EDITORIAL COMMENT

Addressing Coronary Artery Anatomy and Disease in Adult Congenital Heart Lesions*

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There are now more adults than children living with congenital heart disease. As the prevalence of diabetes, obesity, and hypertension are rising, in our patients (1–3) as in the general population, the incidence of coronary artery disease (CAD) will increase as well (4). CAD was diagnosed in 10% of adult congenital heart disease (ACHD) patients over the age of 65 years in a 2011 population-based study in the province of Quebec, Canada, and myocardial infarction in 7%, in whom it significantly associated with mortality (5). In an analysis of ACHD patients over the age of 60 years seen in a tertiary referral care center in the United Kingdom, CAD was also found to be an independent risk factor for mortality, associated with a higher risk of death than lower New York Heart Association functional class, heart failure, and lower ventricular function in a multivariate model (6). In addition, a higher risk of coronary disease has been seen in particular conditions such as coarctation of the aorta (7), which may be due to the higher lifetime prevalence of hypertension with possible superimposed vasculopathy. Despite their risk factors, patients with ACHD may be less likely to have appropriate lifestyle counseling and treated with guideline-based statin therapy (8), possibly because their cardiovascular care is focused on their congenital lesion. Even patients with lower complexity ACHD are at higher risk than the general population, after adjusting for standard cardiovascular risk factors (3). Finally, patients who undergo surgery involving coronary artery manipulation and reimplantation (such as arterial switch or aortic root replacement) are at higher risk of ostial coronary artery stenoses due to fibrosis, inflammation, or early atherosclerosis.

In this issue of *JACC: Case Reports*, Chag et al. (9) report the case of a patient presenting with an ST-segment elevation myocardial infarction who was known to have congenitally corrected (or L-looped) transposition of the great arteries. He underwent emergent coronary angiography and was found to have a severe stenosis in the mid-right coronary artery, which was treated successfully with a drug-eluting stent. Knowledge of the coronary anatomy seen in most patients with congenitally corrected transposition of the great arteries, namely the anterior and leftward position of the aorta and origin of the coronary ostia from the right and posterior-facing sinuses (10,11), facilitated the procedure by an astute choice of catheters. Anomalous coronary origins, either higher in the sinus or from the opposite cusp are also prevalent in these patients (12,13). Prior to cardiac catheterization, a review of the patient's cross-sectional imaging is routinely recommended, whether with contrast and gated for coronary assessment or not, for estimating the position of the coronaries. Nonstandard angulation of the image intensifier is often required, with attention to obtaining perpendicular views.

This patient presenting with ST-segment elevation myocardial infarction appropriately promptly underwent coronary angiography. The usual diagnostic algorithms for non-ST-segment elevation myocardial infarction or suspected CAD, however, including coronary angiography, cardiac computed tomography (CT) and nuclear perfusion imaging, may be altered in

select patients with ACHD. Care must be taken in the face of lifetime cumulative radiation exposure and consideration given to the use of stress echocardiography, for instance. Prior interventions, in particular involving surgical clips, transcatheter coils or valves, or calcifications, should be considered, and previous imaging or a current chest radiograph obtained prior to considering cardiac CT, as they may create artifacts that lower the sensitivity of the CT below useful thresholds. Many ACHD patients have electrocardiography (ECG) changes at baseline, including bundle branch block in patients with a systemic right ventricle, making ST segment interpretation more difficult, and comparison to prior ECGs is especially important. We encourage our patients to have a copy of their ECG, either on paper or in electronic form on their mobile devices. Atypical cardiac locations (including mesocardia and dextrocardia) make interpretation of ECG and echocardiography wall motion abnormalities difficult to read. As in the case of the patient presented in the report by Chag et al. (9), coronary anomalies, including anomalous origin from the opposite sinus, are common in patients with conotruncal anomalies.

Treatment of CAD is complex in patients with ACHD. Optimal medical therapy may be limited by other comorbidities such as: bradycardia or heart block as a contraindication to beta-blockers; elevated risk of bleeding with addition of antiplatelets in patients with indications for anticoagulation; renal dysfunction; and baseline hypotension limiting use or up-titration of beta-blockers and angiotensin-converting enzyme inhibitors. Percutaneous intervention may be complicated by the use of higher contrast and suboptimal guide catheter support in unusual location of the coronaries, and higher risk of contrast-induced nephropathy in patients with baseline renal disease. In addition, vascular access is often limited due to occlusions after multiple access for childhood procedures. Radial artery access for coronary angiography, which has become the standard in adults with acquired heart disease because it is associated with lower risk of access complications and bleeding requiring transfusions, should be assessed with care in ACHD patients depending on their anatomy (potential for aberrant subclavian arteries, ligation of the subclavian artery in those with a prior classic Blalock-Taussig shunt, or subclavian flap repair of coarctation).

As our population of ACHD patients grows and ages, they are faced with the risks of progression of their congenital heart disease and of complications from hypertension, metabolic syndrome, renal disease, and atherosclerosis including CAD. An important part of their holistic care is to discuss risk factors for coronary disease and to form multidisciplinary teams with expertise in the nuances of diagnosis and treatment of CAD in this population.

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