The role of multi-modality imaging to investigate and manage anomalous right coronary artery originating from the pulmonary artery (ARCAPA) anomaly with associated coronary aneurysms presenting as acute left ventricular failure

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A 53-year-old hypertensive gentleman presented with acute left ventricular (LV) failure. His mother died aged 42 following a spontaneous rupture of the pulmonary artery (PA). A transthoracic echocardiogram found severe LV dilatation (7 cm) and impairment. A computed tomography (CT) angiogram was performed (Panels A1–A3) to exclude significant coronary disease and demonstrated an aberrant origin of his right coronary artery (RCA) from the main PA (MPA) with an aneurysm in its proximal part. In addition, there was an aneurysm in the proximal left anterior descending (LAD) artery. Both aneurysms were communicating with each other via a fistula. A coronary angiogram (Panel B) confirmed the presence of a large aneurysm of the proximal LAD and an anomalous origin of the RCA that was associated with a fistula to the MPA and receiving arterial blood from the left circumflex artery (LCx). This was also established by cardiac magnetic resonance scanning. Taken together, multi-modality imaging therefore confirmed an anomalous RCA originating from the PA with a collateral supply from the LCx. Potential-associated intracranial aneurysms were excluded by a CT brain scan. After being commenced on anti-heart failure therapy for a dilated cardiomyopathy, he went on to undergo successful surgical resection of the LAD aneurysm (Panel C), re-implantation of the RCA as well as coronary bypass grafting of both RCA and LAD. In the subsequent year following surgery, the patient has remained well and at most recent follow-up his LV size and function have both returned to normal.

Panel A Computed tomography angiogram demonstrating (A1) the relationship between right coronary artery and main pulmonary artery in coronal and (A2) axial section, where the left anterior descending aneurysm and fistula can also be seen. Three-dimensional computer reconstruction (A3) further reveals both right coronary artery and left anterior descending aneurysms and their fistulous communication in relation to cardiac surface anatomy.

Panel B Left anterior descending aneurysm as seen by coronary angiography.

Panel C Intra-operative image demonstrating the incised left anterior descending aneurysm taken during cardioplegia at coronary bypass surgery. Supplementary material online, Video S1 demonstrating samples from the left heart catheterisation. The fistulous connection between left and right coronary systems is seen at 48s. Filling of the main pulmonary artery following injection of the left coronary artery is seen at 50s. The fistulous connection between left and right coronary systems is seen filling at 55s.

Supplementary material is available at European Heart Journal online.