Coronary artery anomalies in the absence of structural heart defects are rare with an estimated prevalence of 0.6% to 1.2% in the general population. Although the anomalous left coronary artery (LCA) from the right cusp is quite rare (0.03%–0.05% estimated prevalence), it is frequently associated with sudden cardiac death, especially when intramural. A similar presentation has been described in 30% of patients with a right coronary artery from the left cusp. Given the unknown prevalence of this disease, the natural history of these lesions has not been truly defined.

Transthoracic echocardiography remains the primary screening tool in young patients with exercise-induced cardiac symptoms. The accuracy of this modality in identifying the abnormal origin and course of coronaries has not been established, in part due to the unknown prevalence of the disease. The technical challenge of transthoracic echocardiography lies in obtaining reliable images of relatively small structures with translational movement during the cardiac cycle. Few prospective studies have sought to evaluate and overcome the limitations of transthoracic echocardiography. Transesophageal echocardiography,

Figure 1. A–B, Parasternal short axis 2-dimensional image (A) and color Doppler flow (B) at the level of the aortic root falsely demonstrate the origin of the left coronary artery as normal. C–D, Two-dimensional (C) and color Doppler flow (D) imaging in the high parasternal short axis view close to the sinotubular junction demonstrate an intramural left coronary artery traveling between the aorta and pulmonary artery. E–F, Corresponding MRI images from the same patient are presented. The black arrow denotes the origin of the left coronary artery. In the first image (E), it appears to arise from the left cusp. A higher plane (F) demonstrates that it in fact originates from the right cusp.
CT scan, and MRI have been recommended when a high index of suspicion is present. However, the rarity of congenital coronary abnormalities in young patients renders a low yield and cost-effectiveness of additional tests.

To thoroughly evaluate the coronary arteries, it is important to understand the anomalous course of the vessel. Intraoperative assessment has often demonstrated a higher takeoff of the coronary into the intramural segment with a normal-appearing exit from the appropriate sinus. Therefore, 2-dimensional imaging alone can be deceiving; color Doppler flow is imperative to visualize the coronary flow within the aortic wall.

We present our experience with 4 cases of anomalous coronaries in which routine imaging resulted in a normal appearance of the coronary artery origins; false-negative results were avoided by further interrogation of more distal planes of the aortic root.

Echocardiography
Transthoracic echocardiography 2-dimensional images and color Doppler flow mapping were reviewed in patients with confirmed anomalous coronary artery origin from the contralateral sinus. All images were obtained using Sequoia ultrasound equipment (Siemens AG, Mountain View, CA) or Philips iE33. We used 8- and 5-MHz transducers. The coronaries were interrogated in parasternal short axis views in the plane of the aortic root with sweeps extending from the aortic valve annulus to the sinotubular junction. The anterior aortic wall in high parasternal long axis was interrogated for a coronary in cross-section.

Patients
Two patients were diagnosed with an anomalous LCA from the right sinus (Figures 1 and 2), both 15-year-old boys who underwent surgical unroofing of the intramural segment. The other 2 patients were diagnosed with an anomalous right coronary artery from the left sinus (Figures 3 and 4), a 14-year-old girl with a secundum atrial septal defect and a 7-year-old boy with congenital heart block, both followed clinically. Both anomalous left coronaries were confirmed
intraoperatively (one had MRI preoperatively). The anomalous right coronary arteries were confirmed by CT scan.

In our cohort, the coronary artery origins in the standard parasternal short-axis view appeared normal and this false-negative was confirmed by color Doppler (online-only Data Supplement video I). On a higher sweep of the aortic root, the anomalous origin and course was identified by 2-dimensional imaging and confirmed by color Doppler (online-only Data Supplement video II). The aberrant coronary could also be visualized within the anterior aortic wall in parasternal long axis.⁴

The LCA could also be evaluated in the careful parasternal long axis sweep from the aorta to the pulmonary artery. In this plane, the normal LCA travels almost parallel to the transducer (Figure 5A). An interesting, subtle observation in both our patients with the abnormal LCAs was the visualization of the short, anomalous course of the LCA in this view. It travels in a more oblique direction to the probe as it dives posteriorly in its proximal

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Figure 4. A, Parasternal short axis 2-dimensional image at the level of the aortic root falsely demonstrates the origin of the right coronary artery as normal. B–C, Two-dimensional (B) and color flow Doppler (C) imaging in the high parasternal short axis view close to the sinotubular junction demonstrates the interarterial right coronary artery.

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Figure 5. A, Normal left coronary artery course in parasternal long axis view. B–C, Abnormal proximal course of anomalous left coronary artery from the right cusp in the parasternal long axis in Patients 1 (B) and 4 (C).
segment. (Figure 5B–C). In our experience, we have not observed this oblique angle in patients with normal coronaries.

Although parasternal short axis view has been described to delineate the origins of the coronary arteries, emphasis has not been placed on identifying a higher takeoff of the abnormal vessel. We noted that when examined at the level right below the sinotubular junction, an intramural coronary may appear to have a normal takeoff as it exits the aorta. However, a careful sweep to the level above the sinotubular junction identified the intramural component in all patients. We propose this sweep be incorporated into routine echocardiograms; when used in conjunction with color Doppler flow and high parasternal long axis, it should increase the accuracy of the diagnosis. The decrease in false-negative results should improve our understanding of these lesions and further our knowledge of its natural history.

Disclosures
None.

References

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