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Coronary Artery Termination Variations and Anomalies Identified on Coronary CT Angiography

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Abstract

The most common coronary artery termination anomaly is coronary artery fistula (CAF), but other variations or abnormalities of termination are increasingly recognized. The purpose of this article is to describe the spectrum of these termination variations and anomalies with the emphasis on coronary computed tomographic angiography (CCTA) illustration.

Keywords: Angiogram, CT, coronary vessel anomalies

Introduction

Coronary artery termination variations and anomalies can be subdivided as coronary artery fistula (CAF), intercoronary communication (coronary arcade), and coronary artery to systemic artery anastomosis.

Congenital Coronary Artery Fistula

Congenital CAF is an abnormal connection between a coronary artery and a heart chamber, coronary sinus, pulmonary artery, vena cava, or pulmonary veins without normal transition through the capillary bed of the myocardium. The incidence of congenital CAF is between 0.002% and 0.7% on conventional angiography (CA) studies.[1-4] However, in a more recent study, the prevalence has been reported higher up to 0.9% utilizing coronary computed tomographic angiography (CCTA), likely related to improved detection.[5,6] Clinical significance of a CAF is mainly determined by the drainage site, the size of the fistulous connection, and coexisting cardiopulmonary diseases, rather than the artery of origin. More than 90% of the CAFs drain into the systemic venous side of the circulation, including the pulmonary artery (Figure 1). Termination into the pulmonary veins or superior vena cava is very rare; only a few of such cases have been reported.[7-9] Although most cases are unilateral, multilateral CAF, involving more than 1 coronary artery, can also occur[10] (Figure 2).

Coronary artery fistula flow direction is toward the low-pressure side of the communication. Persistent fistulous connection leads to aneurysmal dilatation of the coronary artery due to gradual weakening of the walls from increased blood pressure and flow. The chambers that are involved may become dilated (Figure 3).

Although more than half of the patients are asymptomatic, some patients may have symptoms associated with hemodynamic disturbances. Coronary artery fistulas with drainage into the right heart chambers can cause left-to-right shunt symptoms and findings, while CAFs with drainage into a left cardiac chamber can mimic aortic insufficiency. Aneurysmal dilatation of the involved coronary artery can cause morphologic disruption of the aortic valve, and lead to aortic insufficiency. The hemodynamic indication for intervention is the pulmonary-to-systemic flow ratio greater than 1.5.[4] Congestive heart failure may develop secondary to volume overload. Low-flow CAFs may remain asymptomatic, but high-flow fistulas may cause myocardial ischemia and infarction that can be attributable to the decreased myocardial and endocardial blood flow as a result of coronary steal syndrome.[4,6] The association between coronary atherosclerosis and CAF is uncertain, but coexistence of these two may lead to worsening of ischemic symptoms.[11] Infective endocarditis, arrhythmia, rupture or thrombosis, thromboemboli from the aneurysmal coronary artery, and sudden deaths were also reported.

Coronary computed tomographic angiography is a very useful imaging modality, not only for diagnosis, but also for imaging follow-up after percutaneous or surgical treatment of the CAFs.[5,6] Contrary to CA, CCTA can provide information

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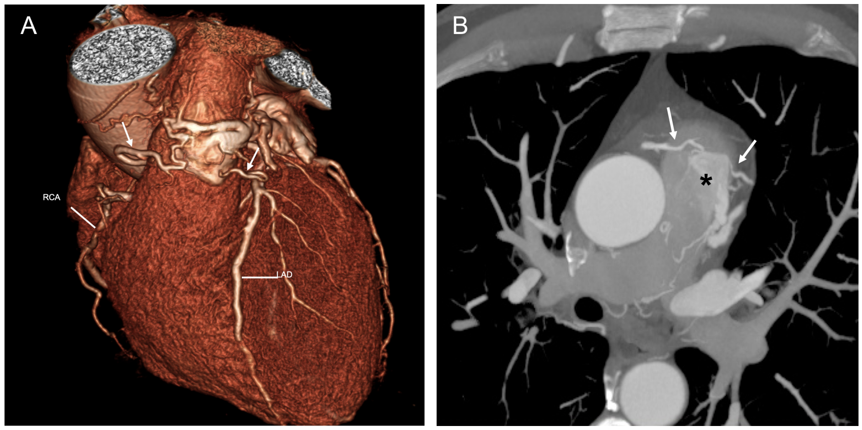

Figure 1. A 67-year-old man with chest pain. Bilateral coronary artery to pulmonary artery fistula. (A) Volume rendered and (B) axial maximum intensity projection (MIP) images show tortuous arteries arising from right coronary artery (RCA) and left anterior descending (LAD) (arrows) and draining into the main pulmonary trunk leading to abnormal contrast in the lumen (*).

regarding CAF complexity and other associated cardiac anomalies (Figure 4).

Intercoronary Communications (Coronary Arcade)

Coronary arcade is an extremely rare condition resulting in an open-ended circulation with unidirectional or bidirectional blood flow in the absence of coronary artery disease. There are only a few cases described in the medical literature. Yamanaka et al.[3] have identified only 3 cases in their 126 595 CA series, corresponding to an incidence of 0.002%. Most of them occur between the atrioventricular branch of the right coronary artery (RCA) and the circumflex coronary artery (CxA),[3,12–14] and rarely between the distal posterior descending branch of the RCA and the distal left anterior descending artery[3] or distal CxA.[14]

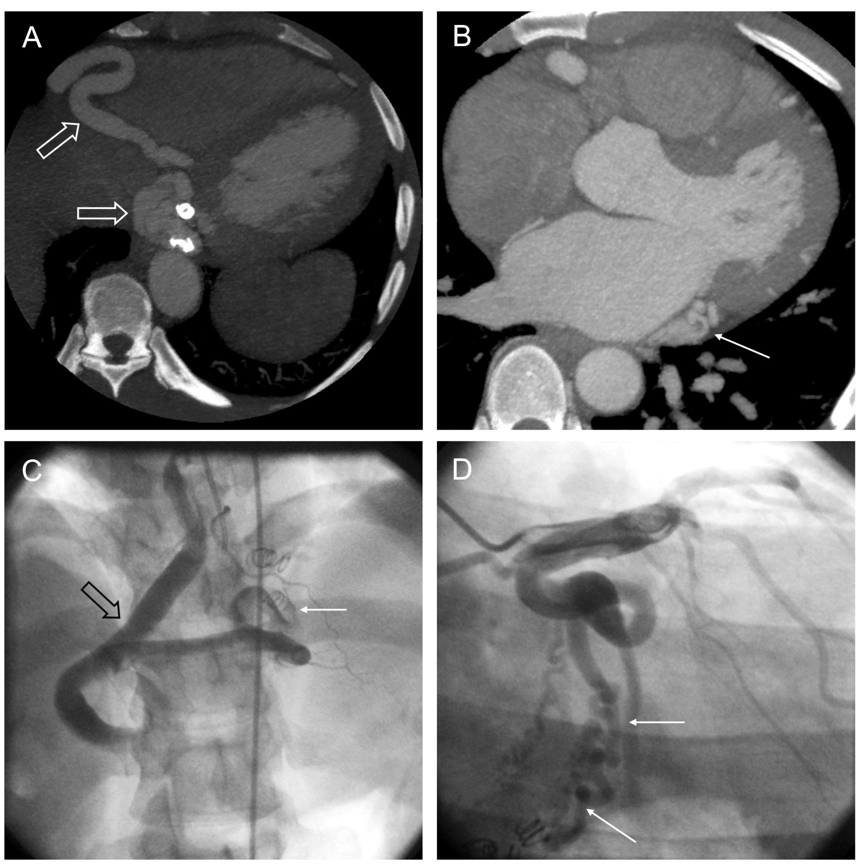

Figure 2. A 46-year-old asymptomatic man with a history of coil embolization of the RCA to coronary sinus fistula and incidentally detected cardiac murmur. (A) MIP image demonstrates dilated and tortuous distal RCA (open arrows), which terminates in the coronary sinus and coils in the distal segment of the RCA. (B) MIP image shows anomalous termination of CxA into the great cardiac vein (arrow). Conventional angiography images show (C) the dilated RCA (open arrow) and CxA (arrow) and (D) dilated tortuous distal RCA (arrows) terminating into the great cardiac vein.

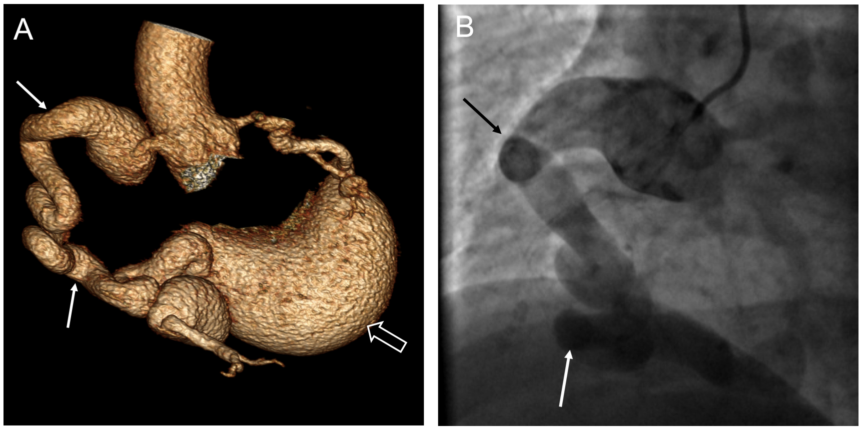

Figure 3. A 58-year-old woman with known RCA to the coronary sinus fistula referred for preoperative evaluation. (A) Volume rendered image of coronary computed tomographic angiography (CCTA) and (B) conventional angiography image demonstrate dilated and tortuous RCA (arrows) draining into the aneurysmal coronary sinus (open arrow).

During fetal life, the human heart contains a network of small interconnecting vessels between the right and left coronary arteries. These communications persist until about 8 months of age and then diminish in caliber.[15] Intercoronary connections may remain prominent and maintain a large caliber into adult life (Figure 5). They are invisible on CA due to their small calibers (<350 μ in diameter).[12] The clinical significance of these communications is not well known. It may provide protection against myocardial ischemia in the setting of coronary artery obstruction.[14] However, if unilateral blood flow is present, it may cause ischemia as a result of coronary steal phenomenon.[16,17]

Differential diagnosis includes collateral circulation associated with occlusive coronary artery disease. Normal coronary

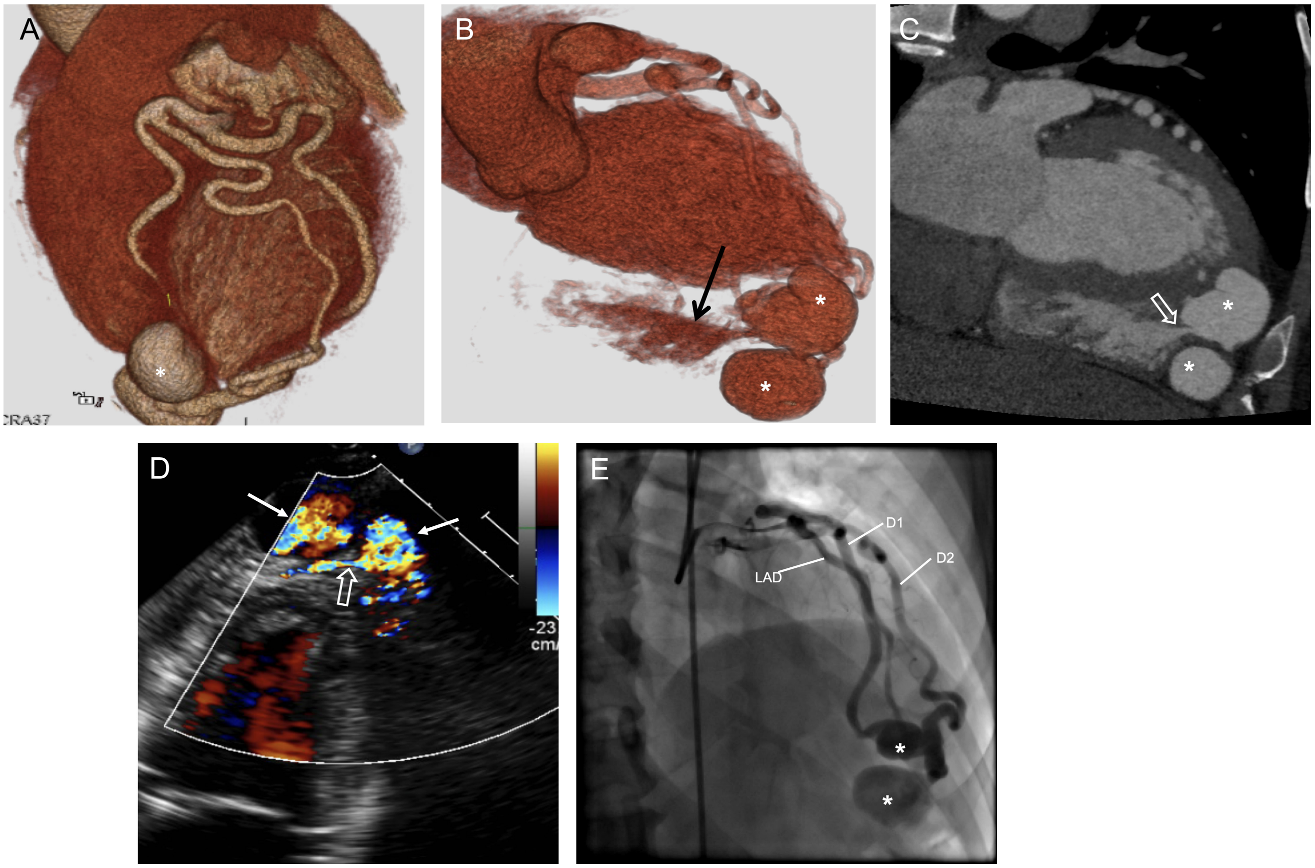

Figure 4. A 63-year-old man with chest pain. (A) and (B) Volume rendered CCTA images demonstrate bilobed aneurysm (*) supplied by the diagonal artery branches of the LAD and the communication between the aneurysmal segment and the right ventricular apex (arrow). (C) Coronal MIP image better demonstrates the communication between the aneurysmal segment and the right ventricular apex (open arrow). (D) Color Doppler echocardiography shows color mixing due to turbulent flow secondary to fistulous connection. (E) Conventional angiography shows ectatic feeding arteries and bilobed aneurysm (*).

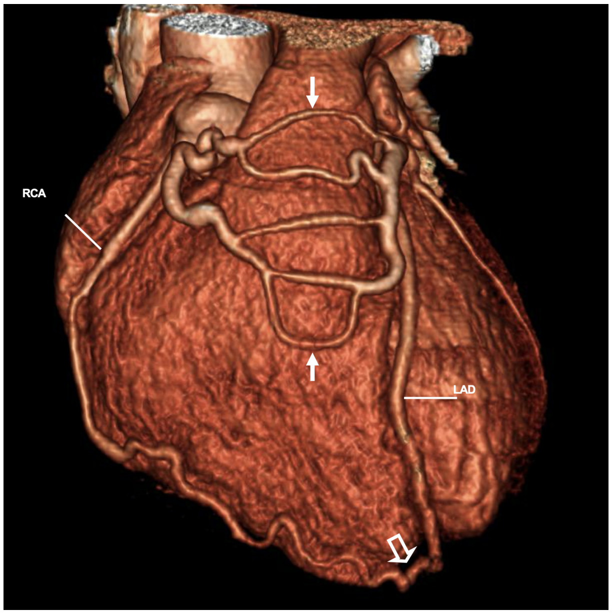

Figure 5. A 17-year-old male with murmur. Volume rendered CCTA image shows multiple large vascular connections (coronary arcade) between the RCA and LAD (short arrows) and extramural large anastomosis between the posterior descending artery (PDA) branch of the RCA and the distal LAD (open arrow).

circulation is functionally closed-ended, while coronary arcade is open-ended.[14] Coronary arcade demonstrates completely extramural, straight or gently curving open-ended morphology regardless of the diameter, while collateral circulations are extensively twisted and corkscrew in shape.[12]

Coronary Artery to Systemic Artery Anastomosis (Extracardiac Termination)

Coronary artery to systemic artery anastomosis can involve a variety of systemic vessels including anterior mediastinal, pericardial, bronchial, intercostal, superior and inferior phrenic arteries, pericardiophrenic branches of the internal mammary artery, and esophageal branches of the aorta.[18] These anastomoses are congenital in origin. Like CAFs, they can become enlarged and functional in either direction when there is a condition creating a considerable and persistent pressure gradient between anastomosing arteries, such as pulmonary artery hypoplasia, tetralogy of Fallot, supravalvular aortic stenosis, Takayasu arteritis, pulmonary thromboembolism, bronchiectasis, and pulmonary tuberculosis[19] (Figure 6). The most commonly recognized communication is an anastomosis between a coronary artery and a bronchial artery. Coronary artery to bronchial artery anastomosis (CBA) is seen in 0.008% to 0.6% of the CA cases.[19-21] Bronchiectasis is the most commonly reported cause of such connections to become functional.[22] The involved coronary artery is usually the left atrial branch of the CxA. RCA or multilateral involvements are rare[19-21] (Figure 7). Coronary artery to bronchial artery anastomoses generally remain hemodynamically insignificant and clinically silent because of the similar filling pressures in the coronary and bronchial circulation. In patients with pulmonary disease, the blood flow from the coronary to the bronchial circulation through the CBA may lead to coronary steal syndrome. Conversely, the blood flow in the CBA can be from the bronchial artery to the coronary artery in patients with occlusive coronary artery disease and may help to preserve the left ventricular systolic function. Associated complications include congestive heart failure, infective endocarditis, rupture of an aneurysmal fistula, and hemoptysis. Awareness of the possibility of CBA is important, since bronchial artery embolization distal to the anastomosis site may induce serious myocardial complications.[23]

In patients with suspected coronary artery to systemic artery anastomosis, the imaging field of view of the CCTA should include the aortic arch and the descending thoracic aorta to identify such extracardiac connections, and whenever such variation is identified on CCTA, careful evaluation for the associated cardiopulmonary disorders is essential.

Conclusion

Coronary computed tomographic angiography not only facilitates the identification of coronary artery termination variations and anomalies but also fully delineates the number of involved vessels and associated complications, such as

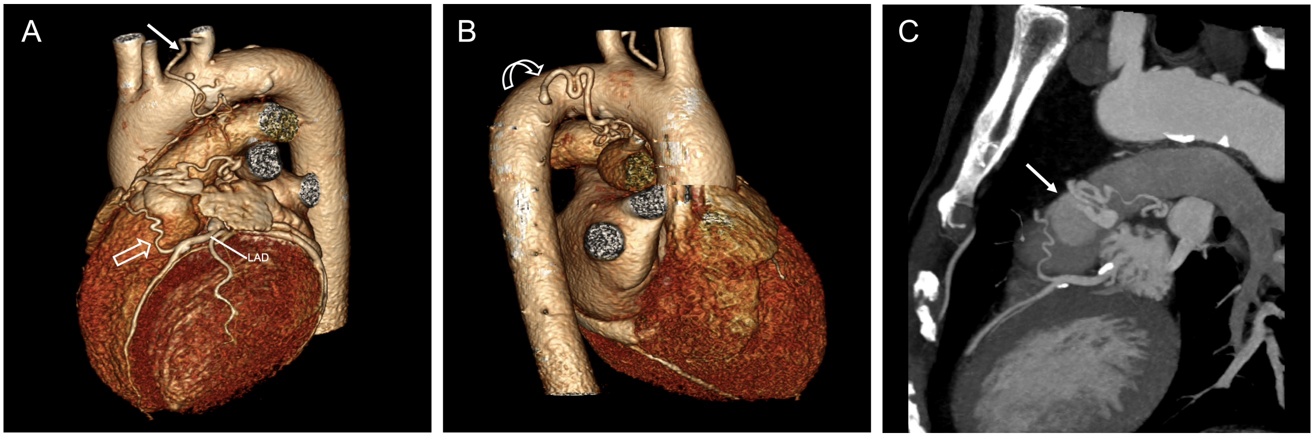

Figure 6. A 67-year-old man with a bicuspid aortic valve and severe aortic stenosis referred for preoperative evaluation of ascending aorta and coronary arteries. (A and B) Volume rendered images show abnormal arterial plexus arising from mid LAD (open arrow), proximal left subclavian artery (arrow), and descending aorta (curved arrow). (C) Sagittal oblique MIP image demonstrates the main pulmonary artery as the drainage site of the abnormal arterial plexus (arrow).

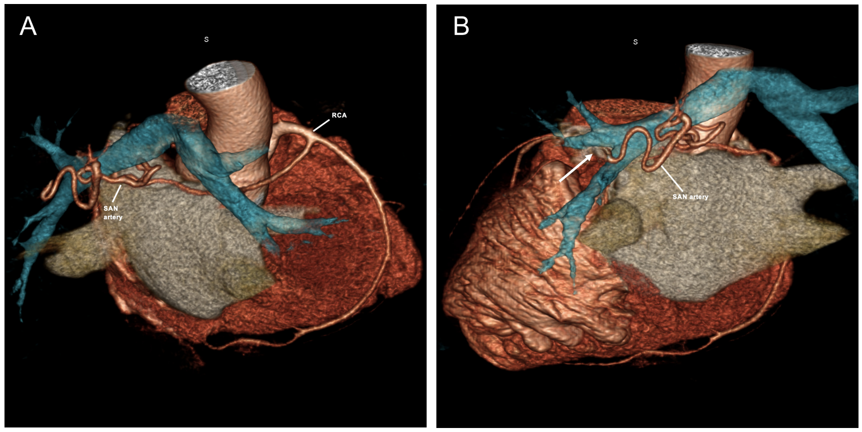

Figure 7. A 37-year-old man with Noonan syndrome, a history of VSD, and subpulmonary repair of congenital pulmonary stenosis presented with atypical chest pain. (A and B) Volume rendered CCTA images demonstrate the connection between the sinoatrial nodal (SAN) branch of the RCA and the left bronchial artery (long arrow).

aneurysms and intravascular thrombosis, that might be missed on conventional angiography, and thus provides essential information for treatment choice.

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