis. Congenital SOVA is associated with other anomalies like Ventricular Septal Defects (VSDs) (in 30-60% of cases), bicuspid aortic valve (in 10% of cases), aortic regurgitation, CAA (Aberrant LCA), Atrial Septal Defects (ASDs), left ventricular non-compaction, Patent Ductus Arteriosus (PDA), left-sided superior vena cava, hypertrophic obstructive cardiomyopathy, aortic/pulmonary stenosis [18].

Acquired SOVAs may occur due to infective (tuberculosis, syphilis, bacterial endocarditis), autoimmune (Behcet’s disease), degenerative (atherosclerosis, cystic medial necrosis), traumatic, or iatrogenic (repair of VSD, valvular repair, dissection, etc.) causes [16,17]. SOVA can be broadly classified into unruptured and ruptured atherosclerotic. Unruptured aneurysms can be symptomatic or asymptomatic. If asymptomatic, they are incidentally detected when the patient is being evaluated for an enlarged cardiac silhouette detected on a chest radiograph or for a murmur. Unruptured aneurysms, when large, become symptomatic due to their mass effect on adjacent cardiac structures.

Ruptured SOVAs present with cardiac failure, dyspnoea, chest pain, and eventually cardiac arrest, depending on the acuteness of the rupture and the chamber they communicate with. Right SOVAs usually rupture into the right atrium, right ventricle, left ventricle, or into the interventricular septum. Left SOVAs rupture into the left atrium, left ventricle, pulmonary artery, myocardium, or epicardium [Table/Fig-6a-d]. When they rupture into the epicardium, the resultant cardiac tamponade can cause coronary arterial compression and myocardial ischaemia. Based on the site of rupture, SOVAs have been classified into four types [16].

Echocardiography (2D, colour Doppler, and transthoracic) is often the initial investigation performed, followed by conventional angiogram for confirmation. ECG-gated cardiac CT and Cardiac MRI are the latest, non invasive, accurate imaging modalities with better spatial and temporal resolution that can define and delineate the SOV, rupture, communication, mass effect on adjacent cardiac structures, and further complications [18]. A precise surgical roadmap can be obtained by the surgeon with 3D volume rendering techniques. MRI has the added advantage of quantifying the haemodynamics, such as the aorto-cardiac shunt in ruptured SOVA and aortic regurgitation. Cross-sectional imaging also aids in postoperative follow-up. The differential diagnoses for ruptured SOVA are aortic root/ascending aortic aneurysm (occur above the sinotubular ridge), prolapsed aortic valve cusps (occur below the aortic annulus), and coronary arteriovenous fistulas (communication between a coronary artery and a cardiac chamber/systemic/pulmonary vessel) [17].

Although both ruptured and unruptured SOVAs have a high fatality rate, prompt surgical intervention ensures an exceptional prognosis. The mean survival duration after the detection of SOVA is 3.9 years [19]. Percardial or polyester patch closure is the surgical treatment for SOVA. The approach is either through aortotomy or via the chamber with which the ruptured aneurysm communicates, or sometimes both. Percutanous transcatheter rupture closure is a new innovative and less invasive technique, with fewer perioperative complications.
complications compared to open surgery. Amplatz occlusion device deployment and coil occlusions were successfully performed, with no reported peri-procedural complications [20]. Immediate postoperative complications include peripatch leakage, low cardiac output, ventricular arrhythmias, and haemorrhage. Late complications include recurrent aortic regurgitation, endocarditis, and para-aortic abscesses. Complications associated with unruptured aneurysms are cardioembolic events (due to thrombi within the SOVA), aortic regurgitation (occurs in 30 to 50% of the patients) [16], ventricular outflow tract obstruction, arrhythmia, and dissection into the Interventricular Septum (IVS). Asymptomatic patients with small SOVAs can be monitored, but those with a larger size must be operated on to avoid complications [18]. Clinical and imaging follow-up of unruptured SOVAs is similar to aortic aneurysms [21].

**Aortopulmonary Septal Defects**

Aortopulmonary Septal Defects (APSDs), also known as aortopulmonary windows, are extremely rare congenital anomalies, constituting about 0.1-0.2% of all congenital heart diseases [22]. They occur when there is an abnormal side-to-side communication between the ascending aorta and the main pulmonary trunk, while the orthotopic semilunar valves develop normally [23]. APSDs happen due to incomplete septation of the truncus arteriosus, which occurs when the opposing conotruncal ridges fail to fuse [23].

This condition was described by Elliotson in 1830 and was first successfully repaired by Gross RE in 1952 [24]. In about 50-66% of cases, APSDs occur in association with other congenital anomalies such as interrupted aortic arch, coarctation of the aorta, anomalous origin of the coronary arteries, atrial and ventricular septal defects, patent foramen ovale, Tetralogy of Fallot (TOF), and Transposition of the Great Arteries (TGA) [25].

Mori K et al., classified APSDs into three types based on the location of the communication: Type-1 involves a proximal communication between the great vessels, Type-2 is a distal type with distal communication between the great vessels, and Type-3 is the total type with communication between the entire ascending aorta and main pulmonary artery (MPA) [26]. Ho SY et al., added a 4th category to this conventional classification, known as the intermediate type, which involves communication in the middle aspect of the vessels with adequate superior and inferior walls [27].

The abnormal communication between the great arteries results in a left-to-right shunt, leading to pulmonary hypertension and congestive cardiac failure [Table/Fig-7a-e]. The clinical presentation includes failure to thrive, diaphoresis, tachycardia, tachypnoea, recurrent infections, bounding pulses, continuous systolic murmur, and cardiomegaly [23].

Pulmonary hypertension is often fatal in childhood and adolescence. If patients survive into adulthood, they become inoperable due to the development of severe pulmonary hypertension and Eisenmenger’s syndrome [28]. Primary diagnosis is often made using echocardiography. However, CCTA and Cardiac MRI can precisely demonstrate the anomaly, quantify the shunt flow, and depict other concurrent cardiac/coronary anomalies [29].

The closest differential diagnoses are Truncus arteriosus (which has a common truncal valve, unlike APSDs that have separate aortic and pulmonary valves) and patent ductus arteriosus (which clinically presents similarly to APSD due to the left-to-right shunt, but the connection occurs between the descending aorta and pulmonary artery) [23]. Surgical patch closure is performed for larger defects, while double ligation/suture closure is used for smaller defects [30]. Medical management with diuretics, digoxin, and Angiotensin-converting Enzyme (ACE) inhibitors serves as a double-edged sword. While these drugs provide symptomatic relief by reducing the afterload, they can result in impaired renal perfusion [23].

**Single Coronary Artery (SCA)**

Single Coronary Artery (SCA) is a rare congenital anomaly with an incidence of 0.024% to 0.06% on coronary catheterisation and 0.024% to 0.098% in CCTA [31]. By definition, it is a solitary vessel that arises from a solitary ostium, either the right or left aortic sinus, and supplies the myocardium of the entire heart with essentially normal distribution [32].

The SCA can occur in isolation or in conjunction with other congenital heart diseases such as Transposition of the Great Arteries (TGA), coronary Arteriovenous Fistula (AVF), Tetralogy of Fallot (TOF), Truncus arteriosus, Ventricular Septal Defect (VSD), bicuspid aortic valve, and patent foramen ovale [33]. Isolated SCA is often silent and asymptomatic. SCA becomes clinically significant when affected by atherosclerosis and/or when the course of the vessel is malignant and interarterial. In the latter scenario, patients can present with angina, syncope, or sudden cardiac arrest [32].

The SCA has been classified by Smith, Ogden and Goodyear, but the accepted classification was published by Lipton MJ et al., [34]. Yamanaka O and Hobbs RE modified Lipton’s classification and added two variant courses, namely transseptal and combined types of SCA [35]. Rigatelli G et al., classified SCA according to their clinical consequences: Type-1 as benign, Type-2 as fixed myocardial ischaemia, Type-3 as sudden cardiac death, and Type-4 with superimposed coronary artery disease [Table/Fig-8] [36].

### Parameters

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolation of ostium</td>
<td>Right</td>
<td>Described in parenthesis as (R) and (L)</td>
</tr>
<tr>
<td>Anatomical course</td>
<td>Type-1</td>
<td>SCA with course of a native RCA or LCA</td>
</tr>
<tr>
<td>Relationship of the anomalous/traverse trunk to MPA</td>
<td>Type-2</td>
<td>SCA originates from right or left aortic sinus; from the proximal SCA, anomalous vessel arises and crosses the base of the heart to assume the distal expected course of the remaining circulation</td>
</tr>
<tr>
<td></td>
<td>Type-3</td>
<td>SCA arises from right aortic sinus and trifurcates into RCA, LAD, LCX</td>
</tr>
<tr>
<td></td>
<td>A</td>
<td>Anterior to MPA</td>
</tr>
<tr>
<td></td>
<td>B</td>
<td>Between aorta and MPA</td>
</tr>
<tr>
<td></td>
<td>P</td>
<td>Posterior to ascending aorta</td>
</tr>
<tr>
<td></td>
<td>S</td>
<td>Trans-septal (above the inter ventricular septum)</td>
</tr>
<tr>
<td></td>
<td>C</td>
<td>Combined Type</td>
</tr>
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</table>

**Table/Fig-7**: Aortopulmonary septal defect - Type I in a 24-year-old male patient with known Patent Ductus Arteriosus (PDA), recurrent respiratory infections, chest pain and pan-systemic murmur. a) Axial CTA image showing a large communication (2.9 cm) between proximal ascending aorta and Main Pulmonary Artery (MPA) (red arrow) and grossly dilated MPA suggesting Pulmonary arterial hypertension (red asterix); b) Oblique coronal VRT image demonstrating APSD type I (white arrow); c) Axial CTA image showing PDA (red arrow) between arch of aorta and pulmonary artery; d) Coronal MIP image demonstrating normal Pulmonary valve (red arrow); e) Coronal MIP image revealing normal Aortic valve (red arrow).

**Table/Fig-8**: Lipton’s classification of Single Coronary Artery (SCA) [36].

The Type-B anomalous course of SCA, located between the aorta and pulmonary artery, is strongly associated with myocardial ischaemia and sudden cardiac death, especially when it originates...
from the right aortic sinus. It has a relatively higher incidence in young athletes. The exact mechanism is unclear, but it is postulated to be due to compression of the interarterial component of the vessel by the dilated main pulmonary artery and ascending aorta during exercise. Hence, prompt detection of this anomalous course is crucial for management [37].

The SCA arising from the left aortic sinus is a rare entity compared to those arising from the right aortic sinus. R-IIIC is the most common type of SCA [Table/Fig-9a, b] [37]. Although conventional angiography is the gold standard for diagnosing SCA, CCTA has superseded it owing to its non-invasiveness and superior spatial and temporal resolution [34]. Surgical management is reserved for symptomatic patients or those with a malignant arterial course. Surgical options include reimplantation of the anomalous vessel into the aorta, osteoplasty, Coronary Artery Bypass Grafting (CABG) of the anomalous artery, and pulmonary artery translocation [38].

**Conclusions**

A high index of suspicion, early diagnosis, and treatment can prevent sudden cardiac death from rare coronary anomalies. Coronary CTA is the imaging investigation of choice as it is non invasive, accurate, provides a surgical roadmap, and also aids in postprocedural follow-up.

**References**

For any images presented, appropriate consent has been obtained from the subjects. Yes

Was informed consent obtained from the subjects involved in the study? Yes

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