Broken heart syndrome

by Kate O’Donovan

Broken heart syndrome, also known as takotsubo cardiomyopathy, apical ballooning syndrome and ampulla cardiomyopathy was first described in Japan in 1991.1 A takotsubo is a round-bottomed narrow necked fishing pot commonly used in Japan to capture octopus. The term takotsubo cardiomyopathy refers to the shape of the left ventricle where the apical segment of the left ventricle balloons out while the base of the heart has preserved shape and function giving it a similar appearance to the Japanese fishing pot.

This form of cardiomyopathy is an acute, transient reversible state of left ventricular dysfunction in the absence of coronary artery disease and is often triggered by profound psychosocial distress or physical stress. Stressors that are commonly associated with the development of symptoms are described in Table 1. In addition to these stressors, physical conditions or diagnoses of a life limiting illness such as cancer may precipitate its occurrence.

The pathophysiology underpinning this cardiomyopathy is largely unknown. Under normal physiological conditions a stressor stimulates the sympathetic nervous system, which triggers the release of catecholamines (adrenaline and noradrenaline). The response is an increase in heart rate and vasoconstriction, which increases blood pressure. But in takotsubo cardiomyopathy Wittstein2 demonstrated that levels of catecholamines were significantly higher in comparison to other causes of raised adrenaline and noradrenaline such as acute myocardial infarction.

It is thought that these high circulating levels of catecholamines have a direct toxic effect on the myocardium, which results in myocardial stunning and wall motion abnormalities (hypokinesia, akinesia). The hallmark response to myocardial stunning is the inappropriate responsiveness of the left ventricular apex to the catecholamine overload, in that the apex dilates and relaxes leading to a reduction in contractility and thus reduced cardiac output. This dilation of the apex is also associated with hypercontractility of the left ventricular base. As the heart rate increases in response to the catecholamines, diastole is shortened. Consequently due to shortened diastolic time the ventricles do not fill adequately for contraction, thus reducing the cardiac output further. In addition, coronary artery perfusion is suboptimal and may result in chest pain.3,4

**Signs and symptoms**

The patient normally presents to hospital within a few hours of the identified stressor and onset of symptoms.3 The majority of patients have no prior cardiac history and present with moderate to severe substernal chest pain and/or dyspnoea. Although the most commonly associated presenting symptom is chest pain its characteristics are not well defined. Other symptoms include diaphoresis, nausea, vomiting, malaise and palpitations. The degree of symptom severity varies widely.7

In most cases patient presentation is stable but approximately one third of patients will present more acutely with symptoms of decreased cardiac output, such as pulmonary oedema, hypotension and cardiogenic shock.2 The signs and symptoms often mimic those of acute coronary syndrome making the differential diagnosis challenging. Derrick7 describes the clinical findings associated with takotsubo cardiomyopathy in Table 2.

**Investigations**

In the majority of cases chest pain is the most commonly presenting symptom and therefore common investigations include an electrocardiogram (ECG), cardiac biomarkers, imaging such as echocardiography and cardiac catheterisation. ECG findings are variable and cannot be used to diagnose takotsubo cardiomyopathy. There are a variety of ECG findings from normal to non-specific ST segment and T wave deviations and may also include right or left bundle branch block. The most common ECG finding is ST segment elevation in the precordial leads especially leads V2-V5 and its incidence has been reported in approximately 50-60%. The literature describes four ECG stages:8

1. Immediate ST elevation (Stage 1)
2. Initial T wave inversion from days 1-3 (Stage 2)
3. Transient improvement in T wave inversion from days 2-6 (Stage 3)
4. A second deeper T wave inversion with QT prolongation persisting for at least two months (Stage 4).

---

**Stressors associated with takotsubo cardiomyopathy**

<table>
<thead>
<tr>
<th>Stressor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Death of a relative or spouse</td>
</tr>
<tr>
<td>Domestic abuse or dispute</td>
</tr>
<tr>
<td>Surprise party</td>
</tr>
<tr>
<td>Car accident</td>
</tr>
<tr>
<td>Natural disasters</td>
</tr>
<tr>
<td>Severe pain/discomfort</td>
</tr>
<tr>
<td>Work-related stress</td>
</tr>
<tr>
<td>Public speaking</td>
</tr>
<tr>
<td>Court appearance</td>
</tr>
<tr>
<td>Being held at gunpoint</td>
</tr>
<tr>
<td>Physical exertion</td>
</tr>
<tr>
<td>Financial difficulties</td>
</tr>
<tr>
<td>Gambling</td>
</tr>
</tbody>
</table>
The majority of the literature reviewed comment that the QT interval may also lengthen. The normal QT interval is between 0.36 and 0.45 seconds. The QT interval represents the amount of time it takes the ventricles to depolarise and repolarise. If the QT interval is prolonged there is a predisposition to ventricular arrhythmias.

As with the management of chest pain cardiac biomarkers troponin T and I are reserved. Although the troponin may be elevated it will not rise to the same level as in acute myocardial infarction. Echocardiography (ECHO) findings may aid in the diagnosis of takotsubo syndrome. Findings demonstrate a reduced left ventricular ejection fraction associated with extensive hypokinesis (slow movement) or akinesis (no movement) of the left ventricular apex in conjunction with compensatory hyperkinesis (dynamic movement) of the left ventricular base. This pattern resembles the fishing pot used to catch octopus – the takotsubo. These findings are transient with ventricular function returning to baseline over a period of days to weeks. It is recommended that a repeat ECHO is performed within days to weeks after the acute event to confirm that left ventricular function is recovering.

Cardiac catheterisation, in particular left ventriculogram, will confirm the ECHO findings, revealing a hyperkinetic left ventricular base and a hypokinetiakinesis left ventricular apex. Coronary angiography illustrates no obstructive coronary artery disease thus supporting the diagnosis of takotsubo cardiomyopathy.

The Mayo Clinic developed diagnostic criteria to aid in the diagnosis of this cardiomyopathy:

- New ECG findings (not evident on previous ECGs or are acute changes) such as T wave inversion or ST segment changes or elevated troponin
- Absence of any obstructive coronary artery disease or angiographic evidence of acute plaque rupture
- Transient hypokinesis, akinesis or dyskinesis of the left ventricular apex and mid ventricular segment including wall motion abnormality
- All other possible causes of the changes have been ruled out (head trauma, intracranial bleeding, phaeochromocytoma, coronary occlusion, myocarditis, hypertrophic cardiomyopathy).

This last point is important to consider in when thinking about differential diagnosis, as ventricular wall abnormalities are commonly associated with these other conditions. Therefore confirming the absence of coronary artery disease and that this acute episode was preceded by a psychosocial stressor confirms the diagnosis of takotsubo cardiomyopathy. For the clinical diagnoses to be made all four criteria must be present.

**Prevalence, prognosis, mortality**

The exact incidence of this form of cardiomyopathy is unknown but it is recognised that the prevalence is increasing with it affecting six to nine times more women than men and may account for 1-2% of patients who present with acute chest pain, ST segment elevation and a rise in cardiac markers. It remains unclear why more women than men are affected but the literature identified those women are often older and post menopausal, average age ranging between 60 and 80 years.

Although the cardiomyopathy is transient and left ventricular function recovers to patient’s baseline mortality rates vary between 2-6%.

Several associated complications are recognised. The majority of complications occur during the acute phase with the reported complication rate approximately 19% with left ventricular failure/pulmonary oedema being the most commonly occurring. Other complications include cardiogenic shock (rare), left ventricular outflow tract obstruction, ventricular arrhythmias, transient complete heart block and left ventricular mural thrombus. More severe complications include mechanical issues such as left ventricular wall rupture and/or ventricular septal rupture. Late complications are unusual because the cardiomyopathy is transient and short lived with ventricular function returning to baseline.

**Patient management**

Specific guidelines regarding management do not exist and in the majority of cases management is based on the management of acute coronary syndromes such as the European Society of Cardiology guidelines. These guidelines include the use of oxygen, aspirin, low molecular weight heparin, angiotensin converting enzyme inhibitor (ACE inhibitor) and a β-blocker. Patients are probably best managed initially by emergent cardiac catheterisation to rule out obstructive coronary artery disease. In relation to the role of β-blockers, some of the literature recommend continuing β-blockade on a long term basis to protect against catecholamine sensitivity which may predispose to recurrence.

In addition to standard treatment, heparin and warfarin may be used to treat or prevent left ventricular apical thrombus, which is a risk of significant left ventricular dysfunction. Apical thrombi form because of a stasis of blood in the akinetic portion of the apex. Anti-coagulation is discontinued once ventricular function has recovered.

Supportive treatment is employed for acute complications such as anti-arrhythmic drugs for ventricular arrhythmias, diuretics for left ventricular failure/pulmonary oedema. In cases of severe left ventricular dysfunction intra aortic balloon pump counterpulsation may be used to improve patient haemodynamics. Inotropes and vasopressors are used with caution as they may induce additional catecholamine stimulation thus potentially worsening ventricular function.

**Nursing implications**

Takotsubo cardiomyopathy is an emerging important health issue involving postmenopausal women without obstructive coronary artery disease, and often without the classic risk factor profile. Thorough history taking in this patient population is vital in determining if a significant psychosocial or physical stressor preceded symptom onset, thus aiding in the diagnosis. Nursing
Care involves cardiac and haemodynamic monitoring, providing supportive measures and watching for complications. Goals of care are similar to those of acute coronary syndrome: alleviating pain, reducing anxiety, preserving myocardial function and preventing/treating complications.

Continuous cardiac monitoring is essential for detecting arrhythmias and/or conduction abnormalities as well as observing for prolongation of the QT interval and ST segment T wave changes. Cardiovascular and respiratory assessment should be undertaken regularly or as acuity determines to detect for respiratory distress or the development of pulmonary oedema. If diuretics, and/or ACE inhibitors, are prescribed, daily monitoring of renal function is necessary so that electrolyte imbalances are detected and treated accordingly. If haemodynamic support is provided nursing care is adapted to the specific type of support employed.

Psychological support in the form of education and counselling is vital to the holistic care of this patient cohort, with referral to the relevant resources that can aid in coping strategies or bereavement counselling. Education consists of informing patients and families about the nature of the cardiomyopathy, including its reversibility and low rate of recurrence, which will also provide reassurance. This can be continued in cardiac rehabilitation and where necessary referral to a stress reduction clinic may be made.

Takotsubo cardiomyopathy is transient reversible form of left ventricular dysfunction that is commonly preceded by a significant emotional stressor. Signs and symptoms mimic that of acute coronary syndrome but angiography demonstrates no obstructive coronary artery disease. This condition should be considered as a possible diagnosis in patients, especially post-menopausal women, whose onset of symptoms coincides with some type of a stressor. Further research is required into the reason why it affects women more than men and also into the exact pathophysiology underpinning this cardiomyopathy.

Kate O’Donovan is course co-ordinator for the postgraduate diploma in cardiovascular nursing in the Mater Hospital, Dublin

References