Trust, but verify

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During the development of the cardiac outflow tracts, the separation into two distinct arterial trunks is a complex process. Fusion of the truncal protrusion with the distal end of the outflow cushions completes this separation.¹ Failure to close this embryonic aortopulmonary foramen leads to the rare congenital defect of an aortopulmonary window (APW). Although the underlying mechanisms of the development of the coronary plexus giving rise to the coronary arteries are not interrelated with those seen in truncal separation, coexistence of APW with coronary artery anomalies has been described in as many as 10% of cases.²

In this issue of the *Journal*, Alhadlaq and colleagues³ present a rare case of a patient with APW and pulmonary origin of the left coronary artery. Echocardiography identified the communication between aorta and pulmonary artery as well as normal coronary anatomy. Severe depression of left ventricular function was noted after initial repair of the APW, and inadequate flow in the left coronary artery was detected by transesophageal echocardiography. The pulmonary origin of the left coronary artery was confirmed by surgical inspection of the left coronary ostium, and a baffle was used to reroute the coronary artery and close the window.

Although the case and its surgical repair do not present an entirely new clinical scenario, several important concepts of the management of APW can be highlighted. Despite the excellence of echocardiography as a noninvasive preoperative imaging technique, coronary anomalies remain challenging to detect. In particular, the physiologic characteristics of APW, with high pressure in the pulmonary artery and diastolic runoff into the pulmonary vasculature, limit the echocardiographic assessment to a direct visualization of the aberrant coronary ostium. Indirect signs seen in isolated pulmonary origin of the left coronary, such as an enlarged right coronary artery, septal collaterals, retrograde blood flow in the proximal left coronary artery, reduced ventricular function, or mitral valve regurgitation, are usually not encountered. The

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Central Message

Intraoperative assessment of coronary artery anatomy is essential, even if suspicion is low and preoperative imaging results are negative.

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anomaly thus may be missed by echocardiography, and any inconclusive imaging should raise the suspicion of a coronary anomaly until demonstrated otherwise. In patients with ambiguous results, transesophageal echocardiography, magnetic resonance imaging, computed tomography, or even diagnostic angiography may be considered.⁴ Although the coronary anomaly can be identified intraoperatively, the upcoming use of transcatheter devices to close the APW makes a thorough assessment of the coronary anomaly even more important, because coexistence changes the therapeutic strategy. Irrespective of the preoperative workup, the ostia of the coronary arteries have to be identified intraoperatively to confirm or refute the initial diagnosis. Unexplained reduced ventricular function, although uncommonly associated with APW preoperatively, should also raise concerns of impaired coronary blood flow, either because the repair itself compromises the coronary artery or because an unrecognized congenital anomaly exists.

Preoperative diagnostic imaging of patients has achieved outstanding reliability. Nevertheless, the case presented by Alhadlaq and colleagues³ demonstrates that preoperative imaging and intraoperative assessment are not redundant but rather complement each other. Sometimes, the surgeon's vigilance can make all the difference between a grim and an excellent outcome.

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