

## CLINICAL VIGNETTE

# Heart failure manifestation of carcinoid tricuspid dysfunction

Katarzyna Starzyk<sup>1</sup>, Iwona Gorczyca<sup>1</sup>, Beata Wożakowska-Kapłon<sup>1,2</sup>

<sup>1</sup>1<sup>st</sup> Cardiology Clinic, District Hospital, Kielce, Poland

<sup>2</sup>Jan Kochanowski University, Medical Faculty, Kielce, Poland

Neuroendocrine neoplasms (NENs) originate in the small intestine, pancreas, or large intestine. Hormonally active NENs secrete substances (serotonin, kinins, prostaglandins, and histamine) causing symptoms known as carcinoid syndrome. The symptoms occur if biogenic amines are secreted directly to the systemic circulation, most often with hepatic metastases. Hedinger syndrome is a heart disease characterised by plaque-like deposits of fibrous tissue, localised mainly on the tricuspid and pulmonary valves. The prognosis depends on the stage of the disease; according to European data, the five-year survival rate in those with liver metastases is 15% to 25%. The prognosis may improve if the primary tumour is removed. In patients with New York Heart Association (NYHA) class III/IV heart failure, the prognosis is unfavourable (median survival rate without cardio-surgical treatment is 11 months). We report a case of a 60-year-old patient with carcinoid disease involving grade 2 neuroendocrine tumours and disseminated disease, with liver metastases. His comorbidities were type 2 diabetes, hypertension, and hypercholesterolaemia. He was admitted to the Cardiology Department due to a month-long history of worsening exertional dyspnoea and fatigue (NYHA class III). He had had prior partial resection of the small intestine (2011), had finished a third course of peptide receptor radionuclide therapy with radioisotope-labelled somatostatin analogues (PRRT) two months before hospitalisation, and had been chronically treated with long-acting somatostatin analogues. On physical examination, blood pressure was 130/80 mmHg and heart rate was 90/min. Clinical pathological findings included muted breathing murmur over the right lung, hepatomegaly, and oedema in both legs. Blood tests showed features of cholestasis, mild increase in troponin T levels, and elevated B-type natriuretic peptide levels. Electrocardiography did not show any specific signs of right ventricular or atrial enlargement (Fig. 1). The patient underwent transthoracic echocardiography, which revealed thickening and retraction of the tricuspid valve leaflets, with thickening of subvalvular chords. The planimetry of the tricuspid orifice showed mild stenosis (a valve area of 1.8 cm<sup>2</sup>) (Figs. 2, 3). The mean pressure gradient was 6 mmHg. There was also moderate tricuspid regurgitation, caused by morphological changes (Carpentier's classification: type III). The maximum velocity of tricuspid regurgitation jet was 3.0 m/s and the vena contracta of colour Doppler jet was 6 mm (Figs. 4, 5). The pulmonary leaflets were thickened and shortened, with moderate valve regurgitation (jet occupied < 50% of the right ventricular outflow tract). Colour Doppler showed also mild mitral regurgitation. The symptoms improved (NYHA class I/II) after standard heart failure treatment; further monitoring was recommended with a follow-up echocardiography scheduled at three months or in case of symptom worsening. Echocardiography should be performed in all patients with carcinoid syndrome. Routine repeated cardiac screening may help determine the optimal timing of referral to surgical intervention.

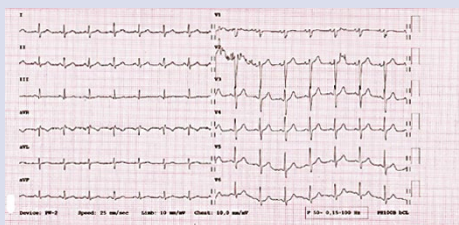


Figure 1. Electrocardiography; no specific signs of atrial or right ventricular enlargement

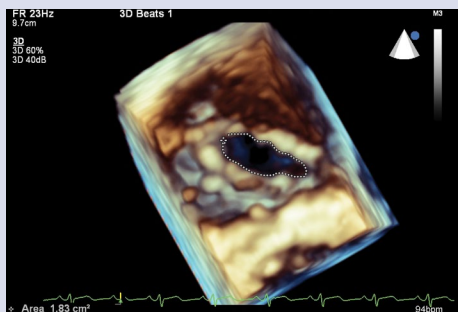


Figure 2. Thickening of tricuspid valve leaflets, with mild stenosis and valve area of 1.8 cm<sup>2</sup> on three-dimensional echocardiography recording

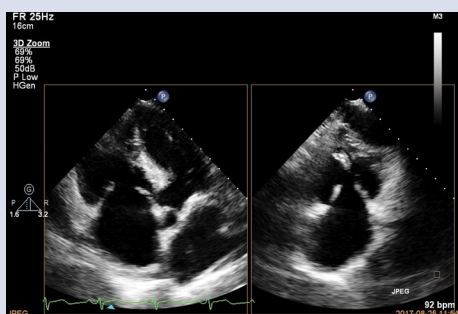


Figure 3. Tricuspid valve with leaflet immobility on three-dimensional echocardiography recordings

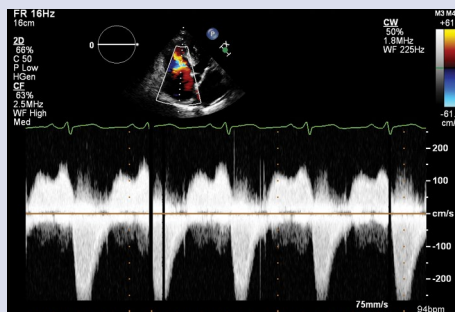


Figure 4. Pulsed-wave Doppler; peak velocities of tricuspid flow > 1.0 m/s due to tricuspid stenosis and regurgitation

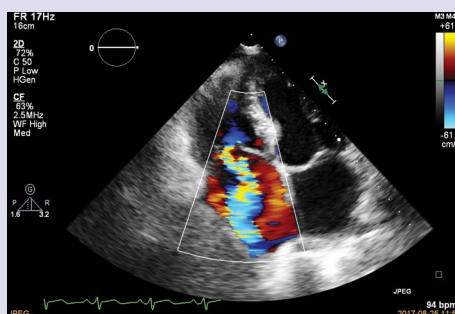


Figure 5. Moderate tricuspid regurgitation on colour Doppler echocardiography

#### Address for correspondence:

Katarzyna Starzyk, MD, PhD, 1<sup>st</sup> Clinical Department of Cardiology, Swietokrzyskie Centre of Cardiology, ul. Grunwaldzka 45, 25-736 Kielce, Poland, e-mail: zikas@poczta.onet.pl

**Conflict of interest:** none declared

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