Takotsubo syndrome (TTS) is a stress-induced reversible cardiomyopathy, characterized by a transient reversible left ventricular dysfunction. We present a case series of systemic right ventricular TTS (RV-TTS). A 40-year-old woman with history of transposition of the great arteries corrected by an atrial switch (TGA-AS) was referred for acute chest pain after an emotional stress. Troponin T level was increased and electrocardiogram (ECG) modified (Panel 3A). Coronary angiography was normal. Transthoracic echocardiography (TTE) showed decreased of systemic RV function with ballooning of the apex with normal left ventricle (LV) function confirmed by magnetic resonance imaging (MRI) without late contrast enhancement (Panels 1A, 1C, and 1E, Supplementary material online, Video S1A). She was diagnosed as a systemic RV-TTS and received a medical therapy. Six months later, TTE and MRI exhibited a complete restoration of the RV kinetics, and ECG was normalized (Panels 1B, 1D, and 3B, Supplementary material online, Video S1B).

The same year, a 46-year-old woman with previous history of TGA-AS was referred for acute pulmonary oedema associated with chest pain after a stress. ECG, TTE, coronary angiography, and MRI diagnosed a systemic RV-TTS with complete clinical and imaging recovery 1 month later (Panel 2, Supplementary material online, Video S2).

To our knowledge, these are the two first described cases of systemic RV-TTS. In systemic LV classical apical TTS, the most common hypothesis is the high level of circulating catecholamine during a stress leading to a negative inotropic effect through the cardiac β-adrenoceptors which density is higher in the apex. We hypothesized, that in systemic position, the number and function of β-receptors in RV cardiomyocytes is modified with an increase sensibility to catecholamine responsible of TTS as for systemic LV. Nevertheless, understanding the pathogenesis of systemic RV-TTS has to be elucidated. Acute heart failure or chest pain in systemic RV patient should suggest a TTS.

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(Panels 1A–1D) MRI of systemic RV of the first patient in systole, during TTS in long axis (Panel 1A) and short axis at the apical level (Panel 1C) and recovery 6 months later (Panels 1B and 1D). (Panels 1E and 2E) MRI showing no late contrast enhancement. (Panels 2A–2D) MRI of systemic RV of the second patient in systole during TTS in long axis (Panel 2A) and short axis (Panel 2C) and recovery 1 month later (Panels 2B and 2D). (Panel 3) Electrocardiogram during the acute chest pain (Panel 3A) and 6 months later (Panel 3B).

Supplementary material is available at European Heart Journal online.

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