Pivotal role of echocardiography for the diagnosis and follow-up of the anomalous origin of the left main coronary artery from the pulmonary artery

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A 36-year-old woman presented with typical unstable angina. Transthoracic echocardiography in both the parasternal short-axis view and the four-chamber view depicted large multiple lacunae in the interventricular septum with intense turbulent continuous flow (Panel D; see Supplementary data online, Movie S1 and Panel G, respectively). Congenital malformation of the coronary arteries or abnormal communications resembling fistulas was implied. From the parasternal long-axis view, only the enormous origin of the right coronary artery was identified (Panel A). From modified parasternal view, above the pulmonary valve a cavity-like structure interrupting the continuity of the posterior wall of the pulmonary artery (Panel B) was visualized. The structure was fully occupied with blood flow (Panel C) and represented the origin of the left main artery from the posterior aspect of the pulmonary artery, as shown by computed tomography (Panel I), and the flow depicted the retrograde filling of the left main coronary artery to the pulmonary artery as seen in angiography (Panel F; see Supplementary data online, Movie S2).

The patient was surgically treated and is still, 30 months after, free of symptoms, has no signs of ‘collateral vessels’ (Panels E and H) and her stress echo is faultless.

Although sophisticated imaging techniques are utilized in the management of the syndrome, echocardiography can be uniquely practical, contributing both in the diagnosis and the follow-up of these patients. A high suspicion index is only required. The immense turbulent flow in the interventricular septum and the detection of flow towards the pulmonary trunk warrant the diagnosis.

Panel A. Echocardiography, in parasternal long-axis view with zoom, identified an enormous origin of the right coronary artery (RCA) from the right sinus of Valsava without any evidence for the left coronary origin.

Panels B and C. From modified parasternal long-axis view, above the pulmonary valve a cavity-like structure interrupting the continuity of the posterior wall of the pulmonary artery (arrow) was visualized. The structure was fully occupied with blood flow in colour Doppler imaging (Panel C) and represented the origin of the left main artery from the posterior aspect of the pulmonary artery, as shown by computed tomography, and the flow depicted the retrograde filling of the left main coronary artery to the pulmonary artery as seen in angiography (Panels I and F, respectively).

Panels D and G. With colour Doppler imaging a significant flow (arrow) is detected, mainly in diastole, throughout the interventricular septum (arrows) illustrating the collateral circulation (both in parasternal short-axis and apical four-chamber view).

Panels E and H. The turbulent flow of the extensive collateralization, seen in Panels D and G, disappears after surgical treatment and throughout follow-up. (LV, left ventricle).

Panel F. Selective right coronary (RCA) catheterization showed a widely dilated artery, filling retrograde, through dense collaterals, the left coronary artery and finally, roughly portraying the pulmonary artery by draining there the contrast agent. (PA, pulmonary artery, Lm, left main coronary artery).

Panel I. Cardiac-computed tomography confirmed that the left coronary artery (Lm) originated from the pulmonary artery (PA) while the dilated right coronary artery (RCA) originated normally from the aorta (Ao).

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.