Takotsubo Cardiomyopathy
Natural History

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KEYWORDS

• Takotsubo cardiomyopathy • Stress cardiomyopathy • Congestive heart failure

KEY POINTS

- Takotsubo (stress) cardiomyopathy is a recently recognized cardiomyopathy with acute onset, distinctive left ventricular (LV) contraction profile, and a predilection for women older than 50 years. It is often triggered by a stressful event and is typically completely reversible.
- At presentation, takotsubo cardiomyopathy resembles acute coronary syndrome with ischemic electrocardiogram changes and troponin elevation. Abnormal regional LV contraction is in a non-coronary distribution and occurs in the absence of acute coronary artery obstruction.
- A triggering stressful event, either physical or emotional, is frequent but not universal.
- Complications include congestive heart failure, ventricular arrhythmias, left ventricular outflow tract obstruction, and ventricular mural thrombi with the potential for embolization.
- Hospital rate of mortality is low but after hospital survival is less than the general population and rate of recurrence is 5% to 10%. Complete recovery of LV systolic function is a hallmark.

INTRODUCTION

Takotsubo (stress) cardiomyopathy (TTC) is a recently recognized cardiomyopathy with acute onset, characterized by a distinctive left ventricular (LV) contraction profile, a predilection for middle-aged and older women, which is triggered by a stressful event and typically completely reversible. The clinical presentation resembles that of acute coronary syndrome, yet this cardiomyopathy occurs in the absence of significant obstructive coronary artery disease.[1–6] Although uncommon, 5%–10% of women with suspected acute coronary syndrome are ultimately proven to have TTC.[7–10]

The first reports of this cardiomyopathy emerged from Japan in 1991 at which time Dote and colleagues[11] reported 5 patients with a novel acute cardiac condition characterized by distinctive LV dysfunction in the absence of atherosclerotic obstructive coronary artery disease. These and other investigators were intrigued by the unusual end-systolic LV shape (as captured on right anterior oblique left ventriculogram), which resembled the Japanese takotsubo, a pot with a narrow neck and round bottom used for the harvest of the octopus (**Fig. 1**). Consequently, the term takotsubo cardiomyopathy was introduced to describe this condition.[12,13]

For several years, recognition of TTC was confined to Japan and much of the early literature was written in the Japanese language. Beginning in the late 1990s, publications first emerged in the English literature; thereafter, TTC became widely recognized with greater than 1000 reports by 2011.[13–16] The condition is now recognized worldwide and during this process has acquired a variety of additional names (at least 75), the most common being stress cardiomyopathy and apical ballooning syndrome.[16] With this increased attention has come the recognition that TTC has

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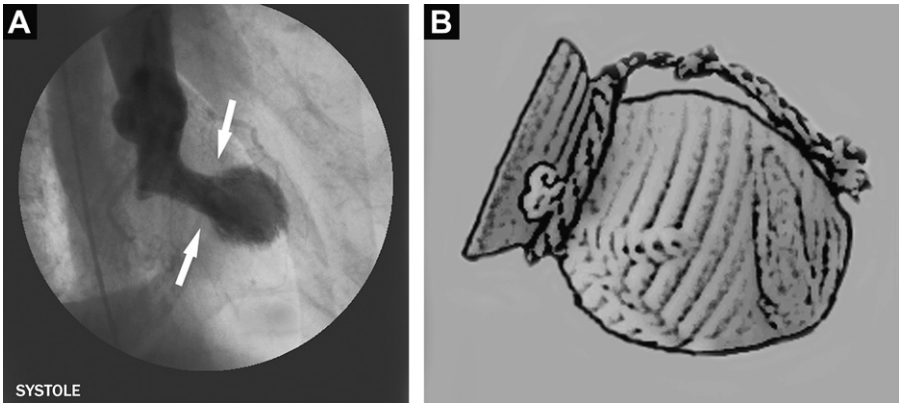

Fig. 1. (*A*) Left ventriculogram (right anterior oblique projection) at end-systole demonstrating classic apical ballooning pattern. Arrows depict demarcation between hypercontractile basal myocardium and dyskinetic ballooning apex. (*B*) The Japanese takotsubo has a shape that resembles the left ventricle.

a much more diverse clinical presentation than initially considered.[2,5,17–20]

ACUTE PRESENTATION

The clinical features of TTC are summarized in **Table 1**. Patients usually present to the emergency department with sudden onset of chest discomfort or shortness of breath, symptoms indistinguishable from acute coronary syndrome. In an important minority, TTC occurs during hospitalization for a noncardiac illness, in which case symptoms may be atypical or absent, and cardiac evaluation is triggered by hypotension, tachycardia, heart failure, abnormal electrocardiogram (ECG), or unexpected troponin elevation.[18] The ECG often shows ischemic changes, most frequently ST-segment elevation. Furthermore, the initial troponin is elevated in 90% of patients. By currently established guidelines, patients with TTC usually meet the definition for acute myocardial infarction.[21]

The clinician may suspect TTC because of its predilection for older women and its usual association with an antecedent stressful event. The average age at onset is 65 to 70 years, although the condition has been described in younger patients, including a 2-year-old girl.[2,17,22] Men comprise only 10% to 15% of patients.[2,5] The inciting stressor (**Table 2**) may be physical (40%–45% of patients), such as sepsis or acute respiratory failure, or emotional (40%–45% of patients), such as anger or grief from a death of a family member.[8,17] Questioning the patient and family is useful because some emotional stressors may be intensely private. It is now recognized that TTC also occurs spontaneously (in the absence of an overt stressor) in up to 30% of patients, consequently the term stress cardiomyopathy does not accommodate all patients.[2,8,17]

DIAGNOSIS

The hallmark is a reversible LV contraction abnormality extending beyond the geographic territory of a single epicardial coronary artery, occurring in the absence of significant obstructive coronary artery disease or acute plaque rupture (**Box 1**). An urgent coronary angiogram with left ventriculogram is necessary to differentiate TTC from acute coronary syndrome. It is not unusual to note reduced epicardial coronary artery flow rate (reduced thrombolysis in myocardial infarction frame count) despite absent obstruction, likely signifying microvascular dysfunction.[23–25] Until the diagnosis is verified, it is reasonable to treat the patient with suspected TTC as you would a patient with acute coronary syndrome (aspirin, heparin, β-blocker, and clopidogrel). A 2-dimensional echocardiogram is useful to assess LV outflow tract obstruction, mitral valve regurgitation, LV and right ventricular (RV) thrombus, and RV dysfunction.

LV AND RV CONTRACTION PATTERNS

Three distinct patterns of abnormal LV contraction (ballooning) characterize TTC.[2,8,17] The LV apical ballooning pattern was the first described pattern, currently the most common and observed in 70% to 80% of patients (**Fig. 2**). More recently, the midventricular ballooning pattern has been defined[26] and is present in a substantial minority of patients (20%–30%) (**Fig. 3**). A third pattern, inverted ballooning, is rarely encountered (1%–2%) and seems to occur predominantly in younger women (**Fig. 4**).[27]

Each of these LV contraction patterns is, in general, unlike that caused by obstructive coronary artery disease and can be recognized by