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Percutaneous transcatheter pulmonary and tricuspid valve replacements in a patient with carcinoid heart disease

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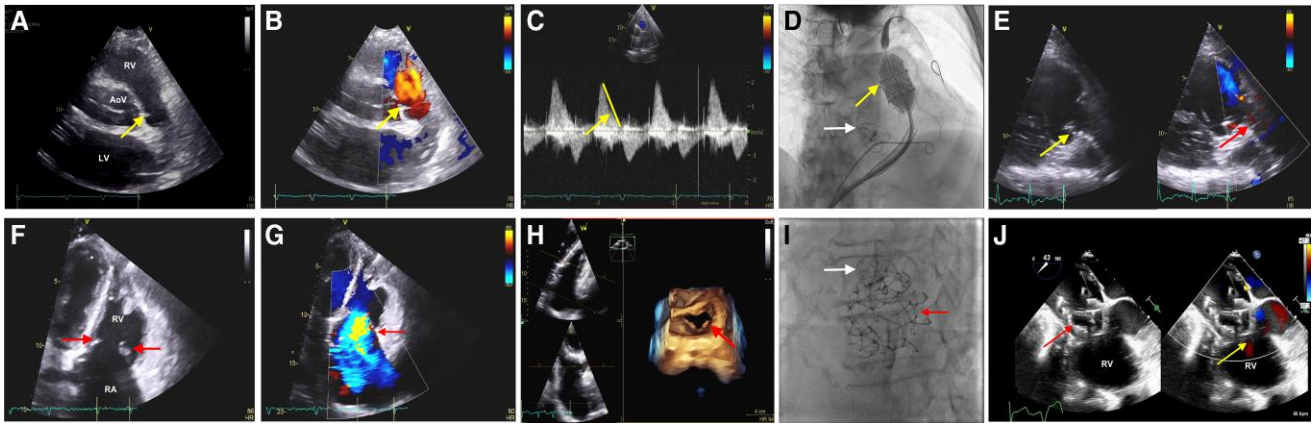

(*Panel A*) Transthoracic echocardiography 2D parasternal short-axis view prior to intervention showing thickened, retracted, carcinoid pulmonary valve cusp (yellow arrow). (*Panel B*) Pre-intervention transthoracic echocardiography with colour flow Doppler on the parasternal short-axis view demonstrating severe pulmonary regurgitation (yellow arrow). (*Panel C*) Pre-intervention transthoracic echocardiography continuous wave Doppler across the pulmonary valve demonstrating very short pressure half-time (yellow line and arrow) consistent with severe pulmonary regurgitation. (*Panel D*) Catheterization image showing pulmonary prosthesis deployment (yellow arrow). The Amplatzer from prior patent foramen ovale closure can also be seen (white arrow). (*Panel E*) Post-transcatheter pulmonary valve replacement 2D and colour Doppler transthoracic echocardiography images showing prosthetic pulmonary valve (yellow arrow) in diastole with only trivial periprosthetic regurgitation (red arrow) and no prosthetic regurgitation. (*Panel F*) 2D transthoracic echocardiography prior to transcatheter tricuspid valve replacement four-chamber right ventricle focused view showing thickened and retracted tricuspid valve leaflets that do not close in systole (red arrows). (*Panel G*) Transthoracic echocardiography prior to transcatheter tricuspid valve replacement with colour Doppler demonstrating wide-open torrential tricuspid regurgitation (red arrow). (*Panel H*) Transthoracic echocardiography 3D imaging demonstrating carcinoid tricuspid changes and restricted, immobile valve leaflets that do not coapt in systole. (*Panel I*) Catheterization image showing deployed percutaneous tricuspid prosthesis (red arrow), and patent foramen ovale closure device sits above (white arrow). (*Panel J*) Post-transcatheter tricuspid valve replacement transoesophageal echocardiogram images showing suboptimally visualized but well-seated prosthetic tricuspid valve (red arrow) with no significant regurgitation on colour Doppler (yellow arrow). RV, right ventricle; AoV, aortic valve; LV, left ventricle; RA, right atrium.

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Carcinoid syndrome (CS) is a paraneoplastic syndrome associated with neuroendocrine tumours (NETs) that arise predominantly in the gastrointestinal tract. Carcinoid heart disease (CHD) arises when NET metastases to the liver leads to reduced clearance of vasoactive substances produced by the tumour, with serotonin (5-HT) playing a prominent role in endocardial proliferation mainly in the right side of the heart before it is deactivated in the lung circulation.[1] Consequently, plaque-like fibrous deposits develop on the tricuspid and pulmonic valves that lead to primarily valvular regurgitation but also stenotic physiology in some.[2–4] This in turn may result in right ventricular volume overload and progression to symptomatic heart failure.[1]

A 79-year-old male with CS from small bowel NET was referred to our clinic for progressive exertional dyspnoea and fatigue. Diagnosis of well-differentiated NET was made 15 years prior with biopsy of hepatic metastases that were identified on imaging to investigate abdominal pain. Other significant past medical history was persistent atrial fibrillation, pectus excavatum, and percutaneous patent foramen ovale closure. Transthoracic echocardiography (TTE) demonstrated moderate to severely enlarged right ventricle (RV) with reduced RV systolic function (RV free wall longitudinal peak systolic strain of −15%, fractional shortening of 14%, and fractional area change of 44%). Pulmonary valve cusps were thickened and immobile due to carcinoid plaque with severe regurgitation (*Panels A–C*), and the carcinoid affected tricuspid valve leaflets, which were thickened, retracted, and fixed with severe regurgitation.

Given his symptoms, comorbidities, and prohibitive surgical risk, percutaneous transcatheter pulmonary valve replacement (TPVR) was performed in a staged approach to treat the valves; the pulmonic regurgitation was treated first with a rationale of potentially improving RV volumes and function prior to percutaneous tricuspid procedure and taking into consideration that there is commercially available pulmonic bioprosthesis whereas the tricuspid bioprosthesis still under clinical investigation. Transcatheter pulmonary valve replacement was undertaken with left femoral access, and a pre-stent was deployed into the RV outflow tract as a scaffold given pulmonary valve annular size was too large to accommodate a dedicated pulmonary valve bioprosthesis; a 29 mm S3 Sapien bioprosthesis was then implanted into the pre-stent (*Panel D*).

However, heart failure symptoms persisted after TPVR with repeat TTE demonstrating trivial pulmonary peri-prosthetic valve regurgitation (mean gradient of 2 mmHg) (*Panel E*), and carcinoid thickened and retracted tricuspid valve with wide-open regurgitation (*Panels F–H*) (see Supplementary material online, *Video*). Accordingly, 6 months after TPVR, a percutaneous transcatheter tricuspid valve replacement was undertaken with a 52 mm bioprosthesis that is currently under investigation (*Panels I* and *J*). The patient tolerated the procedure well, was discharged home without complications, and on 3-week follow-up reported significant improvement in symptoms.

Serotonin and other neuroendocrine substances produced by NETs lead to fibrotic, thickened, retracted, and immobile pulmonary and tricuspid valves, associated with limited life expectancy. Advanced age and severe right heart failure, as in this patient, are well-known risk factors for surgical valve replacement. In this high-risk population, percutaneous treatments (especially when double valve replacement is needed) could be beneficial in the setting of right heart failure and improve quality of life.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: Written consent was provided by the patient for publication of this case report. All authors approve the publication of this report, with all patient identifiers kept confidential and material presented solely for educational purposes arising from the clinical encounter.

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Data availability

No new data were generated or analysed in support of this research.

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