The unexpected in grown-up congenital heart disease: Takotsubo syndrome

Lilia Oreto, MD,^{a,b} Claudia Parrella, MD,^{a,c} Roberto Ricci, MD,^d and Marcello Chinali, MD,^a Rome, Messina, and Naples, Italy

From the ^aMedical-Surgical Department of Pediatric Cardiology, Bambino Gesù Pediatric Hospital, Rome; ^bClinical and Experimental Department of Medicine, University Hospital of Messina, Messina; ^cPediatric Science Department, Federico II University, Naples; and ^dSanto Spirito in Saxia Hospital, Cardiology Unit, Rome, Italy.

Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication March 20, 2017; revisions received May 22, 2017; accepted for publication June 2, 2017; available ahead of print June 30, 2017.

Address for reprints: Lilia Oreto, MD, Department of Clinical and Experimental Medicine, University Hospital, Via Consolare Valeria 100, Messina, Italy (E-mail: liliaoreto@hotmail.com).

J Thorac Cardiovasc Surg 2017;154:e107-9

0022-5223/\$36.00

Copyright © 2017 by The American Association for Thoracic Surgery

<http://dx.doi.org/10.1016/j.jtcvs.2017.06.001>

Left ventricular systolic Takotsubo shape.

Central Message

Takotsubo syndrome is a stress-related, acute, and reversible heart failure, usually described in postmenopausal women. We report the first case of Takotsubo syndrome in a woman with operated pulmonary atresia.

See Editorial Commentary page e111.

▶ Video clip is available online.

CLINICAL SUMMARY

Takotsubo syndrome (TS) is a transient, reversible, stress-induced heart failure syndrome.¹ Although TS frequently is described in adults, it rarely is reported in grown-up patients with congenital heart disease.²⁻⁴ We report the case of a 43-year-old woman with operated pulmonary atresia and ventricular septal defect who was admitted for acute chest pain and dyspnea lasting for 2 hours.

The patient was born with pulmonary atresia and major aortopulmonary collateral arteries and underwent multiple surgical interventions: systemic to pulmonary McGoon shunt at 3 months, right Blalock-Taussig shunt at 8 months, and left Blalock-Taussig shunt and left pulmonary artery reconstruction 2 years later. At 35 years, she underwent surgical repair with ventricular septal defect closure, unifocalization by 24-mm homograft, and ascending aorta replacement with a 26-mm Hemashield aortic graft. One year later, stenting of the right pulmonary artery was performed. At presentation, she was on aspirin 100 mg daily and sotalolol 160 mg twice daily for occasional episodes of atrial fibrillation.

A few minutes before the onset of symptoms, the patient was having an angry debate. Physical examination evidenced poor clinical conditions, pale skin, tachycardia, and tachypnea. Oxygen saturation was 60% on room air, 68% on oxygen 18 L/min, and blood pressure was 140/80 mm Hg.

The electrocardiogram showed mild ST depression and negative symmetric T waves in lateral and inferior leads (Figure 1, A). Echocardiography revealed a dilated left ventricle (Video 1) with severely depressed ejection fraction (30%); there was evident asymmetry of regional function with typical akinetic mid-to-apical segments and normally contracting basal segments, resembling the traditional Japanese octopus trap called Takotsubo. Troponin was still normal at admission.

With suspicion of acute coronary syndrome, the patient was quickly transferred to a coronary intensive care unit. After 3 hours, troponin I turned mildly positive (0.13 ng/mL, range 0-0.04), symptoms resolved, and normal epicardial vessels on coronary angiography confirmed the diagnosis of TS (Figure 1, B and C).

As expected, after 4 days echocardiography showed normal left ventricular volume and improved systolic function with 50% ejection fraction, whereas apical hypokinesia persisted. The patient was discharged on aspirin, metoprolol, and ramipril. After 45 days, systolic function recovered (except for pre-existing mild septal dyskinesia) with 55% ejection fraction and no more apical dysfunction (Video 2).

Diagnostic criteria for TS recently were updated by the European Society of Cardiology and include¹ (1) transient wall motion abnormalities, often preceded by emotional

FIGURE 1. A, Electrocardiogram. B, Left and (C) right coronary angiography. Left ventricular diastole (D) and systole (E) on echocardiography; arrows indicate basal segments contracting normally, resembling the neck of takotsubo pot.

stress; (2) wall motion abnormalities extended beyond a single coronary artery distribution; (3) absence of culprit coronary artery disease; (4) acute and reversible electrocardiographic changes; (5) elevated brain natriuretic peptide;

(6) mild troponin elevation; and (7) recovery of systolic function during follow-up. Sympathetic activation and catecholamines have a central role in the pathophysiology of

VIDEO 1. Left ventricular apical ballooning and systolic “Takotsubo” shape. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(17\)31158-3/addons](http://www.jtcvsonline.org/article/S0022-5223(17)31158-3/addons).

VIDEO 2. Recovered left ventricular systolic function after 45 days. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(17\)31158-3/addons](http://www.jtcvsonline.org/article/S0022-5223(17)31158-3/addons).

TS, producing a complex interaction with cardiovascular system.

TS already has been reported in 2 cases of Fallot tetralogy and 1 Fontan circulation. This is the first report of TS in pulmonary artery atresia. Of note, all 4 reported cases involved young women in premenopausal age, which is quite unusual because postmenopausal women are more likely to be prone to TS.^{1,5} In our patient, the provoking event was related to the emotional stress caused by the angry debate, whereas in the report by Aregullin and colleagues,⁴ TS occurred after a physical stress determined by transcatheter intervention. Perhaps the profound and transient decrease of oxygen saturation was enhanced by the underlying congenital pulmonary vascular disease.

It should be considered that the number of grown-up patients with congenital heart disease is dramatically

increasing, and, therefore, we might expect to observe TS even more frequently and in older ages.

References

1. Lyon AR, Bossone E, Schneider B, Sechtem U, Citro R, Underwood SR, et al. Current state of knowledge on Takotsubo syndrome: a Position Statement from the Taskforce on Takotsubo Syndrome of the Heart Failure Association of the European Society of Cardiology. *Eur J Heart Fail.* 2016; 18:8-27.
2. Nguyen LT, Schelbert EB, Cook SC. Takotsubo cardiomyopathy in an adult woman with repaired tetralogy of Fallot. *World J Pediatr Congenit Heart Surg.* 2016;7:381-4.
3. Derk GR, Aboulhossn J, Reardon LC. Takotsubo cardiomyopathy in a 22-year-old single-ventricle patient. *Tex Heart Inst J.* 2016;43:61-4.
4. Aregullin EO, Garg R, Berman D. Takotsubo cardiomyopathy after transcatheter Edwards Sapien pulmonary valve placement in a patient with tetralogy of Fallot. *Pediatr Cardiol.* 2013;34:1972-5.
5. Pizzino G, Bitto A, Crea P, Khandheria B, Vriz O, Carerj S, et al. Takotsubo syndrome and estrogen receptor genes: partners in crime? *J Cardiovasc Med (Hagerstown).* 2017;18:268-76.