A 69-year-old man complaining of dyspnoea on moderate exertion was referred to our department due to a suspicion of right coronary artery (RCA) malformation based on transthoracic echocardiography.

During transthoracic and transoesophageal echocardiography (TTE and TEE), we found a severe dilatation of the proximal part of RCA, vascular structure located near the free wall of his right ventricle and also posterior to the heart, increased, turbulent flow from the coronary sinus (CS) to right atrium (RA) and small atrial septal defect (Panels A–C; see Supplementary data online, Video S1). Estimated pulmonic:systemic flow ratio was 2:1, and right ventricular systolic pressure was 60 mmHg. The multi-detector coronary angiography (MDCT) revealed particularly well the serpiginous appearance of a giant, tortuous RCA (Panels D–F; see Supplementary data online, Video SII) connected to the CS. Coronary angiography as well as a cardiac magnetic resonance (CMR) imaging were also performed (Panels G and H; see Supplementary data online, Videos SIII and SIV). The overall length of the fistula calculated from the RCA ostium to the CS ostium was over 78 cm; the maximum RCA width was 2.7 cm. We found also marked dilatation of the right atrium and ventricle (RA 84 mL/m², RV 117 mL/m²).

Surgical treatment was advised to the patient (Panel I). The proximal RCA was cut off and closed. Communication point with the CS was identified from inside and ligated. A saphenous vein graft was made to the posterior descending artery as well as an ASD closure was performed. The patient was discharged from hospital in a very good condition.

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