JACC: CASE REPORTS VOL. 4, NO. 7, 2022

EDITORIAL COMMENT

A Road Less Traveled*

Maurizzio Taramasso, MD, PhD,[a] Saad S. Ahmad, MD[b]

In this issue of *JACC: Case Reports,* Bouaouina et al[1] describe a patients with stable angina whose work-up revealed a coronary artery-to-pulmonary artery fistula from the left anterior descending (LAD) artery. To confirm the ischemic burden by steal phenomenon, the preintervention fractional flow reserve (FFR) was confirmed to be 0.8. The closure method described by the group used a liquid embolic agent. After closure of the fistula, the FFR in the LAD territory improved to 0.95, thus providing confirmation of the hemodynamic significance of the fistula. Bouaouina et al[1] also highlight other noninvasive and invasive modalities used to evaluate the hemodynamic significance of a coronary artery fistula (CAF).

CAF traditionally has been identified as a rare incidental finding on coronary angiography, with an incidence of 0.1% to 0.2%.[2,3] The appreciated incidence by coronary computed tomography (CT) angiogram is rising with increased use of the modality and has been reported to be 0.3% to 0.9%, with coronary artery-to-pulmonary artery fistula the most common CAF type.[4,5] CAFs have been associated with angina, dyspnea, arrythmia, possible heart failure, and endocarditis.[6]

The effect of a CAF is determined by its size, the origin of the fistula, and the distal segment associated with it. CAFs have been associated with increased flow across the fistula, thus resulting in patients presenting with angina secondary to steal phenomenon.[7,8] Sometimes the assessment is not definitive and the contribution of the symptoms that are the result of the angina is difficult to discern Published reports show that the hemodynamic significance of the fistula has been investigated by various modalities. Elevated Doppler flow velocities and depressed coronary reserve have been demonstrated in symptomatic patients before fistula closure.[8] Ischemic burden was assessed with 13N-labeled ammonia positron emission tomography in a series of 11 patients, demonstrated only one showed flow abnormality.[9] FFR assessment along the donor vessel has also been used as a means of assessing ischemic burden in the region supplied by the donor vessel of the fistula.[10] FFR has been used to establish the significance of a lesion by calculating the ratio of pressure drop across it relative to the aortic pressure. FFR has been validated and studied across multiple randomized controlled trials to guide the appropriateness of intervention. By current guidelines, treating a lesion when the FFR is ratio <0.8 reduces events of urgent revascularization driven by ischemia.

If it is deemed appropriate to close the fistula, the options include surgical closure, coil embolization, and use of a vascular plug. The outcomes of the surgical cases have been reportedly poorer than the percutaneous procedure outcomes, and it is postulated that there may be a selection bias.

As mentioned in the case report by Bouaouina et al,[1] in 2008 the adult congenital heart disease guidelines did mention consideration for closure of a CAF. However, in the 2018 guidelines, the discussion of fistula management was refined to include a caution that increased mortality and an increased risk of myocardial infarction are noted with surgical closure and to emphasize the need to individualize patient care with a team-based approach.[11] After closure of a large-flow fistula, the donor vessel develops a risk for sluggish flow, and thus thrombosis and infarction may occur.

CAF is a rare finding that may be noted incidentally during imaging. Unlike the data that have led to guideline recommendations for using FFR in

*Editorials published in *JACC: Case Reports* reflect the views of the author and do not necessarily represent the views of *JACC: Case Reports* or the American College of Cardiology.

From the [a]Heart Center Hirslanden Zurich, Zurich, Switzerland; and the [b]Division of Cardiovascular Disease, University of Cincinnati, Cincinnati, Ohio, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2021.12.026

JACC: CASE REPORTS, VOL. 4, NO. 7, 2022
APRIL 6, 2022:395–396

coronary artery disease, there is a lack of registry or randomized controlled trial data to allow for incorporation of the management into current guidelines. Just as a large randomized controlled trial reversed the routine practice of thrombectomy in ST-segment elevation myocardial infarction, it is always important to realize that studies with a single treatment arm and studies with small numbers of participants may obscure the pitfalls on the road to treatment for the patient.[12]

Just as the DEFER[13] trial was a pivotal study for the evaluation and management of coronary artery disease, patients with CAF would benefit from a multicenter study with longitudinal follow-up on the basis of treatment options including medical management. A heart team approach is critical for all cardiac patients, but it becomes more important in areas of the field where there is a paucity of data and the interest of the individual becomes the guiding principle.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr Taramasso has consulted for Abbott, Edwards Lifesciences, Boston Scientific, Shenqi Medical, Medira, 4tech, Simulands, MTeX, Occlufit, and CoreMedic; and has received consulting fees from Cardiovalve. Dr Ahmad has is on the Speakers Bureau for AstraZeneca and Pfizer.

ADDRESS FOR CORRESPONDENCE: Dr Maurizio Taramasso, Clinic of Cardiac Surgery, HerzZentrum Hirslanden Zurich, Witellikerstrasse 36, Zurich 8008, Switzerland. E-mail: maurizio.taramasso@usz.ch. Twitter: @m_taramasso.

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KEY WORDS coronary vessel anomaly, fractional flow reserve, percutaneous coronary intervention