

# Carcinoid heart disease: a rare cause of isolated right heart failure

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## History

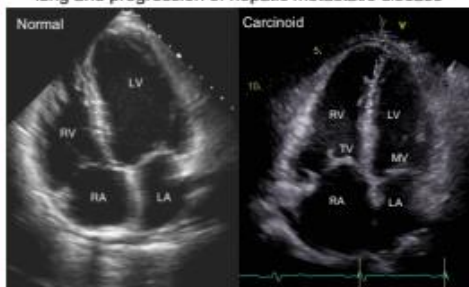
- 68-year-old man with a history of:
  - carcinoid tumor with metastasis to the bones and liver, and carcinoid syndrome
  - carcinoid removal with ileo-colonic resection and hepatectomy 7 years ago
- Presented with 1 year of progressive exertional dyspnea and lower extremity edema

## Physical exam

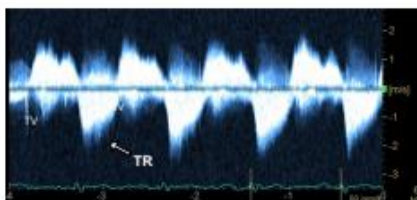
- Prominent v-waves to the level of the mandible
- JVP of 8-10 cm
- Grade 3/6 systolic and 2/6 diastolic murmurs
- Lungs clear to auscultation bilaterally

## Investigations

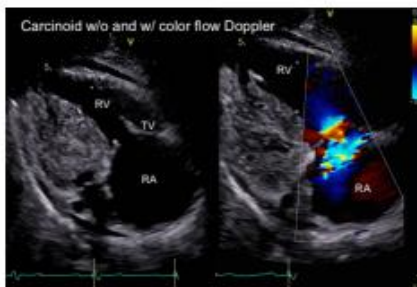
- ECG: incomplete right bundle branch block
- Transthoracic echocardiogram (TTE):
  - thick and immobile tricuspid valve (TV) leaflets, severe tricuspid regurgitation (TR)
  - severe enlarged RV, trace pulmonary regurgitation
  - RV systolic function was low-normal
  - LV: normal size and function
  - normal aortic and mitral valves
- Imaging: new 1.4 cm nodular density in the left lower lung and progression of hepatic metastatic disease



Transthoracic echocardiogram with apical 4 chamber view showing thickened, fused and non-coapting tricuspid valve leaflets.



Continuous flow Doppler image of severe tricuspid regurgitation showing the characteristic dagger shape with an early peak pressure and rapid decline.



Transthoracic echocardiogram of the carcinoid patient with RV inflow view showing color flow Doppler imaging of severe tricuspid regurgitation.

### Inactivation



### Neuroendocrine tumors

• 2.5 – 5 cases / 100,000

### 1. Carcinoid tumor

### 3. Carcinoid heart disease

- 20-50% of patients with carcinoid syndrome
- Right sided heart disease more common as vasoactive substances are inactivated by the lung
- Involves myofibroblast infiltration and activation of valvular interstitial cells leading to fibrosis, and worse outcome

### 2. Carcinoid syndrome

- 30-40% of patients with carcinoid tumors
  - Flushing
  - Secretory Diarrhea
  - Bronchospasm

## Discussion

### Pathogenesis

- Hypothesis: hepatic metastases release vasoactive agents, such as serotonin
- This leads to the deposition of fibrotic plaques on the valvular endocardium
- The pulmonary circulation inactivates the vasoactive hormones, thus limiting disease progression to the left heart

### Screening and follow-up

- Screening: 6-monthly NT-proBNP in all patients with carcinoid syndrome.
- NT-proBNP >260 ng/ml → TTE
- TTE remains the gold standard for diagnosis and follow-up.

### Treatment

- Therapeutic options:
  - somatostatin analogues (octreotide)
  - pharmacotherapy for heart failure
  - cardiac valve replacement
- Without treatment, prognosis is poor:
  - 3-year survival of 31%
- In severe cardiac involvement but well-controlled systemic disease, valve replacement can relieve symptoms and improve outcome
- However, in extensive systemic disease, as in this clinical vignette, tricuspid valve replacement has a primary goal of improving symptoms, but without significant survival benefit

## Teaching points

- Carcinoid heart disease is a rare cause of right heart failure
- It is caused by vasoactive agents that result in valvular fibrosis
- Screen with a proBNP, and follow-up with TTE
- Treat with somatostatin analogues, heart failure medications, and/or valve replacement

## References

- Davari J, et al. JACC 2017;69:1288-1304
- Hassan SA, et al. Heart 2017;103:1488-1495.