

Carcinoid heart disease: the winking heart

A 70-year-old woman with a metastatic neuroendocrine tumour presented with heart failure. A 2D transthoracic echocardiogram (TTE) showed thickened, retracted and fixed tricuspid valve (TV) leaflets with free-flowing tricuspid regurgitation (figure 1A, online supplementary videos 1 and 2), and thickened and retracted pulmonary valve (PV) cusps with severe pulmonic regurgitation (figure 1B, online supplementary videos 3 and 4). A 3D TTE en face view of the mitral valve (MV) and TV from the ventricles showed a wide-open fixed TV, while the MV opens and closes normally, giving rise to an image that simulates the heart

winking at you (figure 1C, online supplementary video 5). A 3D TTE in long-axis view (upper) and en face view (lower) of the PV showed a mildly stenotic pulmonary valve, with a diastolic regurgitant orifice and reopening in telediastole (figure 1D, online supplementary video 6). These findings were confirmed with continuous Doppler and colour M-mode imaging (figure 1E). High telediastolic right ventricular pressure secondary to both regurgitations, shown by the severe dilatation of the inferior vena cava and abrupt mid-diastolic ending of pulmonary regurgitation (figure 1E), can explain the diastolic ejective pulmonary flow that reopens the PV. The rare carcinoid tumours release serotonin that can produce fibrotic endocardial plaque in the right cavities with thickening, retraction and severe regurgitation of right heart valves.¹ More than 50% of cases develop carcinoid heart disease and this is the major cause of death. A TTE allows detailed and quantitative

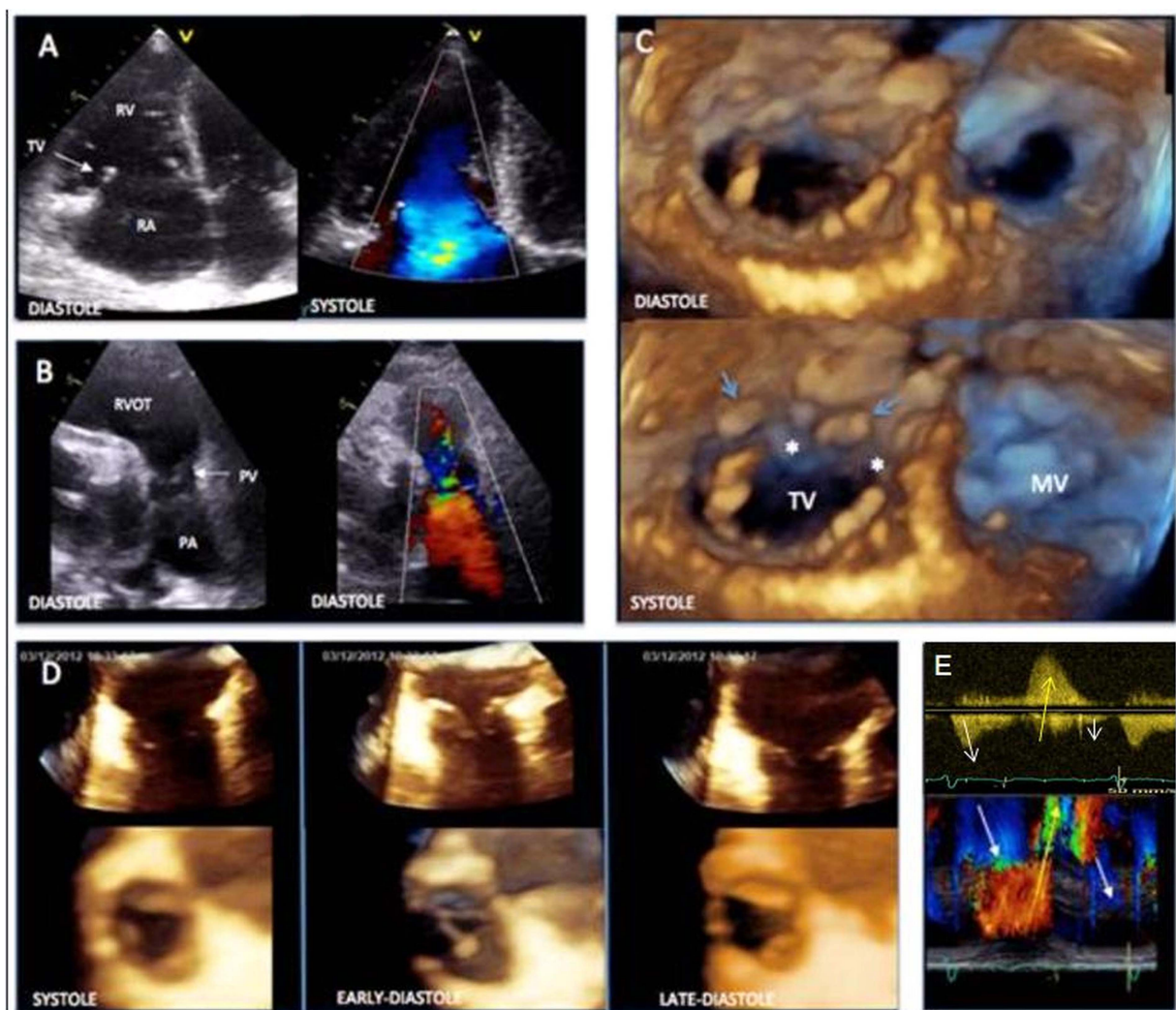


Figure 1 (A, left) A 2D transthoracic echocardiogram (TTE) from a four-chamber view focusing on the right chambers in diastole, showing thickened, retracted and fixed tricuspid valve (TV) leaflets. (A, right) A 2D TTE with colour Doppler focusing on the right ventricle, showing free-flowing tricuspid regurgitation in systole. (B, left) A 2D TTE from a parasternal short-axis view focusing on the pulmonary valve (PV), showing thickened and retracted PV cusps. (B, right) A 2D TTE with colour Doppler from the parasternal short-axis view focusing on the PV, showing severe pulmonic regurgitation. (C) A 3D TTE showing an en face view of the mitral valve (MV) and TV from the ventricles. In diastole (C, top) both valves are opened, and in systole (C, bottom) the TV is fixed opened, showing the absence of parts of the tricuspid leaflets (*) and prominent echogenic areas suggestive of carcinoid deposits (arrow), while the MV closes normally. (D) A 3D TTE showing the PV in long-axis (top) and short-axis (bottom) views. The opening of the PV cusps is limited in systole, does not coapt in diastole and opens again in late diastole. (E) Continuous Doppler and colour M-mode imaging through the PV showing ejective flow in systole (first white arrow), regurgitant flow with abrupt ending in mid-diastole (yellow arrow) and telediastolic ejective pulmonary flow that reopens the PV (last white arrow).

morphofunctional analysis,² and a 3D TTE provides a more graphic presentation than conventional 2D TTE.

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