Imaging of an unusual case of a completely unroofed coronary sinus without persistent left superior vena cava

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A 66-year-old male with a history of surgically closed secundum type atrial septal defect (ASD) during childhood and a percutaneous coronary intervention complained of recurrent stable angina. Clinical investigation and 12-lead ECG were unremarkable. He was referred for hybrid myocardial perfusion SPECT (SPECT) and 64-slice CT coronary angiography (CCTA).

SPECT showed normal perfusion of the left ventricle (LV) and an enlarged right ventricle (RV). Gated images showed a decreased LV ejection fraction of 50% due to septal wall motion abnormalities suggestive of RV overload. Prospectively ECG-triggered CCTA revealed minor coronary atherosclerosis and a patent stent. Severe dilatation of the RV (388 mL mid-diastolic) was confirmed. By coincidence, an inferior sinus venous defect with an unroofed coronary sinus with the right-sided superior vena cava was observed (situs solitus, atrio-ventricular, and ventriculo-arterial concordance). Middle and great cardiac veins were inserted into the roof of the left atrium and a clear left-to-right shunt was visualized by CCTA. Three-dimensional transoesophageal echocardiography showed the circular ASD (14 mm) near the inferior vena cava. Right heart catheterization confirmed the left-to-right shunt (Qp:Qs 2.1) and pulmonary arterial hypertension (mean PAP 29 mmHg and PVR 106 dynes/s/cm°5). Invasive coronary angiography confirmed the absence of recurrent significant CAD.

This case was discussed in our grown-up-congenital-heart-disease team, and the patient underwent surgical closure of the defect, which is the treatment of choice in patients without Eisenmenger syndrome. Currently, 4 months after surgery, the patient is free from anginal complaints.

Conflict of interest: none declared.

Panel A. Quantitative gated SPECT showing bulls eye of left ventricle (LV) motion with a septal wall motion abnormality (black arrow).
Panel B. Myocardial perfusion SPECT, attenuation corrected, showing an enlarged right ventricle (RV) and normal perfusion of the LV.
Panel C. Computed tomography coronary angiography showing an interatrial connection with the left-to-right shunt (black arrow) and insertion of the great cardiac vein in the left atrium (LA) (white arrow).
Panel D. Computed tomography coronary angiography showing an enlarged RV, 388 mL mid-diastolic.
Panel E. Transoesophageal colour-Doppler echocardiography showing severe interatrial left-to-right shunting (white arrow).
Panel F. The three-dimensional transoesophageal echocardiography view from the LA towards the interatrial septum showing an ASD (black arrow).

IVS, interventricular septum; RVOT, right ventricular outflow tract; AoV, aortic valve; RA, right atrium; LA, left atrium; LV, left ventricle; RV, right ventricle; and IAS, interatrial septum.

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