

Hybrid cardiac imaging-guided optimal management of right anomalous coronary artery origin from the opposite sinus with interarterial course (R-ACAOS): a case report

Cai De Jin ^{1,2}, Moo Hyun Kim ¹, *Xuan Jin ¹, and Kyungil Park ¹

¹Department of Cardiology, Dong-A University Hospital, 26 Daesingongwon-ro, Seo-gu, Busan 49201, Republic of Korea; and ²Department of Cardiology, Affiliated Hospital of Zunyi Medical University, Zunyi, Guizhou 563003, China

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Background

Determining the optimal management of right anomalous coronary artery from the opposite sinus (R-ACAOS) with an interarterial course (IAC) in middle-aged adults remains elusive. Hybrid cardiac imaging combining non-invasive and invasive approaches to identify high-risk anatomic features, as well as functional testing to assess potential ischaemic status by dynamic compression, can guide therapeutic decisions.

Case summary

A 65-year-old female was newly diagnosed with R-ACAOS with IAC, accompanied by suspected angina and two syncope episodes. She was initially considered as non-specific chest pain based on negative treadmill test (TMT) taken 10 years earlier. An anomaly of R-ACAOS with IAC travelling between the aorta and pulmonary artery was detected by coronary computed tomography angiography with a severe stenosis at the ostium, but with little evidence of atherosclerotic plaque. Exercise test (TMT) and single-photon emission computed tomography (SPECT) results were negative. Invasive imaging revealed a luminal area stenosis of 45% at the ostial right coronary artery, and a slit-like orifice anatomical feature, with a minimal lumen area of 5.81 mm² at diastole determined by intravascular ultrasound. Based on hybrid cardiac imaging results and previous data from a case series, conservative management was recommended with strenuous exercise restrictions. The patient fared well during 12 months of follow-up after discharge.

Discussion

Hybrid cardiac imaging-guided conservative management including exercise restriction appears justifiable in such middle-aged adults with R-ACAOS accompanied by suspected angina in absence of ischaemia in stress-induced tests (TMT or SPECT), despite high-risk anatomical features of an IAC and slit-like orifice being present.

Keywords

Hybrid cardiac imaging • Anomalous coronary artery origin • Interarterial course • Case report

* Corresponding author. Tel: +82 51 240 2976, Fax: +82 51 255 2177, Email: kimmh@dau.ac.kr

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Learning points

- Hybrid cardiac imaging derived anatomical and functional information may assist in managing a right anomalous coronary artery from the opposite sinus (R-ACAOS) with interarterial course (IAC).
- Conservative management seems justifiable for R-ACAOS (IAC) with negative stress-induced ischaemic test (treadmill test or single-photon emission computed tomography) results, as well as relatively preserved luminal area of intravascular ultrasound criteria, despite a slit-like orifice feature is present.

Introduction

Malignant anomalous coronary artery origin from the opposite sinus (ACAOS) with interarterial course (IAC, a course between the pulmonary trunk and aorta) in young athletes is a potential underlying cause of sudden cardiac death (SCD).¹ Other high-risk anatomical features including slit-like ostium, acute take-off angle, and intramural course are widely known to increase the risk of adverse cardiac events.² In middle-aged and older adults with suspected coronary artery disease undergoing coronary computed tomography angiography (CCTA), the prevalence of newly and incidentally diagnosed ACAOS with IAC has been reported at 0.64%.³ The recommended management approach varies based on the presence or absence of ischaemic evidence, including symptoms, anatomical features, and stress-induced ischaemic test results. Here, the authors present a case of hybrid cardiac imaging-guided optimal management of anomalous right coronary artery (RCA) origin from the left coronary sinus (R-ACAOS) with IAC in a middle-aged adult who presented with suspected angina symptoms.

Timeline

31 December 2009	Non-specific chest pain, syncope, initial evaluations including treadmill test (TMT) were negative
12 November 2019	Right anomalous coronary artery from the opposite sinus with interarterial course was confirmed by coronary computed tomography angiography at a different hospital
23 December 2019	Referred to outpatient clinic for suspected angina and syncope. Brain computed tomography was normal. No ventricular arrhythmias or abnormal findings were detected by Holter monitoring
6 January 2020	Admission to Dong-A University Hospital. The physical examination, cardiac

Continued

7 January 2020	biomarkers, electrocardiogram, and echocardiogram were unremarkable No stress-induced ischaemic evidence on negative results of TMT and single-photon emission computed tomography
8 January 2020	Invasive cardiac imaging revealed a luminal area stenosis of 45% at the ostial right coronary artery, and slit-like orifice anatomical features, with a minimal lumen area of 5.81 mm ² at diastole on intravascular ultrasound
9 January 2020	Patient was discharged with avoiding strenuous exercise. Beta-blocker was prescribed
22 December 2020	No further complaints during follow-up

Case presentation

A 65-year-old female presented with suspected angina symptoms and two syncope episodes. She had a history of hypertension and dyslipidaemia with beta-blocker and statin treatment, and was initially considered as having non-specific chest pain based on a negative treadmill test (TMT) result 10 years ago. Recently, she complained of resting chest pain for 2 min at night, and was newly diagnosed as R-ACAOS with an IAC (a vessel traverse between the pulmonary artery and aorta) with severe stenosis at the ostium by CCTA and was referred from a different hospital (*Figure 1*). The physical examination, cardiac biomarkers, electrocardiogram, and echocardiogram results were unremarkable. There was no stress-induced ischaemic ST-segment change or arrhythmia on TMT, and no regional myocardial perfusion defect on single-photon emission computed tomography (SPECT). Subsequent coronary angiography showed a normal left coronary artery (LCA) and a moderate stenosis at the ostial RCA (*Figure 2*). In detail (*Supplementary material online*), a JL 3.5 guide catheter (GC) was kept afloat near ostium of RCA, one guidewire was initially inserted into LCA to stabilize the GC. Then the second guidewire with floating wire technique was smoothly inserted into distal RCA for additional intravascular ultrasound (IVUS) study, and the GC was non-selective engagement in order to check the features of proximal to ostial RCA by IVUS. Subsequently, elliptical stenosis with slit-like orifice features with a minimal lumen area of 5.81 mm² and stenosis of 45% in diastole was detected by IVUS (*Figure 3*). Based on hybrid cardiac imaging (*Table 1*) and previous data from case series,⁴ conservative management (e.g. beta-blocker, nicorandil, and statin) including exercise restriction was recommended. In addition, the patient, who previously suffered from syncope, characterized by sudden onset, short duration, and spontaneous complete recovery during the meal, after bath and resting state, was considered as vasovagal syncope due to dizziness caused by a postural hypotension without any determinable reason during index hospitalization, and haemodynamic stability was recovered by saline infusion. The patient had no complaint at 12-month follow-ups.

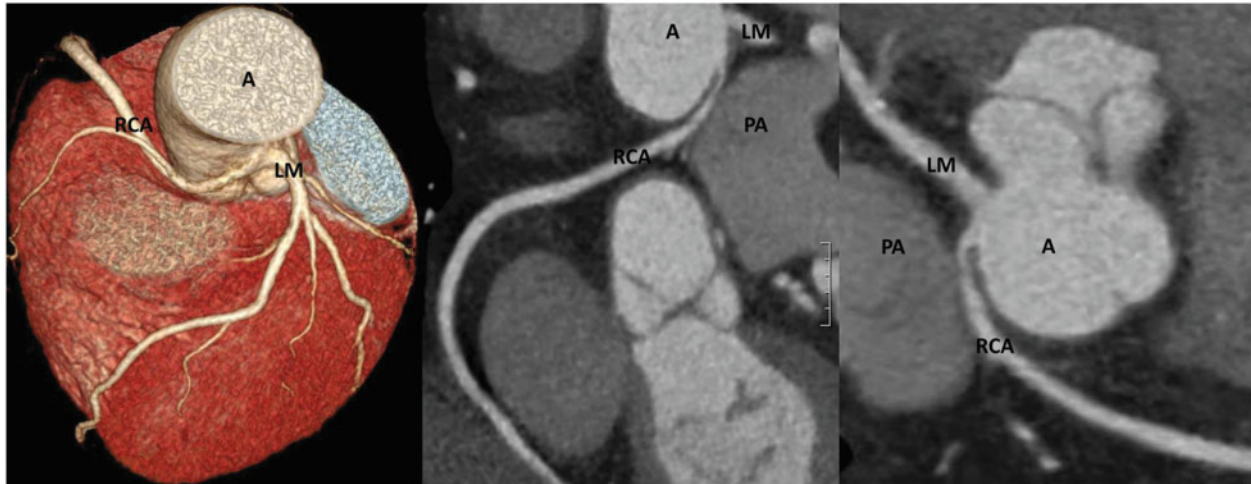


Figure 1 Three-dimensional constructed computed tomography imaging acquisition of the heart on volume rendering view showed right coronary artery origin from the left sinus (left). Curved multi planner reformation view showed right coronary artery anomalous at the proximal segment traverse between the pulmonary artery and aorta, and severe stenosis at the ostial right coronary artery (middle, right). A, aorta; LM, left main; PA, pulmonary artery; RCA, right coronary artery.

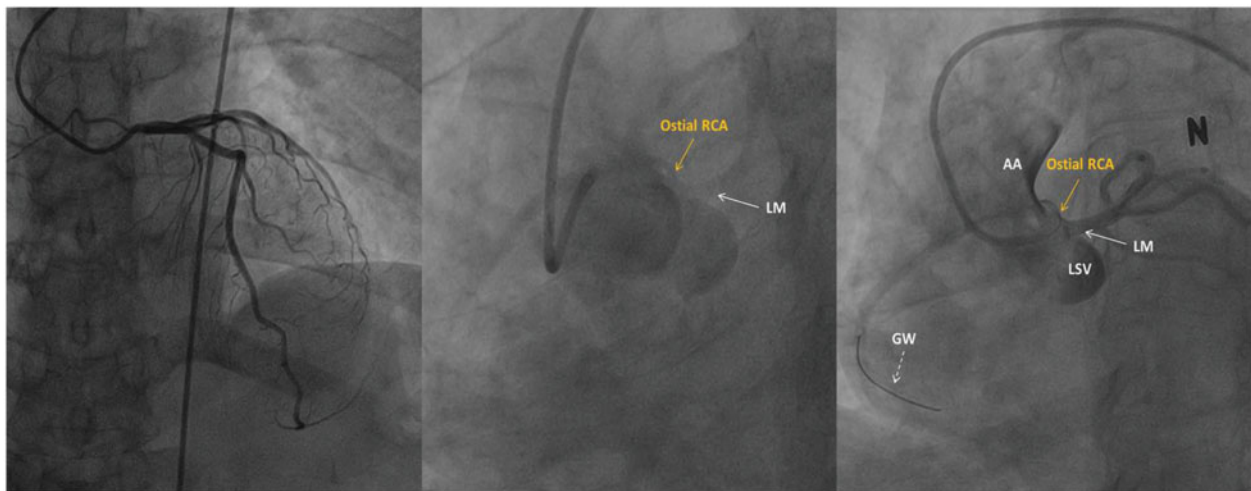


Figure 2 The patient's left coronary angiogram was normal (left). Non-selective right coronary angiogram revealed right coronary artery of anomalous origin from left sinus of Valsalva, a moderate stenosis by visual assessment (middle). Angiography was assisted with guidewire (Runthrough NS, Terumo, Japan) anchoring (right). Yellow arrows indicate ostial right coronary artery; white arrows indicate left main; dotted arrow indicates guidewire. N indicates intracoronary nitrate injection. AA, ascending aorta; GW, guidewire; LM, left main; LSV, left sinus of Valsalva; RCA, right coronary artery.

Discussion

Malignant ACAOS with IAC is rare, but may cause complaints and increase the risk of SCD in young individuals, as anomalous vessels are prone to dynamic compression during physical exercise. Outcomes are also dependent on the presence of anatomical high-risk features such as slit-like ostium.² However, more middle-aged and older adults with these anomalies are found incidentally with increasing use

of CCTA to evaluate symptomatic patients,⁵ gaining better prognoses during 49 months follow-up, and adverse outcomes have not been statistically different to matched controls without coronary artery anomalies regardless of whether the ACAOS occurs in the presence or absence of IAC variants.⁶ A retrospective observation in this patient with ~10 years of clinical outcomes was favourable, despite high-risk anatomic features including an IAC and slit-like orifice being present.

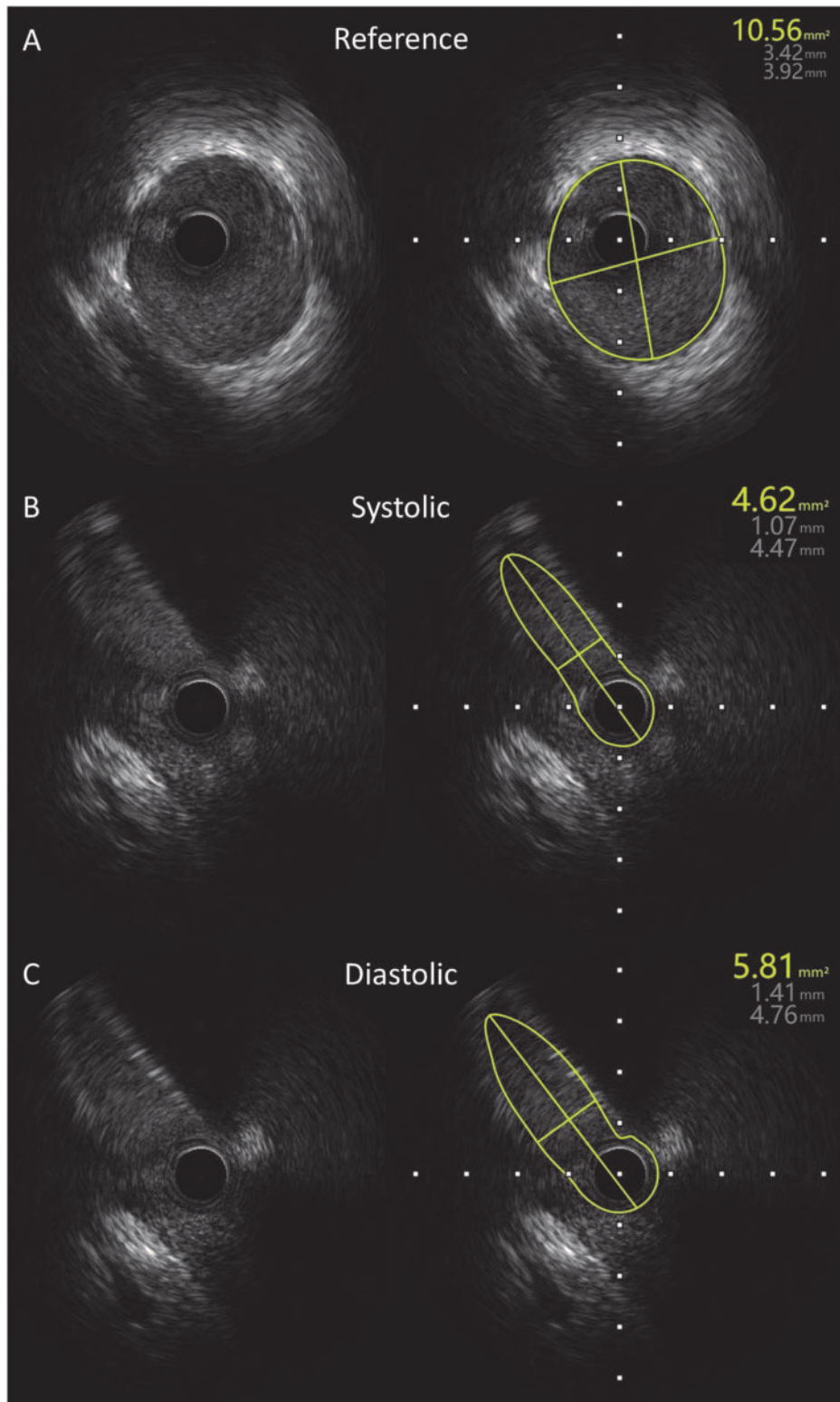


Figure 3 Intravascular ultrasound imaging acquisition from the proximal to ostial segment of the right coronary artery. The reference and minimal lumen diameter (D) and cross-sectional area were determined using Boston Scientific Image Viewer software. (A) Normal vessel without plaque burden at the distal reference ($D = 3.42/3.92$ mm, cross-sectional area = 10.56 mm²). (B) Slit-like orifice anatomic features close to the ostium at systolic phase ($D = 1.07/4.47$ mm, cross-sectional area = 4.62 mm², stenosis = 56%) and (C) at diastolic phase ($D = 1.41/4.76$ mm, cross-sectional area = 5.81 mm², stenosis = 45%). Interarterial segment length is 17.18 mm. Percent lumen area stenosis of anomalous coronary region determined by formula $(RA - SA)/RA \times 100\%$, where RA is the reference area and SA is the stenosis area.

Table 1 Hybrid cardiac imaging assessment of anatomical and functional significance in our patient diagnosed right anomalous coronary artery from the opposite sinus with interarterial course

Cardiac imaging	Anatomical significance	Functional significance
CCTA	Severe stenosis	No visible perfusion defect, no myocardial thinning
SPECT	—	No myocardial perfusion defect, preserved wall motion
CAG	Moderate stenosis	—
IVUS	Elliptical stenosis, slit-like orifice MLA: 5.81 mm ² at diastole (area stenosis = 45%)	Dobutamine stress FFR levels ≤ 0.80 showed area stenosis $>50\%$ by IVUS ⁴

The surgical correction is suggested if in the presence of functional significance and high-risk feature (slit-like orifice), and conservative management is recommended if in the absence of those.

CAG, coronary angiography; CCTA, coronary computed tomography angiography; FFR, fractional flow reserve; IVUS, intravascular ultrasound; MLA, minimal lumen area; SPECT, single-photon emission computed tomography.

To assess the risk estimation is crucial, and treatment options are difficult as there is no uniform way to stratify such patients particularly in the absence of complaints or atypical ones. In general, treatment options for patients with ACAOS and IAC include surgical or percutaneous correction and exercise restriction in those presenting with symptoms such as chest pain, syncope, ventricular arrhythmia, and/or the presence of ischaemia on stress testing.⁷ However, surgical treatment is recommended for patients with LCA originating from the right sinus, regardless of the symptoms or evidence of ischaemia. A treatment dilemma occurs when patients presented R-ACAOS with IAC and negative stress test. In fact, stress testing is difficult to assess as it would likely fail to detect any right ventricular ischaemia.⁵ Therefore, multi-modality cardiac imaging combined with stress-induced ischaemic test is required, which can help to guide optimal management.

Driesen *et al.*⁸ recommend that IVUS and fractional flow reserve (FFR) guided decision-making be followed as a standard work-up for middle-aged adults presenting with ACAOS with an IAC. The surgical correction was suggested if at least two of FFR ≤ 0.8 , slit-like orifice on IVUS and typical angina symptoms. Consistent with Korean data focusing on FFR-guided treatment for R-ACAOS with IAC, the authors suggested that more conservative strategies be considered in adults unless definitive myocardial ischaemia is present.⁴

Stenting in an IAC segment with luminal narrowing and ischaemic burden is challenging. First, the procedure offers technical complexity due to difficult cannulation, and less backup support for guiding correct stent placement. Second, the stent type selection (drug eluting or bare metal) and degree of dilation can be problematic.^{9,10} Third, in-stent restenosis can be more frequent in such cases due to phasic compression forces, since metal fatigue in muscular bridges, stent

fracture limits the efficacy of drug-eluting stents,¹¹ which are anatomically close to the ACAOS situation. There is currently insufficient data available on stenting for ACAOS with IAC.

Previous case series reported a 6-month clinical outcome as favourable after coronary bare metal stenting for R-ACAOS with IAC and slit-like orifice at the ostium, including nine patients aged 44–70 years presenting with myocardial ischaemia caused by anomalous variance. Follow-up angiography after stenting in IAC segment showed stent patency, and non-invasive testing in all patients was normal, suggested stenting may be an acceptable alternative to surgery.⁹ In another rare case that reported successful outcomes, a cypher stent implantation was completed across the IAC segment of R-ACAOS in a middle-aged adult presenting ischaemic symptoms that was not associated with obstructive coronary artery disease. The patient had a favourable outcome during 37 months follow-up, reporting symptom-free and exercise-free outcomes, as well as stent patency determined by CCTA scan at 12 months, while stress myocardial perfusion imaging was negative at 37 months. This suggests that stenting could offer an alternative if patients refuse surgical treatment.¹² Furthermore, 2017 expert consensus suggests that coronary intervention with an IVUS assistant may be considered in selected adults, specifically in R-ACAOS with a partial intramural course and proximal intramural stenosis.¹³

Conservative treatment for normal SPECT and atypical symptoms was undertaken for our patient, despite the slit-like feature being present. The functional dynamic test results in stress-induced ischaemia were normal, and IVUS-derived minimal lumen area was 5.81 mm², and area stenosis was 45% at diastolic phase. Additionally, recent data indicate that SPECT and FFR are significantly similar for the detection of myocardial ischaemia.¹⁴ Thus, conservative management seems reasonable for middle-aged or older adults who present with normal stress-induced ischaemic test results and atypical symptoms, despite an IAC and slit-like orifice being present.

Conclusion

Hybrid cardiac imaging (including CCTA, SPECT, and IVUS) in clinical practice may contribute to optimal guidance for the management of R-ACAOS with IAC in middle-aged adults presenting with suspected angina symptoms.

Lead author biography



Dr Cai De Jin was born in Wangqing, Yanbian Korean Autonomous Prefecture, Jilin, China. He is a physician in the Department of Cardiology, who received his Doctoral degree in Medicine in 2016 and completed his post-doctoral Fellowship in 2017 from Dong-A University (Republic of Korea), and finished coronary interventional training under Prof. Moo Hyun Kim. His

interests include platelet research, cardiac imaging modalities, and coronary intervention.

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 author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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