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LETTERS TO THE EDITOR

The RAC Sign

Retroaortic Anomalous Coronary Artery Visualization by Transthoracic Echocardiography

An unfamiliar finding was noted on the transthoracic echocardiogram (TTE) apical 4-chamber view in several patients. The appearance was that of a highly echogenic tubular structure, located slightly on the atrial side of the atrioventricular groove (Figure 1A). This was not thought to be an artifact because it was seen with a different appearance in other planes (Figure 1C). Although it was tubular and suggestive of a vascular structure, it did not correlate with the typical location for any normal vessel.

Fortunately, one of the first patients with this finding had previously been evaluated with a contrast-enhanced electrocardiography-gated computed tomography (CT) scan of the chest. That study demonstrated an anomalous left main coronary artery originating from the right sinus of Valsalva and coursing posterior to the aortic root. A comparison of the coronary artery on CT with the TTE findings suggested that the echogenic tubular structure clearly represented the posteriorly coursing anomalous coronary artery (Figures 1B, 1D, and 1E).

Although this sign on TTE, henceforth referred to as the RAC (retroaortic anomalous coronary) sign, appeared to represent a retroaortic anomalous coronary artery, more rigorous correlation with coronary CT in other patients was needed to confirm the findings and assess the usefulness of TTE in identifying these anomalies. We subsequently performed a retrospective, investigator-blinded, case-control analysis that was approved by the Mayo Clinic institutional review board. Adult patients with a retro-aortic coronary artery anomaly on a CT scan obtained from 2005 to 2015, as confirmed by a cardiac radiologist (T.A.F.), were included. Patients with congenital anomalies of the great arteries or aortic valve or root surgery were excluded. In a similar manner, a group of patients without a coronary artery anomaly on CT were matched to the case group in a 1-to-1 fashion by age, sex, and date of TTE.

The case and control groups, 49 patients in each, were randomized into a single study population. Two investigators (C.M.W. and L.A.E.), blinded to the coronary artery anatomy, independently reviewed TTE images to determine whether the RAC sign was present, with disagreements reviewed by a third investigator (D.A.F.).

The RAC sign was seen in 31 patients (63.3%) with confirmed coronary anomalies and 3 (6.1%) with normal coronary anatomy. The sensitivity of the RAC sign was 63.3%, and the specificity was 93.9%. The Fisher exact test demonstrated a significant association between the RAC sign and the presence of a retroaortic anomalous coronary artery ($p < 0.001$).

In this retrospective study, we were limited by the images obtained during clinical practice. Imagers, suspecting an artifact, might have intentionally tried to exclude the finding, decreasing the sensitivity of the RAC sign in our study. Furthermore, we had to make a judgment call in equivocal cases, potentially leading to false positives. In clinical practice, further focused imaging can be used to distinguish a true RAC sign from mimics such as calcified valves, normal coronary arteries, or mitral annular calcification. Lastly, although TTE reviewers were blinded, there was an understanding that one-half of the patients had a coronary anomaly, which could have created bias.

Despite these limitations, the RAC sign was strongly associated with the finding of a retroaortic coronary anomaly on CT, confirming the identity of this structure on TTE. The RAC sign also has a high specificity, signifying that its presence could be documented as strongly suggestive of an anomalous coronary artery on echocardiogram reports. This finding, to the best of our knowledge, has not been highlighted previously in the medical literature. However, with coronary anomalies occurring in 1.3% of adults (1), it now may be recognized fairly frequently by imagers aware of its significance.

The clinical implications of the RAC sign are uncertain. Although anomalous retroaortic coronary arteries are believed to be mostly benign, there have been reports of morbidity and mortality (2), as well as damage during valve surgery (3). We have also seen the RAC sign misdiagnosed as coronary artery calcification. Therefore, it is important to understand what the RAC sign represents, so that it is not ignored, dismissed as an artifact, or given an incorrect diagnosis.

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FIGURE 1   **The RAC Sign**

Transthoracic echocardiogram demonstrating the RAC (retroaortic anomalous coronary) sign in the typical apical 4-chamber view **(A)** and a nonstandard view demonstrating bifurcation of the artery **(C)**, along with computed tomography angiogram images **(B, D, and E)** demonstrating the course and origin of the anomalous left main coronary artery.

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REFERENCES

1. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. Cathet Cardiovasc Diagn 1990;21: 28–40.

2. Kejriwal NK, Tan J, Gordon SP, Newman MA. Retroaortic course of the anomalous left main coronary artery: is it a benign anomaly? A case report and review of literature. Heart Lung Circ 2004;13: 97–100.

3. Vaishnava P, Pyo R, Filsoufi F, Sharma S. Compression of an anomalous left circumflex artery after aortic and mitral valve replacement. Ann Thorac Surg 2011;92:1887–9.