The origin of the left main coronary artery (LMCA) from the pulmonary artery (PA), known as Bland-White-Garland syndrome (BWGS), is a rare anomaly with mortality of 90% during infancy (1). If collateral circulation between right and left coronary systems ensues, BWGS patients may reach adulthood (1,2). However coronary flow directs preferentially into the lower pressure PA, away from the left ventricle (LV) creating “coronary steal,” and left-to-right shunt. Mitral regurgitation (MR) is further common in BWGS most likely due to multiple factors (3). Surgical correction of BWGS is strongly recommended, even in asymptomatic patients, because of the high risk of developing heart failure, myocardial infarction, ventricular arrhythmias, and sudden death (1,2).

A 44-yr-old man presented with recent onset of fatigue and atrial fibrillation. Transthoracic echocardiography demonstrated severe MR with a posterior-directed jet, mild anterior mitral leaflet prolapse, a dilated mitral annulus, enlarged left atrium (5.8 cm), preserved global LV function, a moderately dilated right ventricle with moderate tricuspid regurgitation, and estimated right ventricle systolic pressure of 60 mm Hg. Because of the patient’s occasional chest pain, a persantine–thallium study was performed demonstrating a significant perfusion defect of the anterior...
LV wall. Cardiac catheterization revealed the absence of the LMCA in the left sinus of Valsalva. Contrast dye injected into the right coronary artery (RCA) filled the main branches of the LMCA with anomalous drainage through the LMCA into the PA.

The patient was referred for mitral valve (MV) repair and correction of anomalous LMCA. Intraoperative transesophageal echocardiography (TEE) mostly confirmed the preoperative echocardiography findings but now demonstrated a large RCA (17 mm of diameter) that was identified at the right sinus of Valsalva. Color flow Doppler examination showed that the LMCA passed close to the left sinus of Valsalva with no connection to the aortic root. The LMCA origin was identified as arising from the PA, 18 mm distal to the pulmonic valve with retrograde flow into the PA (Figs. 1 and 2; please see video clip available at www.anesthesia-analgesia.org).

The patient underwent uncomplicated reimplantation of the LMCA into the aortic root and MV repair. The postoperative TEE showed LMCA orifice reimplanted into the left sinus of Valsalva, and the MV annuloplasty ring with mild MR (Fig. 3).

Identification of the coronary orifices and normal coronary flow pattern is an important part of a TEE examination. Young adults with no risk factors for coronary disease may present with MR and unrecognized BWGS. In the presence of a single enlarged RCA orifice, BWGS should be considered.

REFERENCES


Figure 3. Postoperative multiplane transesophageal echocardiography (TEE), mid-esophageal aortic valve short axis view. The LMCA orifice now reimplanted into the left sinus of Valsalva can be seen. (LA, left atrium; RA, right atrium; IAS, interatrial septum; LCC, left coronary cusp; RCC, right coronary cusp; NCC, noncoronary cusp; LMCA, left main coronary artery.)