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INTERMEDIATE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Coronary Ostia Localization by 3-Dimensional Transesophageal Echocardiography in a Patient With Quadricuspid Aortic Valve

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ABSTRACT

Quadricuspid aortic valve is rare and requires surgery when symptomatic severe regurgitation/stenosis is present. Associated anomalous coronary ostia location demands accurate diagnosis to avoid intraoperative complications, and several imaging techniques have been used, with drawbacks of low sensitivity, radiation and contrast exposure. We report a pre-operative assessment using 3-dimensional echocardiography. **(Level of Difficulty: Intermediate.)**

A 65-year-old man with a 6-month history of dyspnea on exertion (New York Heart Association functional class II) was evaluated. Physical examination revealed pulsus celer and an early-mid diastolic in decrescendo blowing murmur on the left lower sternal border. Transthoracic echocardiogram (TTE) revealed left ventricular (LV) dilatation, mild LV systolic dysfunction (LV ejection fraction of 50%), and severe aortic regurgitation (AR) (Video 1). A parasternal (long axis) view did not show an eccentric opening of aortic valve (AV) (Video 2), and the short-axis view was limited. Transesophageal echocardiogram (TEE) confirmed severe AR and revealed a type B quadricuspid AV (QAV) according to the Hurwitz-Roberts classification (1) (i.e., 3 equal and 1 supernumerary cusp) with a diastolic trapezoid apposition defect (Video 3). Three-dimensional (3D) TEE achieved optimal visualization of QAV anatomy (Video 4). Multiplanar reconstruction demonstrated normal origin of coronary ostia (CO) (**Figure 1A,** Video 5) with precise takeoff measures (**Figures 1B and 1C**). Coronary angiography was normal (**Figures 1D to 1E**). Surgical replacement with a bioprosthetic valve was performed, and our 3D TEE findings were confirmed intraoperatively (**Figure 1F**).

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DISCUSSION

QAV is infrequent, with a prevalence of 0.006%, and is associated with AV dysfunction and dilatation of the aortic root or ascending aorta (2). New guidelines do not make specific recommendations for surgery in these patients, so indications for valve surgery follow those of a trileaflet aortic valve, and indications for concomitant aortic root or ascending aorta surgery follow those associated with bicuspid aortic valves (3). As in the case presented here, up to 66% of patients require surgery, mainly for AR (2). According to the relative size of the supernumerary cusp, the Hurwitz-Roberts classification divides QAVs into 7 types from A to G—type B being the most frequent (1)—which may help plan surgery. Diagnosis can be made by TTE, TEE, cardiac computed tomography angiography (CCTA), and magnetic resonance (CMR). CMR stands out for its high spatial resolution and quantitative assessment of regurgitant volume by phase-contrast imaging.

Congenital CO anomalies (COAs) affect 2% to 10% of patients with QAV (2). This may be related to common embryology: both abnormal septation of endocardial cushions and mesenchymal proliferation in the truncus arteriosus have been proposed (4). Pre-operative diagnosis of COAs is relevant to prevent inadvertent surgical complications, such as CO obstruction by a prosthetic valve.

Among diagnostic imaging modalities, coronary angiography and aortography have low sensitivity. TTE allows visualization of the CO in the parasternal short-axis view, but is often limited by poor acoustic windows. CCTA offers good accuracy, but with the disadvantages of radiation and iodine contrast exposure. CMR is free from all of these drawbacks because it is not limited by acoustic windows, and it has high sensitivity. Furthermore, technical features such as echo equivalent—reformatted—views, post-processing virtual angiography, and perfusion sequences allow evaluation of CO anatomy and perfusion. Disadvantages include artifacts caused by tachycardia or respiratory disturbances, high cost, and limited availability (5).

TEE has broader availability and lower costs than CMR, better temporal resolution, and 3D TEE has spatial resolution similar to CCTA and CMR. However, the diagnostic role of 3D TEE in evaluating COAs in QAV patients has not been established yet. Here, we were able to identify the precise location of the CO by 3D TEE and measure—using multiplanar reconstruction—the accurate takeoff of both CO with intraoperative confirmation. However, 3D TEE has limited ability to visualize coronary arteries beyond their ostia. Thus, 3D TEE is able to complement, but not replace, coronary angiography, which may still be required in order to evaluate the presence of coronary artery disease and the need for simultaneous coronary artery bypass surgery. This highlights the usefulness of the multi-image approach in the pre-operative evaluation of patients with QAV.

ABBREVIATIONS AND ACRONYMS

3D = 3-dimensional
AR = aortic regurgitation
AV = aortic valve
CCTA = cardiac computed tomography angiography
CMR = cardiac magnetic resonance
CO = coronary ostia/ostium
COA = coronary ostia/ostium anomaly
LV = left ventricular
QAV = quadricuspid aortic valve
TEE = transesophageal echocardiography
TTE = transthoracic echocardiography

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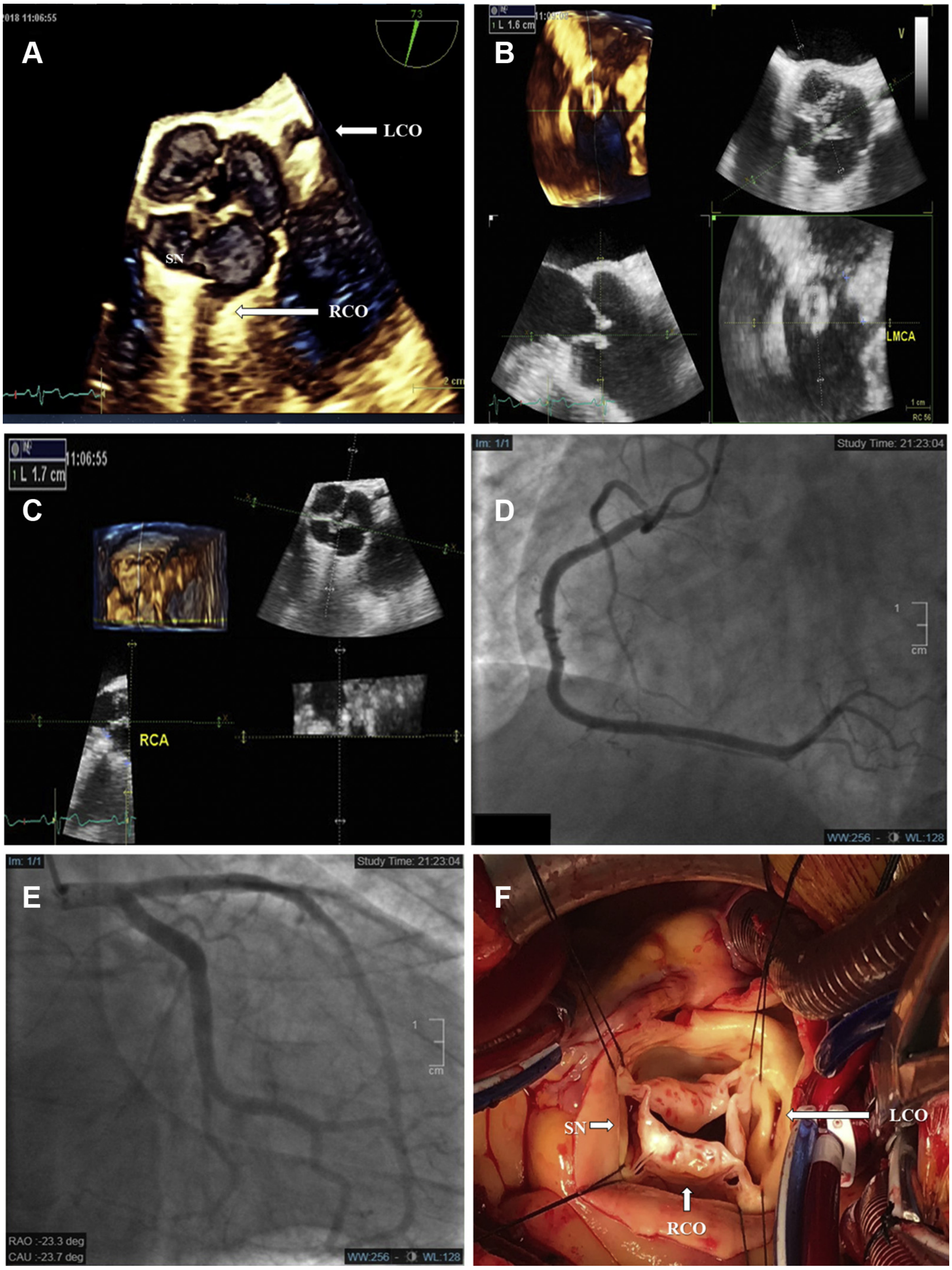
FIGURE 1 3D TEE, Coronary Angiography, and Intraoperative Findings of QAV



(A) 3D TEE: live 3D in mid diastolic frame. Remarkable spatial and temporal resolution, providing optimal visualization of the left coronary ostium (LCO) and right coronary ostium (RCO) **(arrows)**. **(B)** 3D TEE multiplanar reconstruction—left main coronary artery (LMCA) takeoff: the LMCA arises 16 mm above aortic valvular plane. **(C)** 3D TEE multiplanar reconstruction—right coronary artery (RCA) takeoff: the RCA arises 17 mm above aortic valvular plane. **(D)** coronary angiography. right anterior oblique (RAO) view: normal RCA. **(E)** coronary angiography. right anterior oblique view: normal LMCA. **(F)** Intraoperative findings. A type B quadricuspid aortic valve (QAV) was confirmed. 3D = 3-dimensional; CAU = caudal; SN = supernumerary cusp; TEE = transesophageal echocardiography.

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KEY WORDS 3-dimensional imaging, aortic valve, congenital heart defect, coronary vessel anomaly, echocardiography

APPENDIX For supplemental videos, please see the online version of this paper.