



EDITORIAL COMMENT

Coronary artery fistulas: A Portuguese single-center experience

Fístulas coronárias - experiência de centro único português

Sérgio Madeira

Centro Hospitalar de Lisboa Ocidental, Hospital de Santa Cruz, Carnaxide, Portugal

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Coronary artery fistulas (CAFs) are congenital or acquired aberrant connections between the coronary arteries and contiguous structures such as non-coronary vessels or cardiac chambers. The estimated prevalence of CAFs in invasive coronary angiography (ICA) registries is between 0.05% and 0.8%,^{1–3} however non-invasive coronary computed tomography angiography registries suggest that it may be higher, with a prevalence of around 0.9%.⁴ CAFs are usually found incidentally during a study for other reasons,⁵ however depending on the anatomy (donor vessel, recipient vessel or chamber, size and length) and patient characteristics (such as concomitant heart disease), they can cause symptoms and lead to life-threatening clinical scenarios including acute or chronic ischemia, congestive heart failure (CHF), pulmonary hypertension (PH), endarteritis and rupture.^{5,6} Usually small incidentally found CAFs are asymptomatic and do not require intervention, but should be followed up clinically and with echocardiography every 3–5 years.^{5,7} Some CAFs may remain clinically silent for decades, but once they become hemodynamically significant (myocardial ischemia through the coronary steal phenomenon, left ventricular volume overload or significant left-to-right shunt leading to CHF or PH), treatment must be considered.^{1,5} Exceptions are made for pediatric patients, in whom the threshold for intervention may be lower.⁸ Percutaneous treatment is currently the mainstay of therapy when technically feasible and when there are

no other indications for heart surgery.^{1,5} Patients who have undergone intervention should be followed more frequently early after closure to detect possible recurrence of the fistula, persistent dilatation of the coronary artery, thrombus formation, calcification, arrhythmias, and myocardial infarction.^{1,5} Regarding medical therapy, antiplatelet therapy is recommended empirically to prevent thrombosis when there is coronary artery dilatation, antianginal drugs should be used to alleviate angina, and most authors recommend endocarditis prophylaxis.

In this issue of the *Journal*, Torres et al.⁹ report the clinical manifestation, anatomical characteristics and management of 55 patients diagnosed as having at least one CAF among 32 174 ICAs over a period of 12 years in a single tertiary center. This is the largest Portuguese CAF series ever reported, and although the study population is heterogeneous, including pediatric patients, patients with known congenital heart disease and those with previous heart interventions, overall the findings regarding anatomy and clinical manifestations are in line with other international series and management generally reflects current trends.

Excluding pediatric patients and those with congenital heart disease, most of the adult patients referred for ICA for common indications presented with a non-significant bystander CAF, not related to the symptoms or clinical condition. Fifteen patients (~0.05%) underwent treatment. Three pediatric patients with CAFs deemed to be significant in terms of size or shunt magnitude were treated by transcatheter closure irrespective of symptoms (n=3). Six adult patients – four with aortic valve disease, one with

E-mail address: serg.lou.madeira@gmail.com

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stable three-vessel disease, and one with two-vessel disease including left anterior descending disease presenting with non-ST-elevation myocardial infarction (NSTEMI) – had incidental CAFs that were surgically ligated at the time of aortic valve replacement or coronary artery bypass surgery. Finally, only six adults (~0.02%) had a significant CAF that was responsible for their symptoms and/or was the main underlying pathophysiological mechanism for the cardiac abnormality. Of these, two had right chamber volume overload due to left-to-right shunt at the level of the right atrium, three had either chest pain or evidence of ischemia due to a presumed coronary steal syndrome, and one presented with NSTEMI and cardiac arrest. All these patients were treated by transcatheter closure, except one in whom percutaneous treatment was not technically feasible.

Several types of transcatheter devices were used at the discretion of the operators in order to best match the anatomy for complete closure. Four patients had no or trivial residual flow after the procedure (50%), three had a small residual flow and one had a large residual flow. Those with more than trivial residual flow underwent repeat angiography within six months of the procedure; two had no residual flow, and two still presented a small residual flow. One of the latter required reintervention with implantation of a different device, and the other showed complete closure at one-year angiographic follow-up. There was one procedure-related myocardial infarction and pericarditis in the same patient (complication rate 12.5%). Overall, complete transcatheter closure without complications or reintervention at one year occurred in only three-quarters of the patients. There were no specific complications associated with ligation of the CAF during surgery. All symptomatic patients showed improvement.

The most important take-home messages from this report are as follows:

- CAF is a rare entity, and clinically relevant CAF is even rarer.
- In adults the diagnosis is usually made incidentally by ICA, in the course of a routine investigation for causes of chest pain or heart failure.
- Clinical presentation and severity of presenting symptoms depend on the origin, insertion site, size and length of the fistula, as well as the volume of blood that is shunted.
- Management should be supported by an established definite mechanistic correlation of symptoms and/or functional abnormalities of the heart with the anatomical characteristics and magnitude of the flow through the CAF. Functional assessment by right heart catheterization and ischemia assessment may be needed to establish this correlation.
- After a thorough assessment, patients with the following conditions should be considered for intervention: hemodynamically significant left-to-right shunt with right chamber enlargement or dysfunction and/or pulmonary hypertension; congestive heart failure with left ventricular volume overload or left ventricular dysfunction; or myocardial ischemia.
- Percutaneous treatment is the preferred therapy, however there is high variability in anatomy, making device choice and complete closure challenging, and not all anatomies are suited for percutaneous closure.
- Anatomic definition of the CAF with topographic imaging is of paramount importance in challenging anatomies.
- No specific dedicated device exists for this indication; devices from the occluder portfolio, coils, or covered stents are usually used according to the morphologic characteristics of the CAF.
- Safety and efficacy are still a concern given the overall low volume and the challenging technical demands of these cases.
- As with other heart conditions, surgery should be considered when there are other indications for surgery, or when percutaneous closure is not technically feasible or safe.
- Ligation of small incidental CAFs may be considered at the time of surgery for other indications, according to the estimated risk of progression to a hemodynamic significant fistula, based on the size and length of the fistula, the vessels or chamber involved, and the patient's age.
- Treatment should be restricted to highly experienced centers and operators, in order to maximize success and safety.

Conflicts of interest

The author has no conflicts of interest to declare.

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