Broken heart syndrome

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Takotsubo was first described in Japan in 1990, but subsequently it was described globally and in 2013 there were 1879 publications on the subject. The main characteristic is transient reversible systolic dysfunction of the left ventricle and 90% of the cases are in women.

The condition is known by many names and the nomenclature is not universally accepted. It is also known as the 'broken heart syndrome' or apical ballooning syndrome or stress-induced cardiomyopathy.

The name Takotsubo means 'octopus pot' in Japanese and describes the shape of the ventricle. The American Heart Association has labelled it as an acquired cardiomyopathy. The prevalence is not known but it is estimated to make up 2% of patients presenting with acute coronary syndrome.

TYPES

There are at least four types of Takotsubo syndrome:

1. The common classic one with left ventricle apical ballooning.
2. Mid-ventricular type.
3. Basal type.
4. Focal type.

When this condition recurs it can present with a different type each time.

CLINICAL PRESENTATION

The classic clinical presentation is that of acute coronary syndrome but the coronary arteries are open on angiography and the area of akinesia or hypokinesia of the left ventricle does not match any epicardial coronary artery.

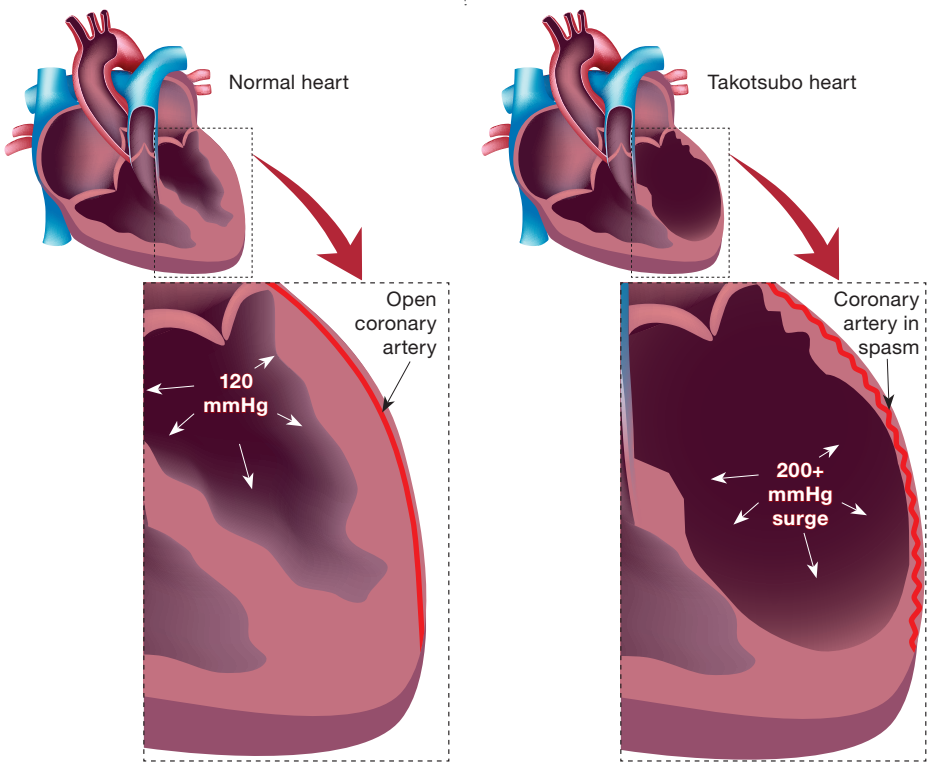
It can also present without any typical symptoms or signs and are discovered incidentally during hospitalisation for another reason such as an operation and then diagnosed because of an abnormal electrocardiography or raised troponins. The clinical presentation is thus very diverse from non-specific to life-threatening with shock.

TRIGGER FACTORS

The unique feature is that there is a preceding severe physical or emotional trigger. Numerous physical triggers have been identified such as surgery, any critical illness, neurological disease and many others. These trigger factors may, however, be absent.

CARDIAC BIOMARKERS

Troponin and creatinine kinase (CK) levels are elevated but only mildly so. Brain natriuretic peptide (BNP) levels are also increased. Despite these raised biomarkers there are no specific biomarkers to diagnose Takotsubo syndrome.



ELECTROCARDIOGRAPHY

Patients can present with S-T segment elevation, S-T segment depression, T-wave inversion, non-specific ECG abnormalities and even a normal ECG. It can also present with QTc prolongation which can lead to malignant ventricular arrhythmias.

INVASIVE IMAGING

Coronary angiography with a left ventricle angiogram is the gold standard for excluding acute coronary syndrome due to atherosclerotic coronary artery disease and for diagnosing Takotsubo syndrome. Echocardiography is also helpful for diagnosing wall motion abnormalities. The role of MRI still needs to be clarified.

PATHOPHYSIOLOGY

The exact pathophysiology remains elusive, but patients have elevated plasma levels of catecholamines sug gesting a role for hypothalamic-pituitary-adrenal axis and stress neuro hormones. Precisely how these neuro-hormones affect the heart is still unclear but there is necrosis of the myocardium on histology.

Epicardial coronary artery spasm has also been proposed as an underlying mechanism. Micro-vascular dysfunction of the myocardium may also play a role.

MANAGEMENT

For patients who survive the acute phase, the long-term prognosis is good and in the majority of cases the left ventricle can recover in four to eight weeks but some may need 12 months.

There are no randomised controlled trials on the treatment of Takotsubo and treatment strategies are therefore based on clinical judgement. Aspirin and heparin can be used in the acute phase. Positive inotropic drugs could increase the ventricle outflow tract and worsen the shock. Chronic therapy with beta-alpha blockers can be used and also ACE-Inhibitors or ARB's. All acute complications are treated as they arise such as heart failure, shock, arrhythmias etc.

Much more research is needed to better understand this condition and to develop more specific therapy.

References available on request. SF