

Right ventricular thrombus-induced myocardial infarction after Fontan surgery in pulmonary atresia with intact ventricular septum

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A 27-year-old man was referred to our hospital for palpitations and chest pain. He was born with pulmonary atresia with intact ventricular septum (PA/IVS), and right ventricle (RV)-dependent coronary artery via coronary-right ventricular fistula. He had undergone extracardiac Fontan procedure at the age of 5 years because biventricular repair was not feasible. Electrocardiogram (ECG) showed incessant ventricular tachycardia (VT) without haemodynamic compromise (Supplementary material online, Figure S1), and therefore the patient received landiolol and there was no recurrence of VT thereafter. ECG in sinus rhythm showed mild ST-segment elevation in V2-4 (Supplementary material online, Figure S2). High-sensitive Troponin I was elevated at 1577.8 pg/mL (normal range <26.2 pg/mL). Invasive coronary angiography showed the left anterior descending artery (LAD) atresia at mid region, and it was occluded at junction of coronary-right ventricular fistula (Panel A, Video 1). 99mTc myocardial scintigraphy revealed myocardial infarction in the LAD territory. Late gadolinium enhancement on cardiac magnetic resonance imaging further confirmed myocardial infarction in the same region (Panel B, red arrowheads). Delayed phase of coronary computed tomography angiography showed organized thrombi that filled the RV (Panel C, yellow arrowheads; Panel D, blue areas). Based on these findings, we concluded that the myocardial infarction was caused by the thrombotic obstruction of the coronary-right ventricular fistula supplying the LAD (Panel E). Direct revascularization for the occluded coronary-right ventricular fistula was difficult. Moreover, myocardial scintigraphy showed poor viability in the anterior territory. Therefore, after discussion in the congenital heart disease team, we decided to perform anticoagulation therapy with warfarin to prevent





further thrombotic event. He was free of cardiovascular events during 18 months of follow-up. The present case illustrates right ventricular thrombus as an unusual cause of ischaemic events. While there are no clear recommendations for use of prophylactic anticoagulation in patients with Fontan, patients with PA/IVS after Fontan surgery are among the high-risk patients for development of thrombosis and therefore long-term

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anticoagulation should be considered in these patients to prevent thrombotic events.

Supplementary material

Supplementary material is available at European Heart Journal - Case Reports online.

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