



# ACC.14

TCT@ACC-12 | Innovation in Intervention

A707

JACC April 1, 2014

Volume 63, Issue 12



## FIT Clinical Decision Making

### A CASE OF CONSTRICTIVE PERICARDITIS IN A PATIENT WITH GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S GRANULOMATOSIS)

Poster Contributions

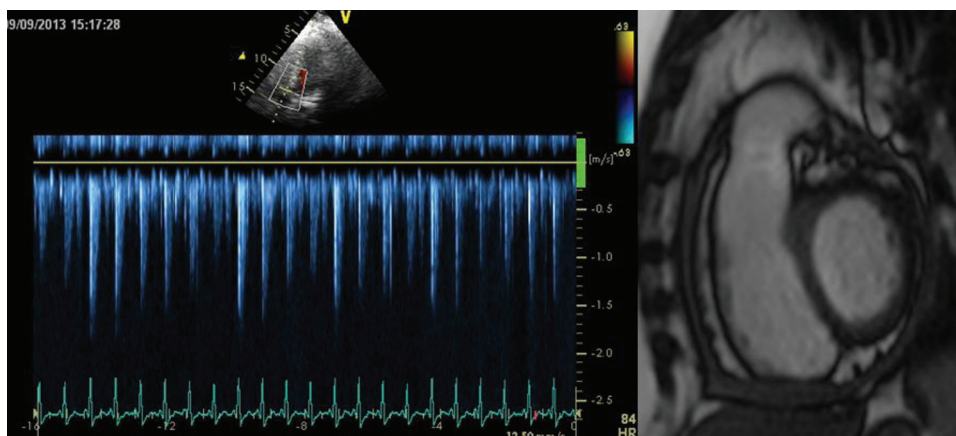
Hall C

Monday, March 31, 2014, 9:45 a.m.-10:30 a.m.

Session Title: FIT Clinical Decision Making: Valvular and Pericardial Disease

Abstract Category: Pericardial Disease

Presentation Number: 1246-07

Authors: *Victoria M. Robinson, Nikhil V. Joshi, Alan G. Japp, Peter A. Henriksen, The Edinburgh Heart Centre, Edinburgh, United Kingdom***Background:** Granulomatosis with polyangiitis (GPA) is a rare vasculitis that can involve cardiac structures.**Case:** A 43 year old lady presented with progressive exertional dyspnoea and orthopnoea. She was receiving treatment with rituximab and corticosteroids for c-ANCA positive GPA. Physical signs included a Cushingoid appearance, extensive peripheral oedema and an elevated JVP.**Methods:** Echocardiography demonstrated good biventricular systolic function, a diastolic septal bounce, dilated atria, exaggerated respiratory variation of Doppler forward flows and a normal E'. CTPA revealed small pleural effusions but no evidence of pulmonary embolism, GPA-related lung disease or pericardial calcification. Cardiac MRI demonstrated pericardial thickening (7mm), and inspiratory septal flattening with no features of intrinsic myocardial disease, corroborating the working diagnosis of constrictive pericarditis. Cardiac catheterisation confirmed typical constrictive haemodynamics. Cyclophosphamide therapy was considered but pericardial biopsies displayed established fibrosis only, with no active inflammation.**Results:** Symptoms persisted despite diuretic therapy and the patient was referred for pericardial stripping.**Conclusions:** This case highlights the need to consider constrictive pericarditis as a potential cause of heart failure with a normal ejection fraction and the utility of multimodality investigation to secure the diagnosis and to guide treatment.

**Exaggerated respiratory variation of LVOT Doppler velocities on transthoracic echocardiogram (left) and circumferential pericardial thickening on cardiac MRI (right).**