Takotsubo Cardiomyopathy in a woman with Pulmonary Atresia, Ventricular Septal Defect, and Major Aorto-Pulmonary Collateral Arteries

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Takotsubo cardiomyopathy (TC) is a transient reversible stress-induced cardiomyopathy. Although TC is frequently described in adults, it is rarely reported in Grown-Up Congenital Heart Disease (GUCH) patients.

We report the case of a 43 year-old woman who came to our emergency department complaining of acute chest pain and dyspnea after an angry debate.

At birth she was affected by pulmonary atresia, ventricular septal defect and major aorto-pulmonary collateral arteries. During childhood she underwent right Blalock-Taussig shunt, left Blalock Taussig shunt and left pulmonary artery reconstruction. When she was 35 years old, ventricular septal defect closure and unifocalization was performed; one year later, replacement of the ascending aorta and stenting of right pulmonary artery were performed.

Physical examination evidenced tachycardia, tachypnea, oxygen saturation 68% on oxygen 8 l/min, blood pressure 140/80 mmHg.

Electrocardiogram showed mild ST depression. Echocardiography revealed severe ventricular dysfunction, akinetic mid to apical left ventricular segments and normally contracting basal segments, left ventricular dilation and severely depressed ejection fraction (EF), 30%. Troponin was still normal even 1 hour after the access to the hospital.

Three hours after symptoms onset, the patient was hemodynamically stable. Troponin I turned mildly positive, electrocardiogram showed negative T waves. Coronary angiography evidenced no coronary disease. According to Mayo Clinic criteria, TC was diagnosed.

After 4 days, echocardiography showed normal left ventricle volume and improved left ventricular systolic function (EF: 50%), whereas apical hypokinesia persisted.

After 45 days, echocardiography documented recovered systolic function (EF:55%) and no more apical dysfunction.

Conclusions: Few cases of TC in GUCH patients have been described. This is the first reported case of TC in a patient with pulmonary atresia, ventricular septal defect and major aorto-pulmonary collateral arteries.

Of note, all reported cases in GUCH patients involved young women, which is quite unusual in the setting of TC, since postmenopausal women are more likely to be prone to TC. It should be considered that the number of GUCH patients is dramatically increasing and therefore we might expect to observe TC even more frequently and in older ages.