A 72-year-old gentleman presented with right heart failure (RHF) from tricuspid insufficiency. He also noted episodes of diarrhoea and flushing. Echocardiogram showed severe tricuspid regurgitation (TR) with thickened tricuspid leaflets (Panel A). The subcostal view revealed a hepatic mass, which, following biopsy and evaluation of 5-hydroxytryptamine, was confirmed as metastatic carcinoid tumour (Panel B).

Carcinoid heart disease (CHD), namely TR, is the initial presentation of carcinoid tumours in up to 20% of patients. Serotonin, by activating the 5-HT(2B) receptor, induces fibrous tissue deposition on the endocardial surface of valvular cusps and cardiac chambers. Pulmonary vascular endothelial cells metabolise serotonin protecting the left heart chambers.

Clinicians need a high index of suspicion for CHD in patients with TR without pulmonary hypertension or core pulmonale. Carcinoid heart disease heralds a decline in clinical outcome with a 3-year survival rate of 31%, half that of patients without CHD. Advanced symptoms, New York Heart Association class III or IV, have a median survival of 11 months.

Echocardiography provides diagnostic and prognostic utility. Thickening of the tricuspid valve leaflets and sub-valvular apparatus leads to incomplete coaptation and regurgitation. The continuous wave Doppler profile of TR shows a characteristic dagger-shaped spectrum with an early peak pressure and rapid decline (Panel C). Pulmonary involvement should be sought as concomitant pulmonary stenosis may exacerbate RHF.

Medical management includes loop diuretics for RHF and somatostatin analogues to reduce circulating levels of serotonin. Surgical management may provide definitive treatment by correcting the cardiac lesion and removing the carcinoid tumour.
Panel A. Transthoracic echocardiography from apical four chamber view shows tricuspid regurgitation.

Panel B. Metastatic carcinoid liver mass (single arrow).

Panel C. Continuous wave Doppler display of tricuspid regurgitation (single arrow).