**Multivalvular Carcinoid Heart Disease: Mending the Floodgates**

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**Clinical Presentation**

A 57-year-old female developed NYHA II dyspnea, menorrhagia, abdominal pain, diarrhea, and weight loss. She was diagnosed with small bowel low-grade functional neuroendocrine tumour (NET), carcinoid syndrome and carcinoid heart disease (CHD) affecting four valves. Over five months, she developed NYHA IV symptoms requiring increased home oxygen from 2 to 9L/minute.

**Imaging Findings**

Pre-operative transthoracic echocardiogram (TTE) demonstrated marked multivalvular thickening and restriction. The tricuspid leaflets were in a semi-open position causing severe regurgitation with a dense triangular Doppler profile. The anterior mitral leaflet domed in diastole and the posterior leaflet retracted with resultant severe regurgitation. The pulmonic and aortic valves were severely and moderately-to-severely regurgitant respectively. There was

mild biventricular dilatation and mild left (EF 47%) and moderate right ventricular (RV) dysfunction with septal flattening. A large patent foramen ovale (PFO) was present. Findings were consistent with advanced multivalvular CHD.

**Summary/Discussion Points**

She was evaluated at Heart Team Rounds with medical and surgical oncology input. Despite cardiac deterioration, tumor biomarkers (urinary 5-HIAA) declined rapidly (2510 to 490 umol/L) with somatostatin analogue therapy. Considering this and her poor cardiac prognosis if unrepaired, she underwent quadruple bioprosthetic tissue valve replacement and PFO repair. Her course was complicated by RV failure, an open chest and 3-week intubation. By two weeks, TTE showed recovery of biventricular size to normal, with improved function (left normal, right mildly-to-moderately reduced). She returned to her local hospital on day 45. Nine months later, she was ambulatory without heart failure symptoms.

Up to half of patients with NET and carcinoid syndrome develop CHD from tumor secretagogues including serotonin.[1] Fibrotic plaques deposit on endocardial surfaces. On TTE, leaflets appear severely thickened and retracted resulting in predominantly regurgitant tricuspid and regurgitant/stenotic pulmonic valves.[2] Left-sided valvular involvement occurs in <10%, including in the presence of a PFO.[3] Valve surgery is considered the only effective treatment for symptoms and survival.[4] Surgical correction for four valves is rare with the largest series reporting on seven patients.[3] To our knowledge, this is the first reported case in Canada.

Given the paucity of evidence and high-risk surgery, multidisciplinary shared decision making is essential.

References:

1. Grozinsky-Glasberg S et al. European Neuroendocrine Tumor Society (ENETs) 2022 Guidance Paper for Carcinoid Heart Disease. *J Neuroendocrinology*. 2022; e13416.

2. Narula N et al. The Thickened Valve. *J Am Coll Cardiol*. 2020;1845-8.

3. Arghami A et al. Quadruple Valve Replacement in Patients with Carcinoid Heart Disease. *J Thorac Cardiovasc Surg*. 2010; 1432-1434.

4. Connolly H et al. Early and Late Outcomes of Surgical Treatment in Carcinoid Heart Disease *J Am Coll Cardiol*. 2015;66 (20):2189-2196.

**Figure**

Legend: Multivalvular CHD with semi-open tricuspid valve (A) and severe regurgitation (B), restricted mitral valve mimicking rheumatic appearance (C), severe pulmonary regurgitation with short pressure half time (E) and tricuspid/mitral prostheses with early right ventricular reverse remodelling (F).