

CASE REPORT

INTERMEDIATE

CLINICAL CASE SERIES

Novel Assessment of Ischemia in Patients With Anomalous Right Coronary Artery



Lerin C. McCray, MD,^{a,b} Delvise T. Fogwe, BSN,^c Kul Aggarwal, MD,^{a,b} Poorna R. Karuparthi, MD^{a,b}

ABSTRACT

Anomalous right coronary arteries are usually benign; however, sudden death owing to myocardial ischemia, especially during exertion, have been reported in patients with intramural or interarterial course, which is likely due to dynamic obstruction. We propose a novel method of physiological evaluation with instantaneous wave-free ratio with dobutamine infusion to simulate controlled dynamic obstruction in anomalous right coronary arteries.

(Level of Difficulty: Intermediate.) (J Am Coll Cardiol Case Rep 2019;1:819-22) Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Anomalous right coronary arteries (ARCAs) account for 0.17% of all anomalous coronary arteries (1,2). Although the majority of ARCA are usually benign and diagnosed incidentally during coronary angiography, sudden death, or myocardial ischemia with exertion has been reported, especially with intramural (through the aorta wall) or interarterial (between aorta and pulmonary trunk) course, which could cause dynamic obstruction during exertion (1,3). Physiological evaluation of dynamic obstruction with ARCAs has not been established.

We present a case series of 2 patients with ARCAs, who underwent evaluation in the cardiac catheterization lab using instantaneous wave-free ratio (iFR) with dobutamine and atropine administration.

LEARNING OBJECTIVES

- To be able to assess ischemia in patients with coronary anomalies that have inconclusive stress testing.
- To expand the role iFR in patients with dynamic obstruction with dobutamine and atropine administration.

PATIENT #1

Patient #1 was a 59-year-old woman with history of coronary artery disease, hypertension, and hyperlipidemia with recurring chest pain despite recent coronary angiography showing nonobstructive lesions. She reported chest pain daily with activity and house cleaning. She described the pain as left chest pressure radiating up to the left neck and down the left arm.

DIAGNOSTIC WORK-UP. Multislice cardiac computed tomography (CT) confirmed the anomalous right coronary artery from the left coronary cusp with interarterial course (Figure 1). Pharmacologic stress testing was suboptimal, as the target heart rate was not reached due to chest pain.

INTERVENTION. iFR was obtained, using Philips Verrata, at baseline and again after achieving a target heart rate of 137 beats/min using dobutamine and atropine administration. Dobutamine infusion was started as a 10 µg/min infusion and increased by 10 µg/min every 3 min to a maximum of 30 µg/min, and 0.25 mg of atropine was also administered to achieve target heart rate.

From the ^aDivision of Cardiology, Department of Medicine, Harry S. Truman VA Hospital, Columbia, Missouri; ^bDivision of Cardiology, Department of Medicine, University of Missouri, Columbia, Missouri; and the ^cSchool of Medicine, University of Missouri, Columbia, Missouri. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Informed consent was obtained for this case.

Manuscript received September 27, 2019; revised manuscript received November 3, 2019, accepted November 4, 2019.

ABBREVIATIONS AND ACRONYMS

ARCA = anomalous right coronary artery

CT = computed tomography

FFR = fractional flow reserve

iFR = instantaneous wave-free ratio

IVUS = intravascular ultrasound

OUTCOME. Baseline iFR was hemodynamically nonsignificant at 0.96. At the target heart rate, iFR became hemodynamically significant at 0.83 (Figure 2). The patient tolerated the dobutamine infusion and atropine administration well. No complications noted at the end of the procedure and patient was discharged the same day.

PATIENT #2

Patient #2 was a 60-year-old man with a history of severe chronic obstructive pulmonary disease (on 3 l of oxygen), hypertension, and dyslipidemia. Initially, his only complaint was worsening dyspnea at rest and with exertion, but he later described a few episodes of sharp, nonradiating chest pain lasting for a few minutes, which would resolve spontaneously. Cardiac catheterization revealed nonobstructive coronary artery disease and an ARCA. The cardiothoracic team recommended medical therapy in view of elevated risk with coronary artery bypass grafting. Due to persistent chest pain episodes while on maximal medical therapy, further evaluation of ARCAs was planned.

DIAGNOSTIC WORK-UP. Multislice cardiac CT confirmed the ARCA from the left coronary cusp with interarterial course (Figure 3).

INTERVENTION. iFR was obtained at baseline and again after achieving target heart rate (142 beats/min) using dobutamine and atropine administration. dobutamine infusion was started 10 µg/min infusion, increased by 10 µg/min every 3 min to a maximum of 50 µg/min; 0.5 mg

of atropine was administered to achieve target heart rate.

Intravascular ultrasound (IVUS) interrogation of the ostial and proximal segments of the ARCA was performed, which revealed evidence of slit like opening and external compression of the ARCA at ostial and proximal segments.

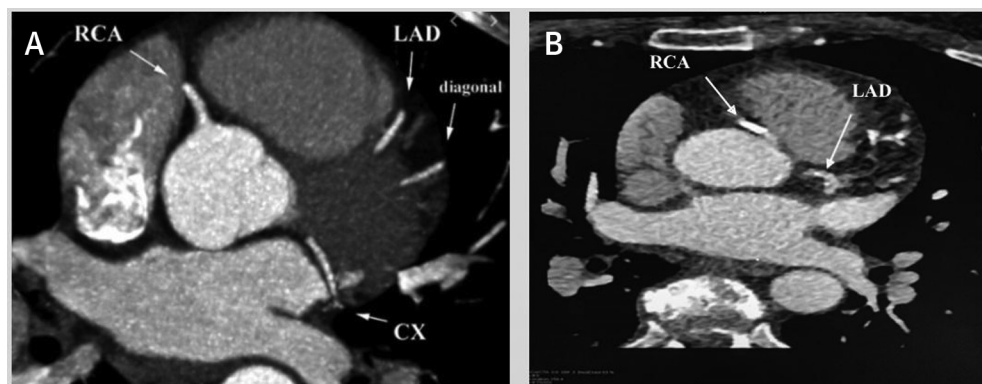
OUTCOME. Baseline iFR was hemodynamically nonsignificant at 0.99. At target heart rate, iFR was hemodynamically significant at 0.77 (Figure 4). No complications were noted at the end of the procedure and patient was discharged the same day.

DISCUSSION

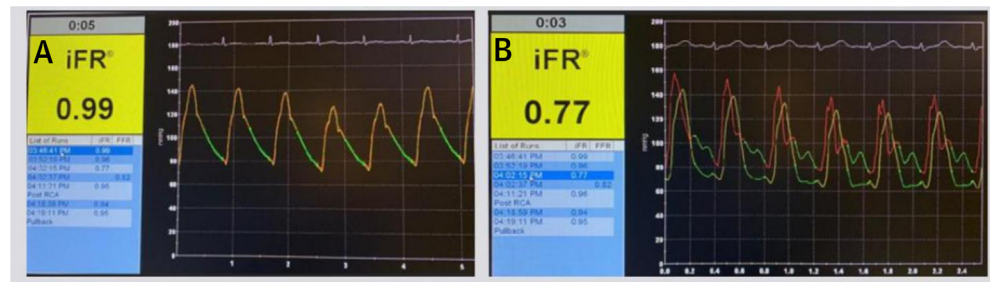
A diagnosis of ARCA can be easily made with coronary angiography or with multislice coronary CT scan, which allows 3-dimensional visualization of the course of the coronary artery (1,3). However, physiological assessment of anomalous coronary artery with interarterial course could be challenging, partly due to the dynamic nature of obstruction. Studies have shown evidence of intermittent ischemia in association with increased aortic wall tension and higher cardiac output with activity (4).

According to the American College of Cardiology/American Heart Association guidelines for patients with an anomalous left main artery originating from right coronary cusp, surgical intervention is favored (Class I recommendation for symptomatic patients and Class IIa for those without symptoms). For ARCAs with an interarterial course, surgical coronary revascularization is only recommended if symptoms or diagnostic evidence of ischemia related to the anomaly are present (2).

FIGURE 1 CT of the Coronary Artery of Patient #1



(A) Chest computed tomography (CT) demonstrating normal origin of coronary arteries (10). (B) Chest CT demonstrating anomalous right coronary artery (RCA) with interarterial coursing (previous stent place in the RCA can also be noted). CX = circumflex artery; LAD = left anterior descending artery.

FIGURE 4 iFR at Baseline and After Target Heart Rate of Patient #2

(A) Baseline instantaneous wave-free ratio (iFR) before dobutamine infusion, hemodynamically nonsignificant. (B) iFR showing significant dynamic stenosis post infusion of dobutamine and achievement of target heart rate.

dobutamine infusion but not at baseline or with adenosine administration (8).

FFR with dobutamine infusion at target heart rate did not show consistent results in patients with dynamic obstruction, such as myocardial bridge, owing to overshooting of systolic pressure and negative systolic pressure gradient across the lesion (9). Diastolic FFR instead of a whole-cycle FFR is proposed to have favorable results with dynamic obstruction but is cumbersome to calculate (9).

We instead used the diastolic pressure gradient study iFR with dobutamine and atropine to evaluate hemodynamic significance of ARCA. In both cases, baseline iFR values were hemodynamically nonsignificant, which became hemodynamically significant at the target heart rate (after dobutamine infusion), simulating peak exertion. IVUS interrogation with patient 2 did confirm evidence of external compression.

Given the positive results noted in our patients, we propose the use of iFR with dobutamine and atropine

infusion as a novel method of evaluating the physiological significance of anomalous interarterial course while inducing controlled dynamic obstruction. This approach may allow further risk stratification of patients with ARCAs before considering surgical coronary revascularization, as the management plan in these patients is not well established.

CONCLUSIONS

iFR interrogation with dobutamine and atropine administration serves as a promising modality to interrogate dynamic stenosis with ARCAs. Further studies are needed to establish safety and effectiveness of the proposed strategy for evaluation of ARCAs.

ADDRESS FOR CORRESPONDENCE: Dr. Poorna R. Karuparthi, Harry S. Truman VA Hospital, 800 Hospital Drive, Columbia, Missouri 65201. E-mail: rkaruparthi@yahoo.com.

REFERENCES

- Pleva L, Jonsza T, Kukla P. Congenital coronary anomalies. *Cor et Vasa* 2014;56:e27-36.
- Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 ACC/AHA Guideline for the management of adults with congenital heart disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation* 2019; 139:e698-800.
- Das A, Joseph A, Jolly N. Stress test to STEMI: Utility of coronary CTA in the diagnosis and management of anomalous right coronary artery from the left coronary cusp. *Echocardiography* 2017;34: 1519-23.
- Lee SE, Koo BK, Yu CW, et al. Physiologic assessment of anomalous origin of right coronary artery from left coronary cusp using dobutamine stress fractional flow reserve. *J Am Coll Cardiol* 2012;59 Suppl:171-9.
- Lee BY. Anomalous right coronary artery from the left coronary sinus with an interarterial course: is it really dangerous? *Korean Circ J* 2009;39: 175-9.
- Davies JE, Sen S, Dehbi HM, et al. Use of the instantaneous wave-free ratio or fractional flow reserve in PCI. *N Engl J Med* 2017;376: 1824-34.
- Angelini P, Walmsley RP, Liberos A, Ott DA. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of Val-salva. Clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J* 2006;33:171-9.
- Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation* 2007;115: 1296-305.
- Escaned J, Cortés J, Flores A, et al. Importance of diastolic fractional flow reserve and dobut-amine challenge in physiologic assessment of myocardial bridging. *J Am Coll Cardiol* 2003;42: 226-33.
- CTsurgery.com. Normal coronary CT. Available at: <https://www.ctsurgery.com/coronary-anatomy.html>. Accessed October 28, 2019.

KEY WORDS anomalous right coronary artery, dynamic coronary obstruction, iFR, instantaneous wave-free ratio