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CONGENITAL HEART DISEASE

CLINICAL CASE

Takotsubo Syndrome in a 47-Year-Old Woman With Repaired Tetralogy of Fallot

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ABSTRACT

Takotsubo syndrome or broken-heart syndrome is a rare form of nonischemic cardiomyopathy characterized by regional systolic dysfunction of the left ventricle without evidence of coronary artery disease or acute plaque rupture. This transient impairment in myocardial contractility leads to symptoms and signs that can mimic a myocardial infarction. We present a case of Takotsubo syndrome in a 47-year-old premenopausal woman with complex congenital heart disease who initially presented with acute onset of shortness of breath and chest tightness after a verbal altercation. Extremely rare cases of Takotsubo syndrome have been described in the congenital heart disease population in premenopausal women. This case emphasizes the need to highlight acquired cardiac disease in patients with adult congenital heart disease as this cohort continues to age. (JACC Case Rep. 2025;30:102804) © 2025 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 47-year-old premenopausal woman with history of tetralogy of Fallot and pulmonary atresia with multiple prior surgical procedures and ultimately complete repair, presented to an emergency department with acute shortness of breath, chest tightness, and 1 episode of nonbloody emesis. Her symptoms started shortly after a verbal altercation with a family member. She was found to have oxygen saturation of 89% on room air, initially requiring bilevel positive airway pressure before being transitioned to 5-L nasal cannula. The rest of her vital signs were stable. She had a loud systolic murmur, clear lungs, and no peripheral edema.

TAKE-HOME MESSAGES

- When an adult patient with congenital heart disease presents with cardiac symptoms, it is imperative to consider common acquired cardiac pathologies as part of the differential diagnosis.
- A thorough work-up must be completed and the Revised Mayo Clinical Criteria should be met to make to an accurate diagnosis of Takotsubo syndrome in a patient.

PAST MEDICAL HISTORY

The patient had a history of tetralogy of Fallot with pulmonary atresia status post initial Blalock-Taussig-Thomas shunt at 16 months of age, a right ventricle to pulmonary artery (RV-PA) conduit at 3 years of age, and surgical ventricular septal defect closure at 4 years of age. She subsequently underwent a RV-PA

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ABBREVIATIONS AND ACRONYMS

CHD = congenital heart disease

ECG = electrocardiogram

EF = ejection fraction

LV = left ventricular

RV-PA = right ventricle to pulmonary artery

TTE = transthoracic echocardiogram

conduit replacement at 8 years of age. She did well for several years and underwent a transcatheter Melody valve implantation in the RV-PA conduit at 42 years of age. She also developed atrial flutter for which she underwent radiofrequency ablation. She had inducible monomorphic ventricular tachycardia on an electrophysiology study leading to an implantable cardioverter-defibrillator.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included the following: acute decompensated heart failure, acute coronary syndrome, acute myocarditis, or an acute pulmonary embolism.

INVESTIGATIONS

An initial 12-lead electrocardiogram (ECG) was notable for an atrial paced rhythm and right bundle branch block with a QRS duration of 144 milliseconds and QTc duration of 471 milliseconds, similar to her prior ECGs. Computed tomography angiography of the chest was negative for pulmonary embolism. Initial laboratory work was notable for high-sensitivity troponin of 211 ng/L that up-trended to 289 ng/L, and N-terminal pro-B-type natriuretic peptide of 2,194 pg/mL. Additional work-up included a transthoracic echocardiogram (TTE) that showed a left ventricular (LV) ejection fraction (EF) of 15% and new wall motion abnormalities of the inferior and anterior lateral walls with severe apical hypokinesis and sparing of the LV base (Video 1). On prior echocardiograms, she had preserved LV EF of 50% to 55%. She was initially given aspirin (81 mg daily) and anticoagulation (dosage included home regimen which was continued of eliquis 5 mg twice daily, diuresed with IV furosemide 40 mg twice daily), and then was managed for acute decompensated heart failure with intravenous furosemide with improvement in her symptoms. Her laboratory work-up the following day showed a down-trending troponin to 190 ng/L, but elevated erythrocyte sedimentation rate to 120 mm/h and C-reactive protein of 57.4 mg/L. Her infectious panel was negative. While on telemetry, the patient experienced paroxysmal atrial fibrillation as shown by 12-lead ECG in **Figure 1**. A follow-up TTE after 24 hours showed improvement in the LV EF to 45% with resolution of apical akinesis (Video 2). A coronary computed tomography examination showed no evidence of coronary artery stenosis and normal LV EF of 60% on the third day after presentation (Video 3). A repeat ECG prior to discharge showed an atrial paced rhythm with similar QRS and QTc values compared with baseline (**Figure 2**).

MANAGEMENT

The patient was found to have transient severe dyskinesis of the left ventricle, regional wall motion abnormalities beyond a single coronary vascular distribution, with moderately elevated troponin. She had no evidence of obstructive coronary artery disease, acute plaque rupture, pheochromocytoma, or myocarditis, thus meeting the Revised Mayo Clinic Criteria for Takotsubo syndrome.[1] After intravenous diuresis, she had rapid improvement in her symptoms with normalization of the LV EF. She was weaned off supplemental oxygen and discharged in stable condition after 3 days on her prehospital medication.

FOLLOW-UP

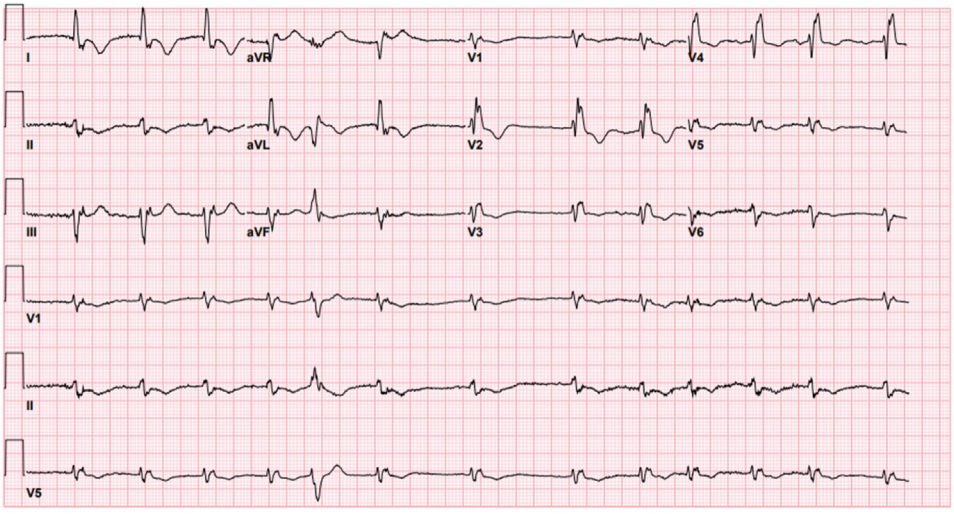
The patient was seen at an outpatient follow-up appointment approximately 2 weeks after discharge. She was found to be feeling overall well and much improved since the hospitalization. Follow-up echocardiogram after 7 months showed markedly improved function with global LV EF of 65% (Video 4).

DISCUSSION

Although the precise etiology of Takotsubo syndrome has yet to be elucidated, it is thought to be at least partially explained by increased sympathetic activity and a catecholamine surge that impairs myocardial function.[2,3] Patients with Takotsubo syndrome commonly present after episodes of significant physical and/or emotional stress. It has been estimated that approximately 2% of patients who present with concern for acute coronary syndrome have Takotsubo syndrome, most commonly seen in postmenopausal women.[4,5]

Advancements in pediatric cardiology and cardiothoracic surgery have extended the average lifespan of patients with congenital heart disease (CHD). However, as this cohort enters middle age, there is limited information on the presentation and management of cardiomyopathies, including nonischemic cardiomyopathies (eg, Takotsubo syndrome). Takotsubo syndrome has not been well described in the CHD population with only rare cases occurring in premenopausal women.[6] Additional research is needed to further elucidate an association, if any, between CHD and Takotsubo syndrome. Although thus far there is little evidence to suggest that pathophysiology may predispose individuals with CHD to

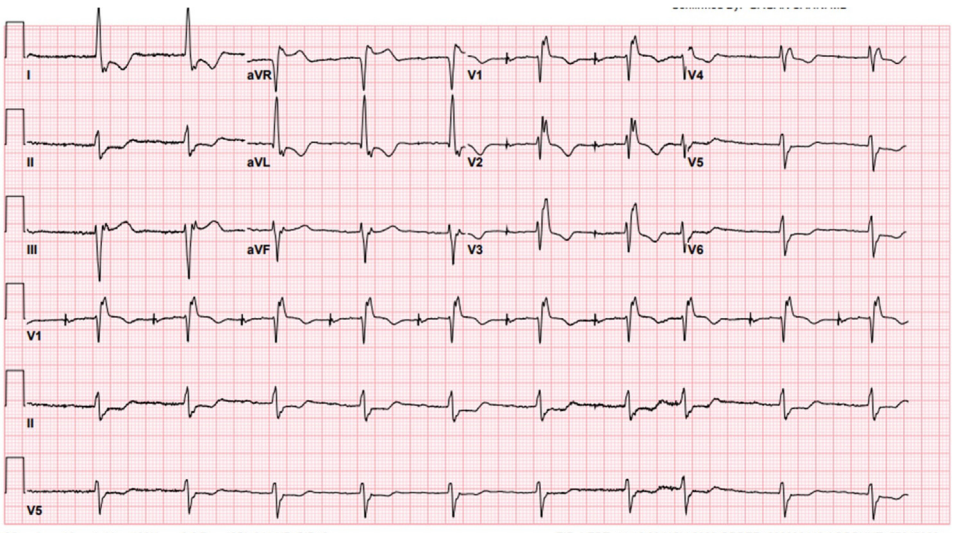
FIGURE 1 12-Lead Electrocardiogram



Twelve-lead electrocardiogram showing atrial fibrillation with a premature ventricular contraction, right bundle branch block, and diffuse T-wave abnormalities. The QRS duration is 154 milliseconds and QTc duration is 500 milliseconds.

an increased risk of having Takotsubo syndrome, it is plausible that patients with CHD have many prominent risk factors that place them at risk for developing the syndrome. For example, evidence suggests that Takotsubo syndrome is associated with a chronic inflammatory state and recent work has shown that CHD demonstrates multiple features of chronic inflammatory disease.[7-9] Furthermore, prior work has identified psychiatric disorders, including anxiety and depression, as common predisposing factors to

FIGURE 2 Repeat 12-Lead Electrocardiogram



Repeat 12-lead electrocardiogram before discharge showing an atrial paced rhythm with premature atrial contraction, right bundle branch block, and diffuse T-wave abnormalities. The QRS duration is 156 milliseconds and QTc duration is 501 milliseconds.

Takotsubo syndrome and are known be more prevalent in CHD compared with the general population.[7,10]

We report a rare case of Takotsubo syndrome in a patient with complex CHD and acute decompensated heart failure with dramatic improvement of the LV EF within 36 hours. Further investigation is needed to determine if patients with CHD are predisposed to nonischemic cardiomyopathies like Takotsubo syndrome particularly at younger ages compared with the general population.

CONCLUSIONS

This rare case highlights that as patients with CHD enter middle age, they will present with acquired cardiac pathologies that are routinely seen in patients without CHD. These patients have to be managed in the context of both acquired cardiac disease and their underlying CHD to improve long-term patient outcomes.

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KEY WORDS congenital, Takotsubo syndrome, tetralogy of Fallot

APPENDIX For supplemental videos, please see the online version of this paper.