Takotsubo cardiomyopathy:
## Presentation, management and outcome

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Introduction

- Takotsubo cardiomyopathy was first described **in Japan in 1990 by Sato et al.**.

- American Heart Association (AHA) classification of cardiomyopathy (2006)

- Also known as broken heart syndrome, apical ballooning syndrome and stress induced cardiomyopathy

- It is generally characterized by **transient** systolic dysfunction of the apical and mid segments of the LV that mimics myocardial infarction in absence of obstructive CAD
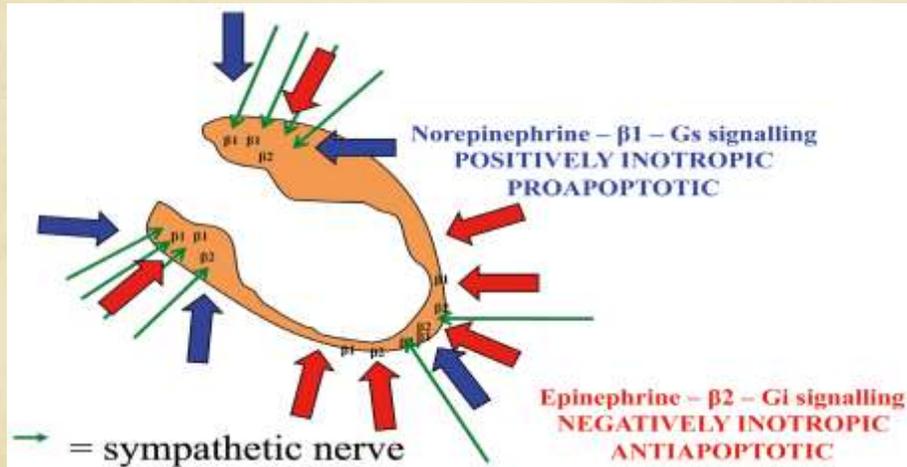
The term **"takotsubo"** was taken from the Japanese name for an **octopus trap pot.**
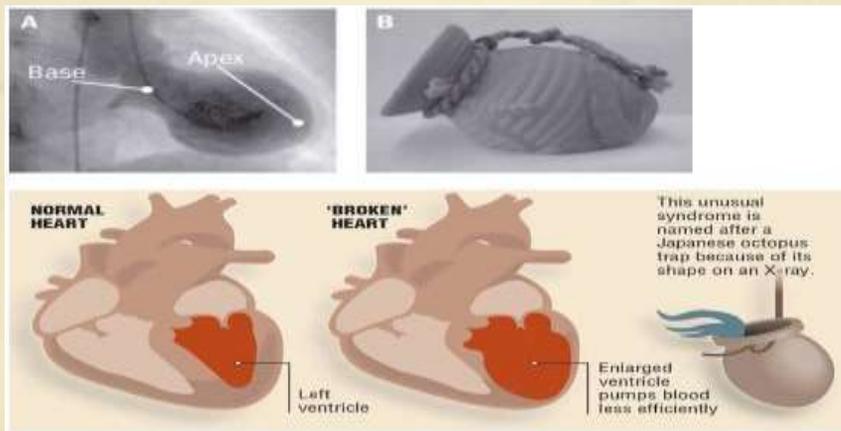

Pathophysiology

○ The exact mechanism of this disorder is **not well understood**.

○ Postulated mechanisms:

1) Excessive sympathetic stimulation

2) Metabolic abnormalities

3) Microvascular dysfynction

Pathophysiology (contd.)
Excessive sympathetic stimulation

Norepinephrine – β1 – Gs signalling
POSITIVELY INOTROPIC
PROAPOPTOTIC

Epinephrine – β2 – Gi signalling
NEGATIVELY INOTROPIC
ANTIAPOPTOTIC

→ = sympathetic nerve

Schematic of the sympathetic innervation of the myocardial regions (Lyon et al , Nat Clin Pract Cardiovasc Med 2008)

Pathophysiology (contd.)
## Metabolism abnormalities
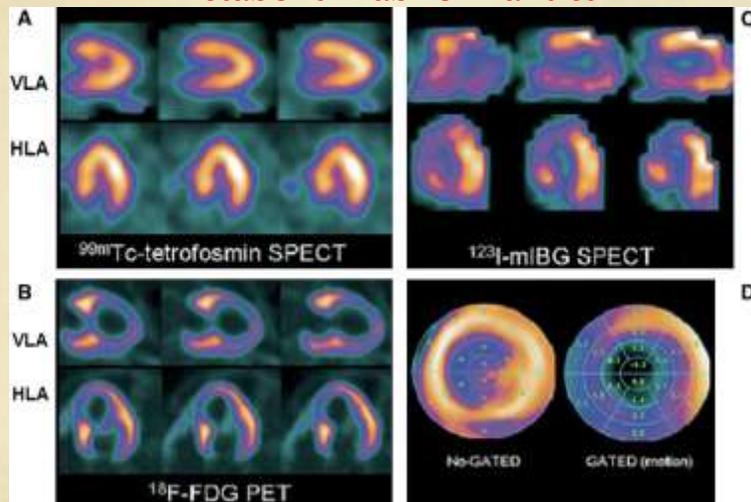
Myocardial perfusion images with Tc-99 m tetrofosmin SPECT (A), F-18 FDG PET images (B), and I-123 MIBG images (C) in a patient with TC. The metabolic and sympathetic images are abnormal, but the perfusion images are normal. Cimarelli et, Nucl Cardiol 2008

Pathophysiology (contd.)
Microvascular dysfunction

○ Reduction in coronary flow reserve

○ Decrease in time of diastolic velocity deceleration

Types of Takotsubo
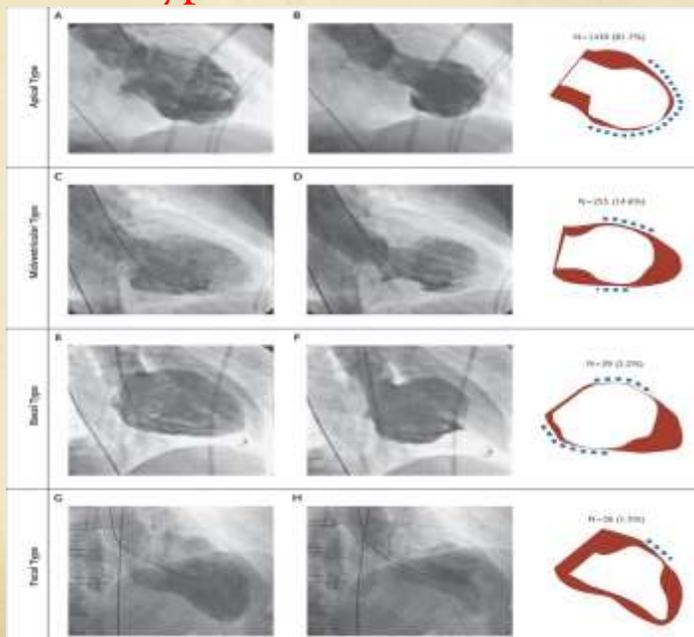
Incidence

- 2.2% of patients admitted with diagnosis of acute MI

- 2% of patients admitted with decompensated heart failure

- 0.02% of all hospitalization

Clinical presentation
Demographics

- Sex : *Female predominance (80-100%)

- Age: *Postmenopausal (62-76 years)

    *5-11% under age of 50 years

- Triggers : * Physical stress (36%)

    * Emotional stress (27.7%)

    * 1/3 of cases no apparent trigger.

Clinical presentation (contd.)
Symptoms and Signs

○ Chest pain (76%)

○ Dyspnea (47%)

○ Syncope (7.7%)

○ Arrhythmia

○ Cardiac arrest
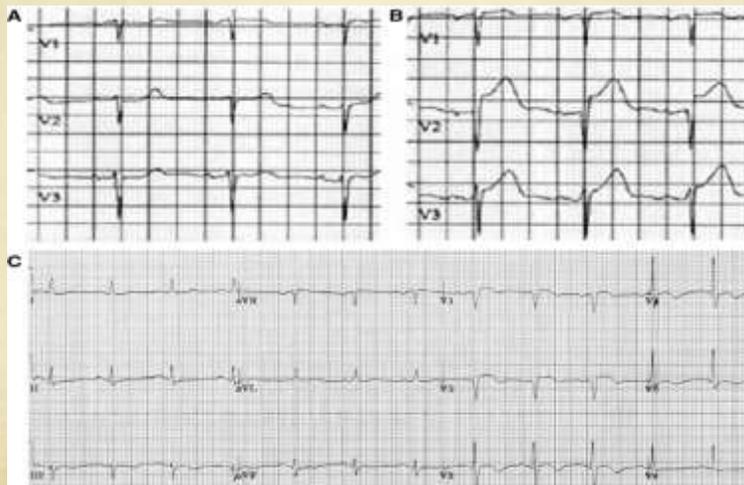
Investigations

○ **ECG**:

- Diffuse T wave inversion (97%)

- ST segment elevation (68%)

- Q waves (27%)

- ST segment depression (10%)

- Conduction abnormalities

- QT prolongation

Investigations
ECG (contd.)

- **Phases of ECG changes** :

- Phase 1: ST segment elevation (acute phase)

- Phase 2: T wave inversion (day 1-3)

- Phase 3: transient improvement of T wave inversion (day 2-6)

- Phase 4: Giant T wave inversion and QT prolongation (tell recovery)

Investigations
ECG (contd.)

Investigations (contd.)

○ **Biomarkers** :

• Cardiac Troponin (86%)

• Creatine kinase- MB (74%)

• Plasma B-type natriuretic peptide (BNP) and N-terminal pro-BNP(NT-proBNP)

Investigations (contd.)

○ **Imaging** :

• Typical wall motion abnormalities

• Assessment of LV systolic function

• Detection of RV involvement (30%)

• Detection of LVOT obstruction

• Complications (LV thrombus, LV free wall rupture)

• Absence of late gadolinium enhancement (LGE) by cardiac MRI

Investigations (contd.)

- **<u>Coronary angiography</u> :**

- Non obstructive coronary artery disease (<50% stenosis)

- Obstructive coronary artery disease (10%)

Investigations (contd.)

- **<u>Left ventriculography</u> :**
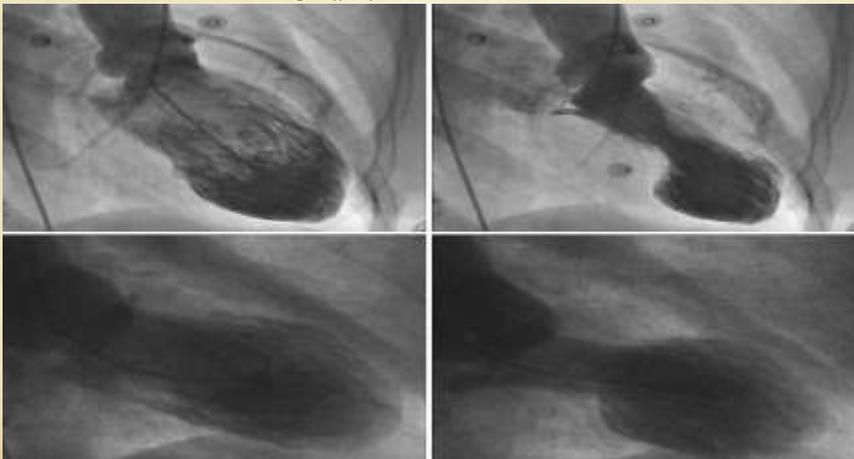
Diagnosis

○ **Mayo Clinic diagnostic criteria**:

1. **Transient** hypokinesis, akinesis or dyskinesis of the **left ventricular mid** segments *with or without* apical involvement. The *regional wall motion abnormalities* **typically extend beyond** a single epicardial coronary distribution. A stressful trigger is **often, but not always** present.

2. **Absence** of obstructive coronary disease or angiographic evidence of acute plaque rupture.

3. **New** electrocardiographic abnormalities (either ST-segment elevation and/or T wave inversion) **or modest** elevation in cardiac troponin.

4. **Absence** of pheochromocytoma or myocarditis.

Complications

○ **Congestive heart failure** (20%)

○ **Cardiogenic shock**

○ **Apical thrombus** (5.3%)

○ **Ventricular rupture**

○ **Arrhythmias:**

1. Life threatening ventricular arrhythmia (8.6%)

2. AV block

Treatment

○ **General principles of therapy :**

1. Initial management should be directed towards myocardial ischemia (Dual anti-platelets , anti-thrombotics, nitrates, statins , ß blockers).

2. IV diuretics in patients with heart failure and volume overload

3. Once the diagnosis of Takotsubo cardiomyopathy has been made >>>> standard medications for left ventricular systolic dysfunction (ACEI/ARBs, ß blockers, diuretics)

4. The appropriate duration of therapy is not known

5. There is no consensus regarding long-term management (??ACEI/ARBs and ß blockers)

Treatment (contd.)

○ **Cardiogenic shock :**

• First step is to rule out left ventricular outflow tract (LVOT) obstruction

1. **No LVOT obstruction:**

• IV inotropic agents and vasopressors (dopamine, dobutrex)

• Mechanical circulatory support (Intra-aortic balloon counter pulsation)

• Inotropic agents and IABP may cause LVOT obstruction >>>> Echocardiography to rule out such adverse effects if there is hemodynamic deterioration

Treatment (contd.)

○ **Cardiogenic shock (contd.) :**

2. **LVOT obstruction:**

• Discontinue IV inotropic agents and nitroglycerine immediately

• ß blockers (in the absence of severe heart failure)

• IV fluids (in absence of significant pulmonary congestion)

• Phenylepherine (IV fluids and ß blockers intolerance)

Treatment (contd.)

○ **Thromboembolism:**

• It is recommended approximately **three months of anticoagulation** if intraventricular thrombus *is detected.* The duration of anticoagulation may be modified based on the rate of recovery of cardiac function and resolution of the thrombus.

• For patients **without thrombus** but with severe left ventricular dysfunction, It is suggested that anticoagulation until akinesis or dyskinesis has resolved or for three months, whichever is shorter.
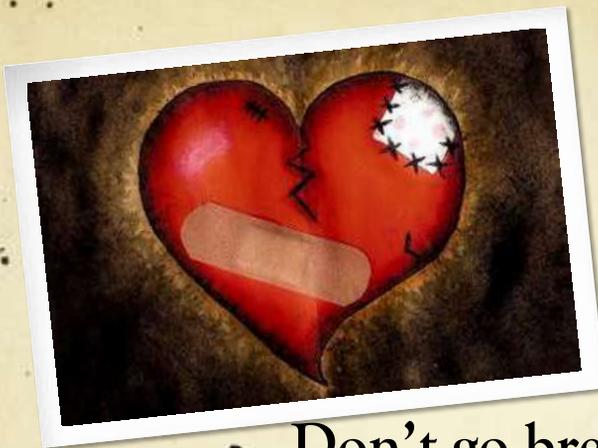
Treatment (contd.)

- **Arrhythmias**

- Should be managed on case by case bases

- Devices implantation is not justified given the reversible nature of the disease

- Correct risk factors of QTc prolongation (hypokalemia, hypomagnesemia, bradycardia, anti-arrhythmic drugs)

- Werabale cardioverter –defibrillators for life threatening ventricular arrhythmia tell recovery.

- Temporary pacemaker implantation if AV block associated with hemodynamic instability or marked QTc prolongation

Prognosis

- In-hospital mortality rates (0- 8%).

- Recurrence rate (0-15%)

- 30 days MACCE (7.1%).

- Long term outcome:
- Death rate from any cause (5.6% per patient-year)
- MACCE (9.9% per patient -year)

- Men have worse outcome than women

Take Home Messages

- Takotsubo cardiomyopathy is currently a diagnosis of exclusion.

- Full understanding of the pathophysiology of takotsubo cardiomyopathy is limited

- There is no specific therapy for takotsubo cardiomyopathy

- Prognosis is generally favorable with complete recovery of LV function but early detection of complications and prompt management are crucial

- Data from randomized trials is needed

Don't go breaking your heart