

Patients with varying courses of single coronary artery: case series

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Background

Single coronary artery (SCA) is a rare congenital anomaly where blood to the heart is supplied through a common trunk. Identifying these abnormalities is important because some variants can compromise myocardial blood flow and increase risk of sudden cardiac death.

Case summary

We present five patients with varying Lipton Group I and Group II SCA subtypes, corroborated on multi-imaging modalities and evaluated with comprehensive non-invasive as well as invasive testing. Their clinical presentations also vary from a spectrum of asymptomatic finding to angina equivalent. The decision for definitive surgical intervention involving unroofing of the involved vessel depends largely on symptoms and evidence of myocardial ischaemia.

Discussion

While SCA findings are often incidental and benign, understanding the origin, branching pattern, and course of the anomalous artery has implications in prognosis and treatment. This usually involves a combination of anatomic assessment with imaging such as coronary Computed Tomography Angiography (CTA), Magnetic Resonance Angiography (MRA), and/or coronary angiography as well as functional assessment with invasive testing using tools like instantaneous wave-free ratio and intravascular ultrasound both at rest and with stress. Individualized treatment plans can then be made through a multi-disciplinary approach involving adult congenital heart disease specialists and congenital cardiothoracic surgeons.

Keywords

Case report • Single coronary artery • Anomalous aortic origin of coronary artery • Interarterial course

Learning points

- Patients with single coronary artery (SCA) can present on a wide spectrum from asymptomatic incidental finding to angina equivalent to sudden cardiac death.
- Comprehensive assessment of SCA should include anatomic definition of its origin, branching pattern, and course as well as functional evaluation using invasive testing like instantaneous wave-free ratio and intravascular ultrasound at rest and with stress.
- Treatment plans including medical therapy and/or surgical intervention should be tailored to each individual's symptoms and functional assessment for myocardial ischaemia through a multidisciplinary approach.

Introduction

Single coronary artery (SCA) is a rare congenital anomaly that supplies perfusion to the entire heart. It is one form of anomalous aortic origin

of the coronary artery (AAOCA) with an estimated prevalence of 0.024–0.066% in the general population.^{1–3} One commonly accepted classification by Lipton *et al.*¹ categorizes SCA into three groups based on location of the ostium and anatomic course of its branches. *Group I*

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follows the course of normal right or left coronary artery (RCA, LCA) but continues in the atrioventricular groove to supply the contralateral territory after giving rise to posterior descending artery (PDA). *Group II* originates as a common ostia from either the right or left coronary sinus and bifurcates into RCA and LCA. The anomalous vessel will then traverse the base of the heart to supply its respective contralateral territory via one of several courses: prepulmonic, interarterial, retro-aortic, subpulmonic, or retrocardiac. *Group III* occurs when the left anterior descending (LAD) and left circumflex (LCx) branches arise separately from a common trunk. Anomalous LCA from right coronary sinus is less common but may confer a slightly higher risk of sudden cardiac death (SCD) than anomalous RCA from the left sinus, particularly if interarterial with a trans-septal or intramural course.^{4,5}

Here, we present five cases of SCA, one Lipton *Group I* and four Lipton *Group II* with varying anatomic courses. We describe in-depth approaches, assessments, and management of their coronary anomaly.

Timeline

Case Presentation

Patient 1

A 71-year-old woman is referred to clinic for abnormal stress test as part of pre-operative workup of a pelvic mass. Her nuclear stress test did not show any ischaemia or scar but a reduced ejection fraction (EF) of 35% without prior EF for comparison. She then underwent coronary angiography, which showed minimal coronary artery disease (CAD) and incidental finding of SCA from the right coronary sinus with continuation of LCx in the atrioventricular groove after giving rise to PDA and a diminutive LAD (*Video 1*). Her Lipton *Group I* SCA anatomy was confirmed by subsequent coronary CTA (cCTA, *Figure 1*). She was started on guideline-directed medical therapy for heart failure with reduced EF.

Patient 2

A 73-year-old man presents to clinic for evaluation of shortness of breath. His pertinent comorbidities include hypertension, hyperlipidaemia, diabetes, persistent atrial fibrillation on warfarin,

Patient	Demographics	Presentation	Single coronary artery (SCA) type and course	Testing	Management
1	71-year-old woman	Asymptomatic, abnormal stress test with reduced ejection fraction (EF) 35%	Lipton Group I SCA	<ul style="list-style-type: none"> • Nuclear stress test • Coronary CTA (cCTA) • Coronary angiography 	Guideline-directed medical therapy for heart failure with reduced EF
2	73-year-old man	Shortness of breath	Lipton Group II SCA, prepulmonic course from right sinus	<ul style="list-style-type: none"> • Nuclear stress test • cCTA • Coronary angiography 	Benign course, abnormal stress likely false positive in the setting of SCA. Reduced EF likely due to tachycardia-induced cardiomyopathy, underwent cardioversion
3	61-year-old man	Asymptomatic, abnormal screening cCTA	Lipton Group II SCA, subpulmonic course w/trans-septal segment from right sinus	<ul style="list-style-type: none"> • cCTA • Coronary angiography • Instantaneous wave-free ratio (iFR) and intravascular ultrasound (IVUS) at rest and pharmacologic stress 	Aspirin and statin for primary prevention, no need for surgical intervention
4	38-year-old man	Palpitations	Lipton Group II SCA, subpulmonic course w/trans-septal segment from right sinus	<ul style="list-style-type: none"> • 30-day event monitor • cCTA • Cardiac magnetic resonance imaging • Coronary angiography 	Trans-conal unroofing of left main

Continued

Continued

Patient	Demographics	Presentation	Single coronary artery (SCA) type and course	Testing	Management
5	73-year-old man	Dyspnoea on exertion	Lipton Group II SCA, interarterial course w/o intramural involvement	<ul style="list-style-type: none"> iFR and IVUS at rest and pharmacologic stress cCTA Coronary angiography iFR and IVUS at rest and pharmacologic stress 	Medical management with risk factor modifications

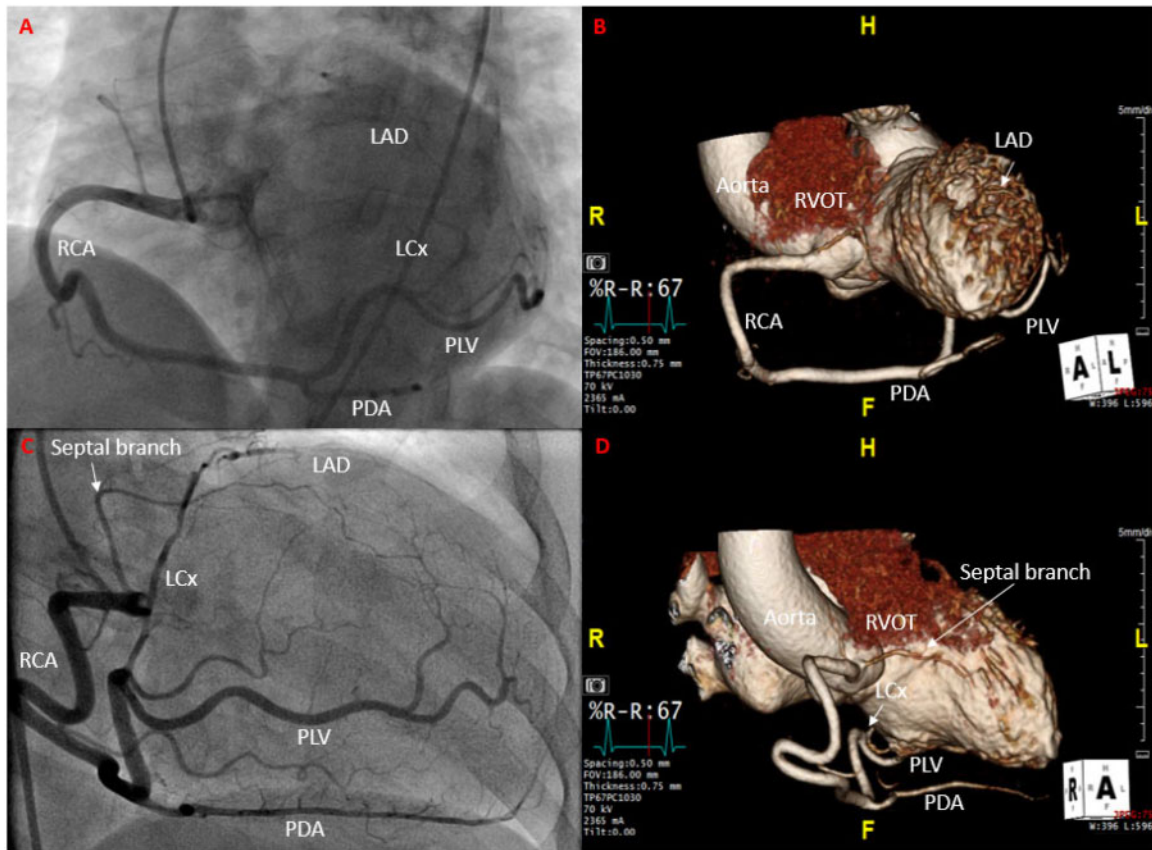


Figure 1 (A) Angiographic LAO view of single coronary artery arising from right sinus, continues after posterior descending artery to give rise to left circumflex and diminutive left anterior descending in the left system territory. (B) Computed tomography representation of similar view. (C) RAO view of single coronary artery. (D) Computed tomography representation. LAD, left anterior descending; LCx, left circumflex; PDA, posterior descending artery; PLV, posterolateral ventricular branch; RCA, right coronary artery; RVOT, right ventricular outflow tract.

and obesity with body mass index of 39. Transthoracic echo (TTE) showed newly reduced EF of 36% compared to normal EF in 2012. Patient then underwent a nuclear stress test, which showed reversible inferior and apical defects with stress. His

coronary angiography showed mild diffuse disease but detected an SCA from the right coronary sinus, which then gives rise to RCA and LCA (Video 2). The latter takes a prepulmonic course anterior to right ventricular outflow tract (RVOT) before bifurcating

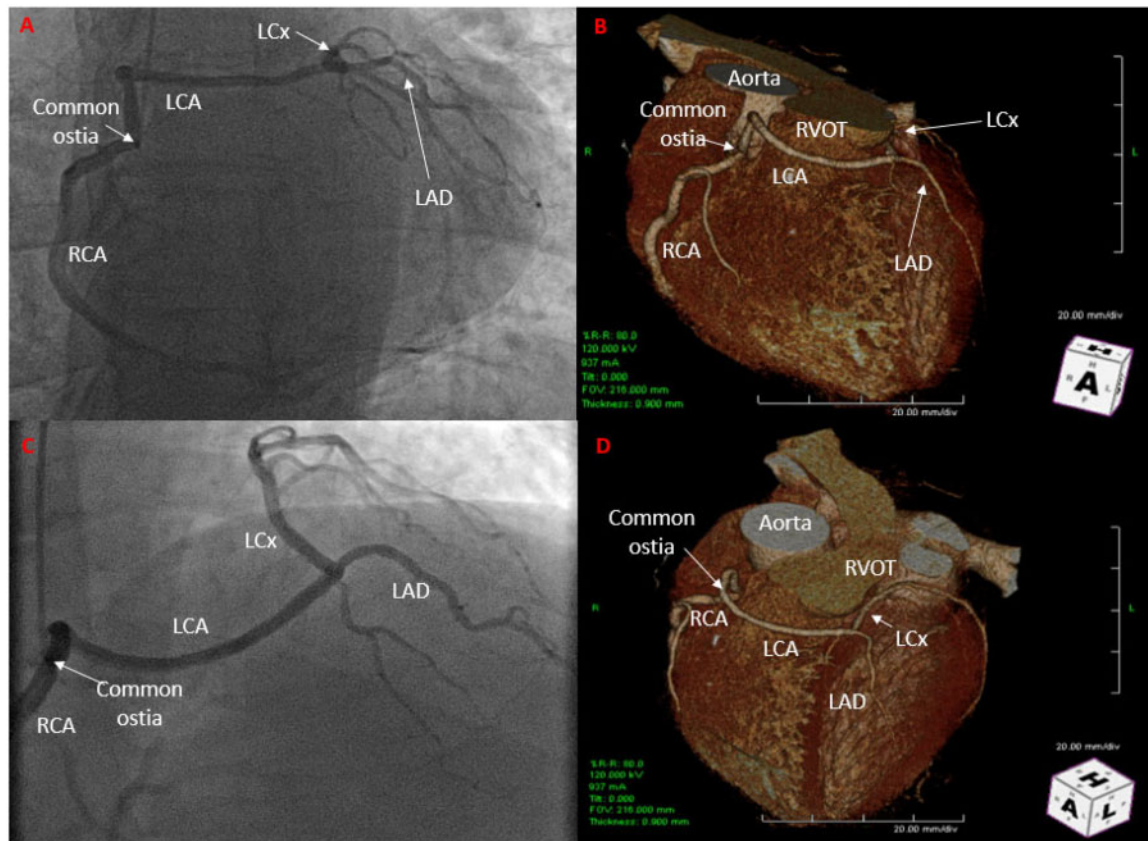


Figure 2 (A) Angiographic LAO view of single coronary artery arising from right sinus. Common trunk gives rise to right coronary artery and left coronary artery, which then takes a prepulmonic course anterior to right ventricular outflow tract before bifurcating left anterior descending and left circumflex branches. (B) Computed tomography representation of similar view. (C) RAO view of single coronary artery. (D) Computed tomography representation. LAD, left anterior descending; LCA, left coronary artery; LCx, left circumflex; RCA, right coronary artery; RVOT, right ventricular outflow tract.

LAD and LCx branches. These findings were corroborated by cCTA (Figure 2). Given the benign coronary course, his abnormal stress was considered a false positive test in the setting of SCA, and his reduced EF was attributed to tachycardia-induced cardiomyopathy, for which the patient underwent a cardioversion restoring sinus rhythm.

Patient 3

A 61-year-old asymptomatic man is referred to clinic after a screening cCTA showed an SCA from the right coronary sinus. Left coronary artery takes a subpulmonic course with a trans-septal segment before bifurcating into LAD and LCx (Figure 3). Patient underwent coronary angiography and invasive haemodynamic assessment using instantaneous wave-free ratio (iFR) at rest and with pharmacologic stress (dobutamine + atropine) achieving 92.4% maximum predicted heart rate (MPHR) as well as intravascular ultrasound (IVUS) imaging (Video 3). His results were negative with iFR of 0.92 and 0.90, respectively. Intravascular ultrasound also corroborated trivial intraluminal compression of trans-septal segment at peak stress. Given constellation of lack of symptoms and

benign invasive findings, patient was managed medically with aspirin and statin for primary prevention without need for surgical intervention.

Patient 4

A 38-year-old man initially presented with palpitations. A 30-day event monitor and TTE were unrevealing. He underwent subsequent cCTA, which showed an SCA from the right coronary sinus. Left coronary artery takes a subpulmonic course with a trans-septal segment before bifurcating into LAD and LCx (Figure 4). A cardiac magnetic resonance imaging did not show any ischaemia on perfusion imaging or fibrosis on delayed gadolinium enhancement. He proceeded to coronary angiography and invasive testing using iFR at rest and with pharmacologic stress, achieving 63.2% MPHR and IVUS (Supplementary material online, Video S1). While there was no angiographic evidence of CAD, there were haemodynamically significant intraluminal compression in the trans-septal segment of the left main (iFR of 0.85 with stress) as well as in the proximal LAD myocardial bridge (iFR of 0.81 at rest and 0.72 with stress). These findings were also confirmed on IVUS. Given the haemodynamic significance of

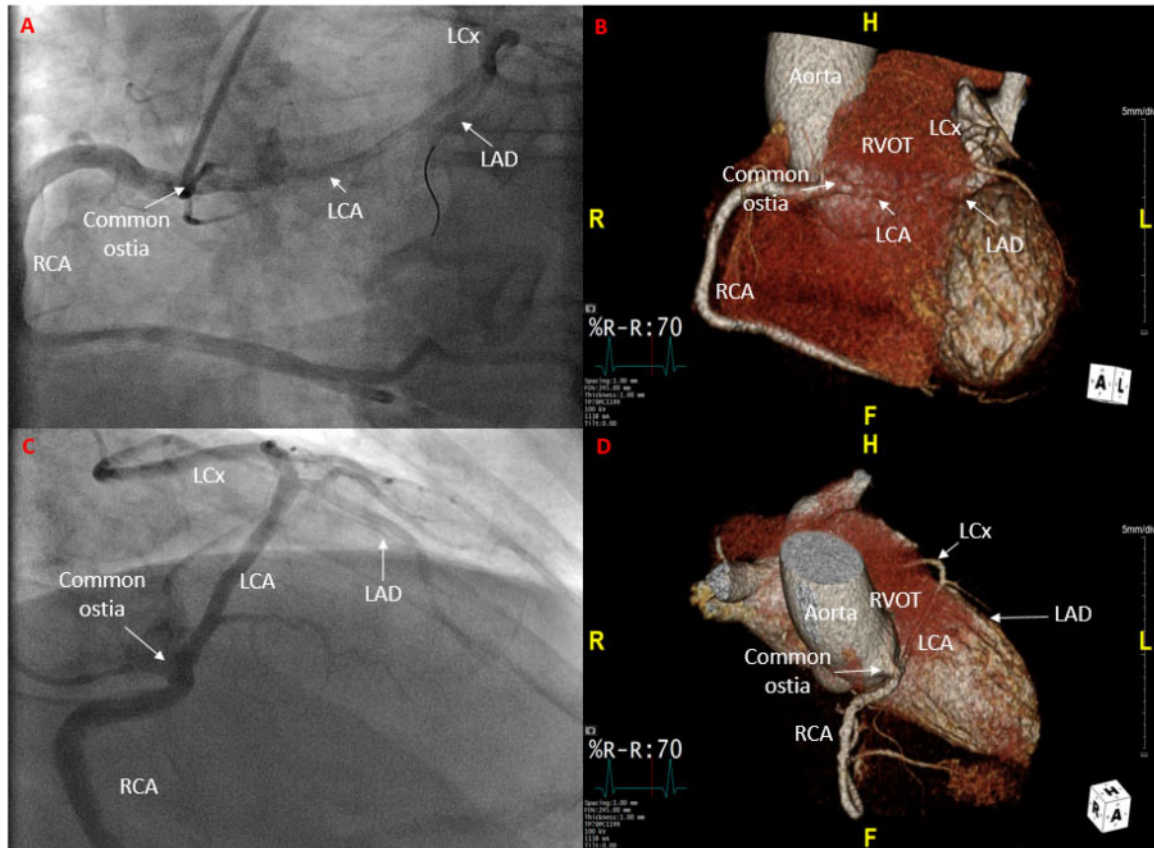


Figure 3 (A) Angiographic LAO view of single coronary artery arising from right sinus. Common trunk gives rise to right coronary artery and left coronary artery, the latter of which then takes a subpulmonic course between the aorta and right ventricular outflow tract before bifurcating left anterior descending and left circumflex branches. (B) Computed tomography representation of similar view. (C) RAO view of single coronary artery. (D) Computed tomography representation. LAD, left anterior descending; LCA, left coronary artery; LCx, left circumflex; RCA, right coronary artery; RVOT, right ventricular outflow tract.

these dynamic obstructions, he underwent successful trans-conal unroofing of the left main with reconstruction of RVOT using pericardium and division of the LAD myocardial bridge.

Patient 5

A 73-year-old man presents for evaluation of dyspnoea on exertion. His history is notable for CAD with anomalous RCA arising from left coronary sinus status post-coronary artery bypass graft (CABG) with RIMA–RCA in 2018, hyperlipidaemia, and poorly controlled diabetes. Per report, he had previously undergone CABG locally due to intermittent chest pain with discovery of the anomalous RCA with concern for intramural course. However, his cCTA here showed an interarterial course without intramural involvement between the PA and aortic root. He proceeded with coronary angiography and invasive testing using iFR at rest and with pharmacologic stress, achieving 100% MPHR and IVUS (Supplementary material online, Video S2). The former was notable for a 50% stenosis in the mid-RCA distal to the interarterial segment and atretic RIMA graft with otherwise mild non-obstructive disease (Figure 5). Instantaneous wave-free ratio was normal at rest 0.97 but at peak stress, iFR was mildly abnormal at

0.86 distal to the mid-RCA lesion and improved to 0.89 distal to the interarterial segment. Subsequent IVUS confirmed a moderate plaque in the mid-RCA and an elliptical luminal appearance within the interarterial segment from mild static compression of adventitial tissue. Intravascular ultrasound measurements showed a minimal luminal area of 2 mm × 4.5 mm with distal reference vessel of 4 mm × 4 mm. Through a multidisciplinary approach and risk–benefit discussion with the patient, it was ultimately felt that surgical intervention was not warranted currently and that he would benefit first from modifying coronary risk factors including poorly controlled diabetes. While he met invasive haemodynamic criteria, it was nonetheless only mildly abnormal iFR achieved at 100% MPHR without other high-risk findings such as intramural course, dynamic compression, or syncope.

Discussion

These five patients highlight how different clinical presentations and variable courses of SCA can affect prognosis and management. Given

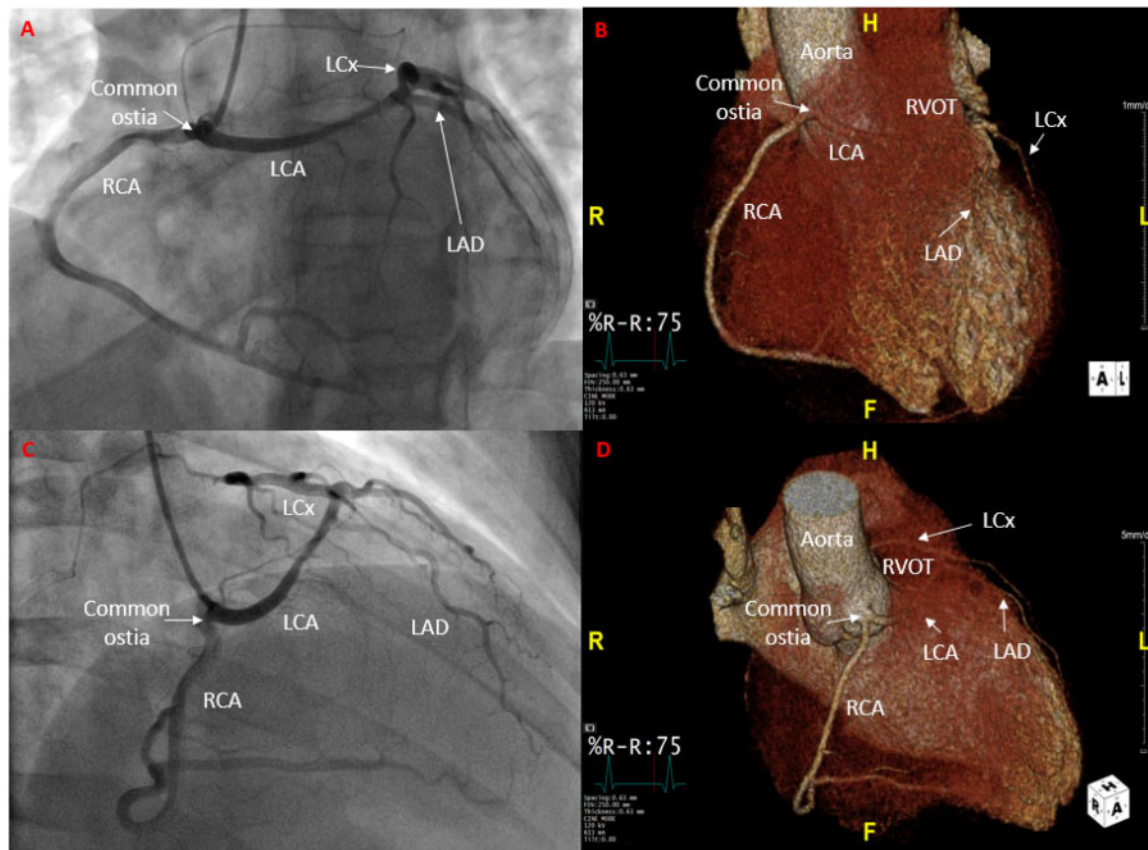


Figure 4 (A) Angiographic LAO view of single coronary artery arising from right sinus. Common trunk gives rise to right coronary artery and left coronary artery, the latter of which then takes a subpulmonic course between the aorta and right ventricular outflow tract before bifurcating left anterior descending and left circumflex branches. (B) Computed tomography representation of similar view. (C) RAO view of single coronary artery. Refer to [Supplementary material online, Video S1](#) but even on still frame, there is already a hint of trans-septal compression in the left coronary artery. (D) Computed tomography representation. LAD, left anterior descending; LCA, left coronary artery; LCx, left circumflex; RCA, right coronary artery; RVOT, right ventricular outflow tract.

the broad spectrum of coronary artery anomalies, it is important to not only define the specific anatomy but also perform invasive testing with pharmacologic stress for functional assessment. Single coronary artery is most commonly classified into Lipton Groups I, II, or III depending on the anatomical course (refer to Aldana-Sepulveda *et al.*⁶ review paper for detail discussion). A comprehensive review of AAOCA from the inappropriate sinus reported that while most coronary anomaly subtypes are benign, interarterial courses of anomalous LCA (prevalence $\sim 0.03\%$) or RCA (prevalence $\sim 0.23\%$) have been implicated with highest risk for SCD.⁷ However, given the rarity of the condition and likely under-diagnosis in the asymptomatic general population, there is a paucity of data to truly characterize the natural history of SCA variants and prognosticate their respective likelihood of SCD. In up to 40% of cases, SCA is associated with other congenital malformations such as persistent truncus arteriosus and tetralogy of Fallot.³

For anatomic assessment of coronary anomalies, cCTA and MRA are currently *Class I* imaging modalities while invasive coronary angiography and IVUS are *Class IIa* per ACC/AHA, depending on resource availabilities.⁸ Compared to MRA, cCTA is typically preferred

due to rapid scan time, high spatial resolution, and lower cost but has the disadvantages of radiation exposure and iodinated contrast use.⁷ Lipton *Group I* SCA is typically a benign finding and can be managed medically if there is any degree of CAD as in our *Patient 1*. Lipton *Group II* SCA can take a variety of courses from prepulmonic, interarterial, retroaortic, subpulmonic, to retrocardiac. Similar to *Group I*, prepulmonic (*Patient 2*), retroaortic, subpulmonic (*Patient 3*), retrocardiac are all usually benign findings, but when there is significant ischaemia as demonstrated by invasive testing leading to intraluminal compression (*Patient 4*) in the trans-septal portion of the subpulmonic course, surgical intervention may be warranted. Conversely, not all interarterial courses (*Patient 5*) require intervention based on a multidisciplinary approach including adult congenital heart disease specialist, congenital cardiothoracic surgeon, and patient him/herself.

Currently, there is no formal established guidelines for treatment of SCA, though revascularization is still recommended if there is significant atherosclerosis and/or documented ischaemia.⁶ To help guide informed decision-making, the Cleveland Clinic Foundation has established an invasive protocol for functional assessment using coronary angiography, iFR, and IVUS compared at rest to at peak stress

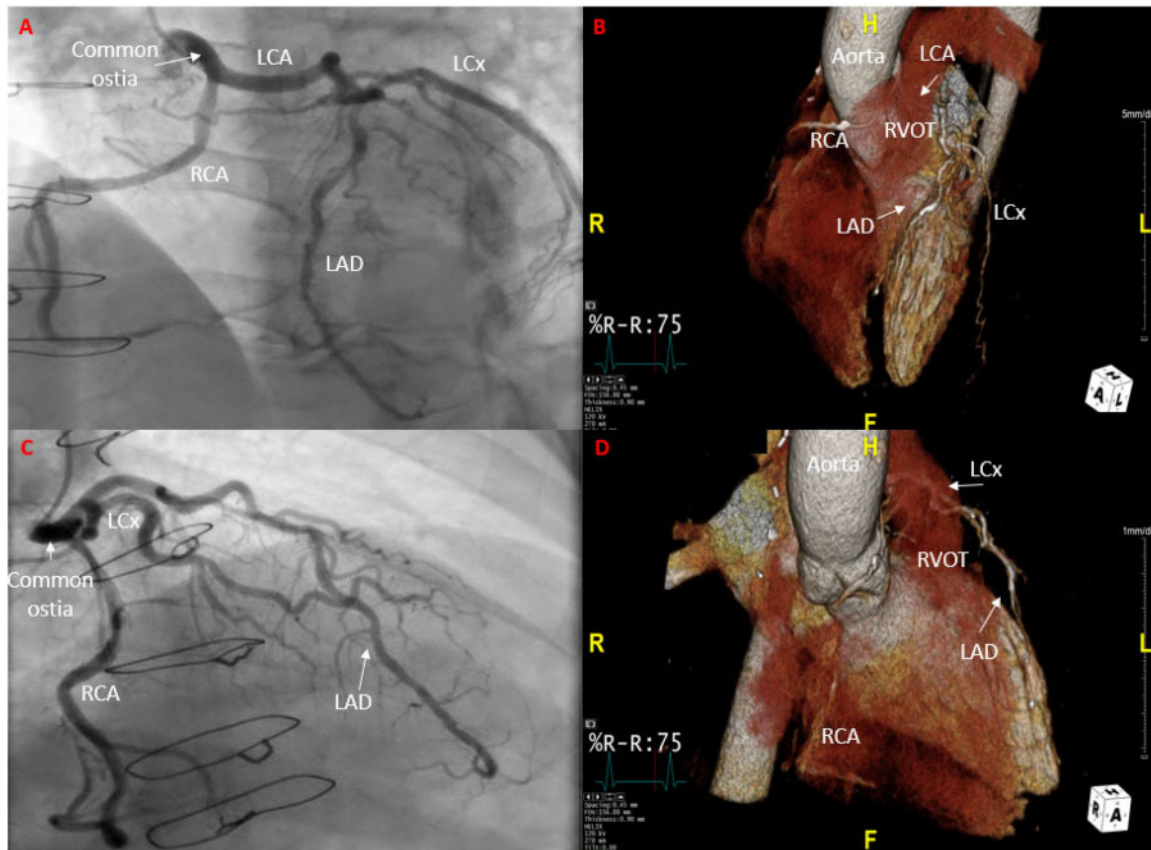
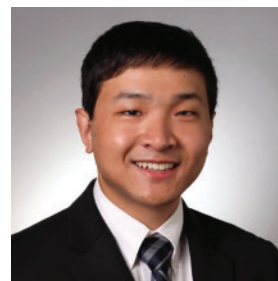


Figure 5 (A) Angiographic LAO view of single coronary artery arising from left sinus. Common trunk gives rise to left coronary artery and right coronary artery, the latter of which then takes an interarterial course between the aorta and right ventricular outflow tract. (B) Computed tomography representation of similar view. (C) RAO view of single coronary artery. (D) Computed tomography representation. LAD, left anterior descending; LCA, left coronary artery; LCx, left circumflex; RCA, right coronary artery; RVOT, right ventricular outflow tract.

using dobutamine infusion \pm atropine with targeted ≥ 80 –85% MPHR, reproducing anginal symptoms and/or achieving haemodynamic significance. As the case in *Patient 4*, while his presenting symptom of palpitations seem atypical to be directly related to his SCA anomaly, his positive iFR test for ischaemia at rest and with stress along with IVUS findings of intraluminal compression, he was offered surgical intervention. Coronary artery bypass grafting was the classic surgical approach but has fallen out of favour due to risk of early graft failure from competitive flow. For interarterial courses with intramural or trans-septal segments, there are various surgical approaches for unroofing the vessel. One novel transconal approach was described by Najm *et al.*⁹ for trans-septal anomalous LCA by unroofing the left main to free it from the compressive effects of the muscular RVOT and extending the RVOT with a pericardial patch.

Comprehensive evaluation of SCA should include an anatomic assessment as defined by cCTA or MRA followed by coronary angiography and invasive functional assessment with iFR and IVUS at rest and with stress. An individualized treatment plan can then be tailored to each patient's symptoms and overall risk profile.

Lead author biography



Dr Calvin C. Sheng is a second year general cardiology fellow at the Cleveland Clinic Foundation in Cleveland, Ohio, USA, with plans to continue training as an interventional cardiologist. He graduated from Vanderbilt University School of Medicine and completed his internal medicine residency at the Johns Hopkins Hospital.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that verbal consent for submission and publication of this case report including images and associated text has been obtained from the patient. This situation has been discussed with the editors.

Conflict of interest: None declared.

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