

Staged treatment of carcinoid syndrome complicated with severe tricuspid regurgitation

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We present a case of a 49-year-old male with symptomatic carcinoid heart disease (CHD), a rare complication of gastrointestinal carcinoid syndrome, which occurs in 1–2 per 100 000 adults per year. The 5-year survival rate depends on staging, its origin, resectability, and the presence of CHD. CHD is characterized by structural damage to the right heart, which is an effect of biogenic amines secretion by the tumor. The presence of CHD is associated with poor survival of 11 months for patients presenting in New York Heart Association (NYHA) functional class IV, and the key prognostic factor is the function of the tricuspid valve [1].

Our patient was diagnosed with a primary tumor in a small intestine with metastases to the liver, bone, and peritoneum and was treated in cooperation with a reference endocrinology center with a somatostatin analog. The serum level of urinary 5-hydroxyindoleacetic acid, the main metabolite of serotonin, was 329.3 mg/24 hours (N: 2–9 mg/24 hours). He presented with facial flushing, diarrhea, and persistent facial plethora on the cheeks, forehead, and chin (Supplementary material, *Figure S1*). He had severe functional impairment: NYHA class III, 6-minute walk test (6MWT) of 180 m, and maximal oxygen consumption of 10.4 ml/kg/min as assessed by ergospirometry. Serum concentration of N-terminal pro-B-type natriuretic peptide was elevated (1326 pg/ml, N: <125 pg/ml).

Echocardiography showed dilation of the right ventricle and right atrium, preserved right ventricular function, moderate stenosis

and regurgitation of the pulmonary valve, severe tricuspid regurgitation [2] with thickened, retracted immobile leaflets with a coaptation gap of 13 mm (*Figure 1A–C*). Right heart catheterization (RHC) showed a reduced cardiac index (1.75 l/min/m²), normal mean pulmonary artery pressure (16 mm Hg), pulmonary vascular resistance (2.2 Wood units), and pulmonary artery wedge pressure (8 mm Hg).

Due to symptomatic tricuspid regurgitation and significant exercise limitation, the perioperative risk of abdominal surgery was considered high [3, 4]. Accordingly, the Heart Team proposed that tricuspid valve replacement should precede abdominal surgery.

The patient was operated on with the use of extracorporeal circulation through the right atrial access. In the operation field, we could see the remains of the tricuspid leaflets adhered to the tricuspid annulus. A Perimount 29 M bioprosthesis was implanted successfully with a trace tricuspid regurgitation (*Figure 1D*). The following hospital stay was uneventful.

Four months after cardiac surgery the patient underwent successful partial resection of the ileum occupied by the primary tumor. Two months afterward, he was reassessed and presented a significant improvement in exercise capacity (NYHA, class II; 6MWD, 375 m). Echocardiography showed a normal gradient of bioprosthesis and a noticeable improvement in heart geometry (*Figure 1E*). RHC showed an improved cardiac index of 2.31 l/min/m². We observed a relapse of the tricuspid regurgitation, which was moderate and stable at the 8-month follow-up (*Figure 1F*).

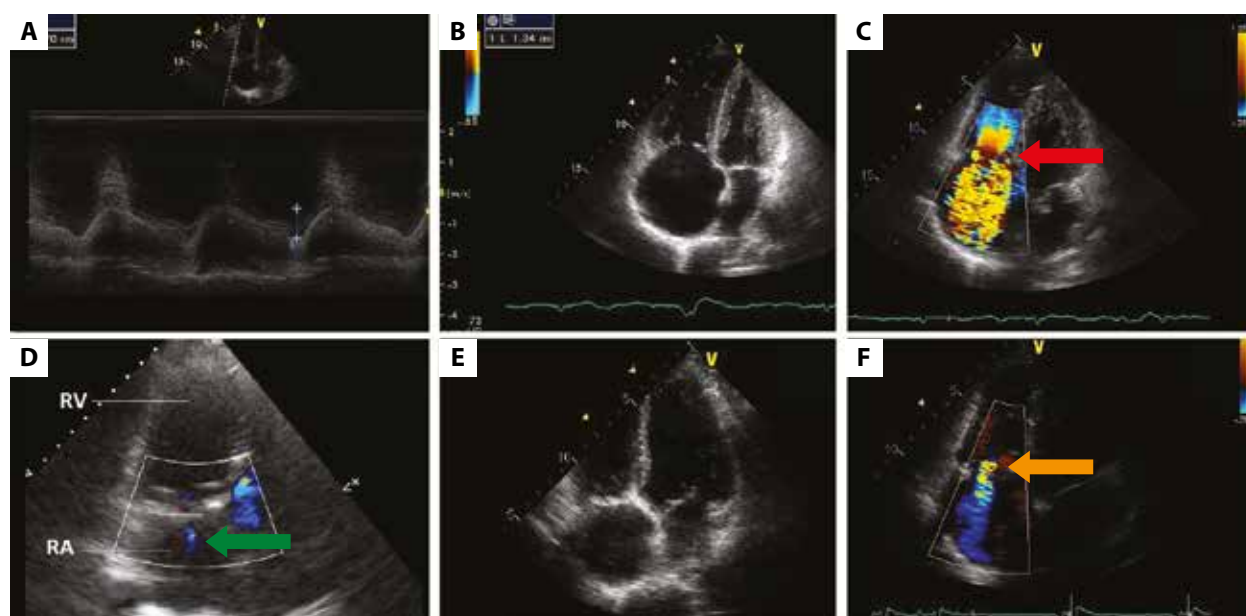


Figure 1. **A.** Normal right ventricle contractility presented as tricuspid annulus plane systolic excursion (TAPSE) of 27 mm. **B.** Coaptation gap of tricuspid valve leaflets. Enlargement of right atrium (area of 37 cm²) and right ventricle (basal dimension in apical 4-chamber view of 50 mm and right ventricular outflow tract diameter of 47 mm). **C.** Colour flow image of severe tricuspid regurgitation of native valve (the red arrow). **D.** Trace tricuspid regurgitation within first day after successful tricuspid valve replacement (the green arrow). **E.** Improvement in chamber and heart geometry after surgical tricuspid valve replacement (right atrium area of 19 cm², right ventricular outflow tract diameter of 40 mm and basal dimension in apical 4-chamber view of 44 mm). **F.** Moderate bioprosthesis regurgitation at 8-month follow-up (the orange arrow)

Abbreviations: RA, right atrium; RV, right ventricle

In our patient, cardiac surgery facilitated a clinical improvement and let the patient be safely operated on for an abdominal tumor. However, at follow-up, we observed deterioration of the tricuspid valve, which could result from the delay between cardiac and abdominal surgery and exposure of the prosthetic valve to high levels of serotonin metabolites before tumor resection [5]. Our results show the need for immediate tumor resection after valvular replacement and strict observation of the tricuspid prosthesis structure and function at follow-up.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

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