

A Murmur in an Elderly Patient: What is the Cause?



Hisham Dokainish, MD, FRCPC, FASE, FACC

Associate Professor of Medicine, McMaster University,
Director of Echocardiography, Hamilton Health Sciences,

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Figure 1: LA=left atrium, LV=left ventricle, MV=mitral valve, RA=right atrium, RV=right ventricle, TV=tricuspid valve

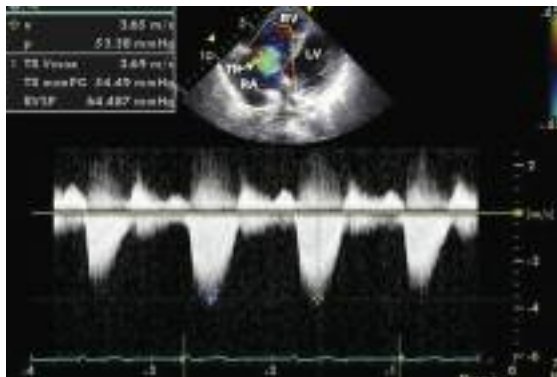


Figure 2: LV=left ventricle, RA=right atrium, RV=right ventricle, RVSP=right ventricular systolic pressure, TR=tricuspid regurgitation

Isolated right-sided valve disease in elderly patients is less common than left-sided valve disease (aortic and mitral valves). Isolated tricuspid valve regurgitation in adult or elderly patients is most commonly due to left-heart disease (left ventricular failure, severe mitral valve regurgitation) or severe respiratory disease (primary or secondary pulmonary hypertension). As opposed to children, significant (moderate or severe) pulmonic valve disease

Meet Vivien

An 88-year-old woman presented with increasing bilateral leg weakness, urine incontinence, and mild back pain; there was no history of stool incontinence. There were no symptoms of dyspnea or chest pain. On examination, heart rate was 72 BPM and regular, and blood pressure was 107/65 mmHg. Jugular venous pressure was 15 cm above the sternal angle, and there was 3+ bilateral peripheral edema to the mid-thighs. Both first and second heart sounds were soft and a III/VI pansystolic murmur was heard along the mid-sternal border. A different II/VI to-and-fro murmur was heard at the left upper sternal border. To investigate the neurologic symptoms, of back pain and urine incontinence, an MRI of the lumbar spine and pelvis was ordered, revealing compression fractures of L4 and L5 roots and a large pelvic mass compressing the bladder with smaller masses in the liver. To investigate the murmurs, an echocardiogram was performed (Figures 1 and 2).

Echocardiography revealed right ventricular (RV) and right atrial (RA) enlargement with thickening and retraction of the tricuspid valve (TV) (Figure 1). Colour Doppler of the TV revealed severe tricuspid regurgitation (TR) (arrow, Figure 2) with an elevated right ventricular systolic pressure of 64 mmHg, indicating moderate pulmonary hypertension. The remainder of the echocardiogram revealed severe pulmonic valve regurgitation; the left ventricle and aortic and mitral valves were within normal limits.

in the elderly is uncommon. However, the combination of significant pulmonic and tricuspid valve disease (stenosis and/or regurgitation) in adults or the elderly should prompt



consideration of carcinoid, a rare neuroendocrine malignancy, with an incidence of 1/75,000.¹ Nearly 90% of all carcinoid tumours are located in the gastrointestinal system, most commonly in the appendix and terminal ileum; less common locations are in the bronchus or gonads. The most malignant of carcinoid tumours arise from the ileum. Importantly, invasive/metastatic disease can cause carcinoid syndrome, characterized by facial flushing, intractable diarrhea, and bronchoconstriction with wheezing. Approximately 50% of patients with carcinoid tumour develop carcinoid syndrome, and approximately 50% of patients with carcinoid syndrome develop heart involvement.

Carcinoid heart disease is characterized by thickening and retraction of the tricuspid and/or pulmonic (right-sided) heart valves with significant (moderate or severe) stenosis and/or regurgitation. The most common clinical presentation in patients with carcinoid heart disease is right-sided heart failure, characterized by severe bilateral peripheral edema, elevated jugular venous pressure, ascites, and liver congestion. In the rare cases of carcinoid heart disease where left-heart valves are also involved (aortic and/or mitral regurgitation/stenosis), extensive liver metastases, bronchial carcinoid, or a patent foramen ovale should be suspected.¹

This patient underwent biopsy of her liver metastases, with pathology confirming carcinoid tumour. What is unusual about this case is that the patient did not have carcinoid syndrome, and presented very late in life, as the mean presentation is approximately 55-years-old.¹ Overall, a high degree of clinical suspicion is needed to diagnose carcinoid disease, and it should be considered in adult or elderly patients with isolated right-sided valve disease with the classic findings of thickened and retracted TV or PV leaflets with significant stenosis and/or regurgitation on echocardiography.

Facts

1. What is a carcinoid tumour?

It is a rare neuroendocrine malignancy; 90% of them are located in the gastrointestinal system, most commonly the appendix and terminal ileum, with less common locations in the bronchus or gonads.

2. What is carcinoid syndrome?

It is a syndrome consisting of facial flushing, intractable diarrhea, and bronchoconstriction, occurring in approximately 50% of patients with carcinoid tumour.

3. What is carcinoid heart disease?

It is a thickening and retraction of the tricuspid and/or pulmonic valve leaflets leading to stenosis and/or regurgitation with the most common clinical presentation being right-sided heart failure.

Take Home Message

Adult or elderly patients presenting with isolated right heart failure who have significant stenosis and/or regurgitation of thickened and retracted tricuspid and/or pulmonic valves on echocardiography should prompt consideration of carcinoid heart disease.

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Reference

1. Fox DJ, Khattar RS: Carcinoid Heart Disease: Presentation, Diagnosis, and Management. *Heart* 2004; 90(10):1224–1226.

Hisham Dokainish, MD, FRCPC, FASE, FACC,
Associate Professor of Medicine, McMaster
University, Director of Echocardiography,
Hamilton Health Sciences, Hamilton, Ontario.