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SHORT COMMUNICATION

Exercise cardiac magnetic resonance imaging with pulmonary artery catheter monitoring in carcinoid heart disease: a shift towards early intervention?

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Abstract

Neuroendocrine tumours are a rare malignancy, which can be complicated by a carcinoid syndrome and, in more rare cases, also valve destruction. The correct timing for surgical repair remains unknown. We report the first-in-men exercise cardiac magnetic resonance imaging with pulmonary artery catheter measurements in order to better understand the haemodynamic impact of isolated tricuspid valve insufficiency in a low symptomatic patient. Not pressure but volume overload is the key factor in the development of symptoms, as long as the right ventricular function is intact. Based on our findings, we referred the patient for tricuspid valve replacement. This case, together with the review of all carcinoid heart disease cases in our hospital (a large tertiary cardiology and oncology centre) since 2000, indicates a potential benefit for early intervention in carcinoid heart disease.

Keywords Carcinoid heart disease; Exercise cardiac MRI; Invasive stress test

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Introduction

Neuroendocrine tumours can cause valve insufficiency due to a carcinoid syndrome. Cardiac manifestation of this rare malignancy is caused by the release of vasoactive substances such as serotonin, which induces plaques of fibrous tissue that distort the valve leaflets and subvalvular apparatus. Usually, vasoactive substances are degraded by the liver or lungs, but liver metastases can cause large quantities of these products to reach the right side of the heart causing primarily tricuspid and pulmonic valve destruction. The optimal timing of tricuspid valve surgery in carcinoid heart disease patients is not clear.[1]

In order to evaluate the haemodynamic impact of early carcinoid heart disease in a case with severe tricuspid insufficiency but without right heart dysfunction, we performed an exercise cardiac magnetic resonance imaging (MRI) with pulmonary artery catheter measurements.

Case report

A 58-year-old woman was referred from the oncology department to the heart failure clinic because of progressive dyspnoea on exertion. She was diagnosed with a neuroendocrine tumour and liver metastases 4 years earlier. After several treatment cycle, she had a sfigle disease at the time of presentation on the cardiology department. The dyspnoea was present since a few weeks. There were no signs of heart failure on clinical examination. Electrocardiogram showed only minor alterations. Echocardiography revealed a structurally normal left heart, but severe tricuspid regurgitation with retraction of the leaflets. Right ventricular (RV) function was normal, and there was no RV dilatation but a clear diastolic D-shaping of the interventricular septum—indicative of volume overload. Cyclo-ergospirometry showed a near-normal exercise capacity (VO_2 max of 27.3 mL/min/kg, 86% of the predicted maximum for age and gender). Coronary

angiography did not show any significant stenoses, and right heart pressures were within normal range. Pre-operative results are summarized in *Figure 1*.

We were left with a patient with objective alterations of the tricuspid valve and subjective complaints of shortness of breath, but without objective parameters that could support referral for tricuspid valve surgery—a potentially dangerous intervention in carcinoid patients.[2]

We proposed the patient to perform an exercise test in the cardiac MRI with measurements of the intracavitary pressures using an MRI-compatible pulmonary artery catheter.

Results are shown in *Figure 2*.

We found that early manifestation of severe isolated tricuspid regurgitation without RV dysfunction leads to a decrease in right ventricle end-systolic volume but an increase in right ventricle end-diastolic volume during exercise, resulting in an absence of stroke volume increase—clearly an abnormal finding—without left-sided or right-sided systolic dysfunction (ejection fraction of both ventricles increased during exercise) or congestion. Left-sided volumes and pressures hardly changed during exercise; this is likely caused by a lack of LV pre-load increase in this case.

Based on the symptoms of our patients and the results of the cardiovascular magnetic resonance, our patient was referred for surgery. Post-operative clinical course was uneventful. Two years later, she is suffering from a progressive disease with lever metastases.

Early detection of carcinoid heart disease is important for a timely referral for valve surgery. After onset of valvular manifestation, these patients may have a limited time frame in which they have a stable oncological process, a still preserved RV function and general acceptable and non-cachectic clinical condition to survive cardiac surgery and still benefit from the intervention.

This seems to be supported by a retrospective analysis of all cases of carcinoid heart disease with surgical repair in our institution since 2000 (n = 15), where we found a very high post-operative mortality both in-hospital and 12 months after surgery. Moreover, pre-operative RV was significantly better in the patients who survived more than 12 months than those who died early, and there was a shorter time from diagnosis to surgery in survivors.[3]

Early referral for surgery may be the way to go for these cancer patients. Multidisciplinary disease programmes

Figure 1 Imaging on diagnosis. (A) Electrocardiogram on diagnosis, showing an rS signal in V1 but no other significant alterations. (B) Transthoracic Echocardiography 4CH view on diagnosis. The right ventricular basal diameter appears to be dilated, but this was not confirmed when measured. On moving images, the retracted septal leaflet was appreciated. (C) Tricuspid insufficiency continuous-wave signal on transthoracic echocardiogram with Doppler measurements. Pressure difference between the right atrium and the right ventricle is 31 mmHg. (D) Cardiovascular magnetic resonance short-axis view, showing the diastolic D-shaping of the interventricular septum—a clear sign of volume overload.

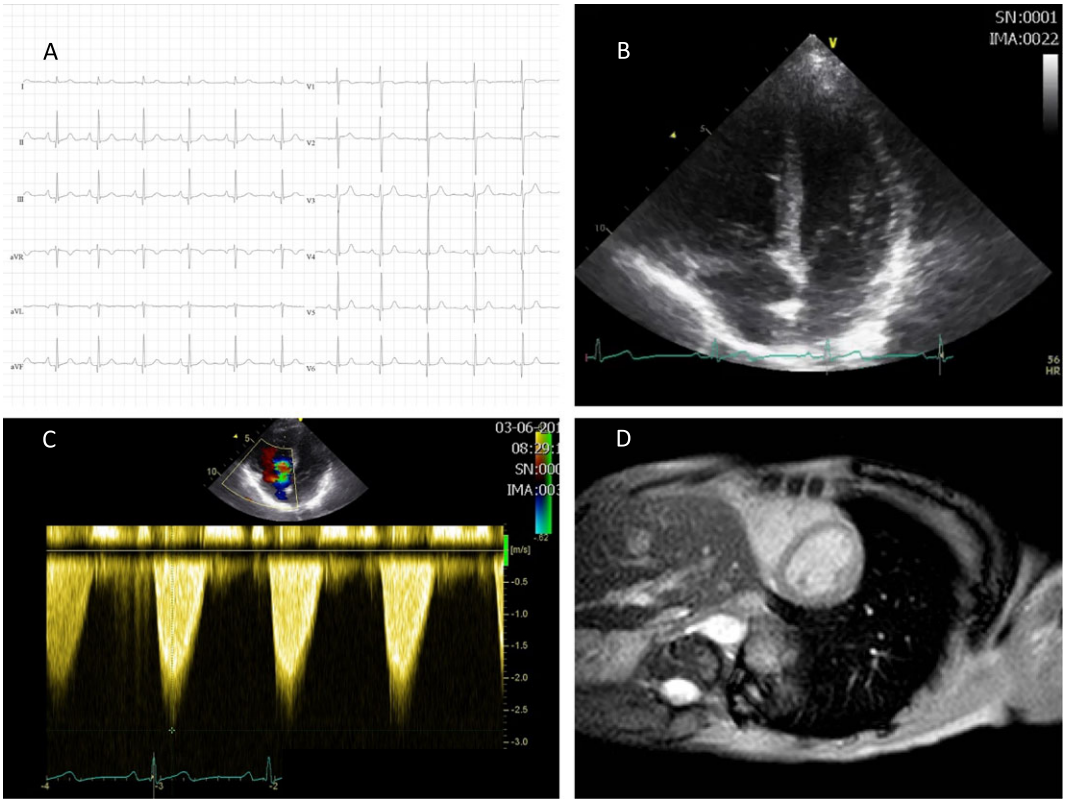

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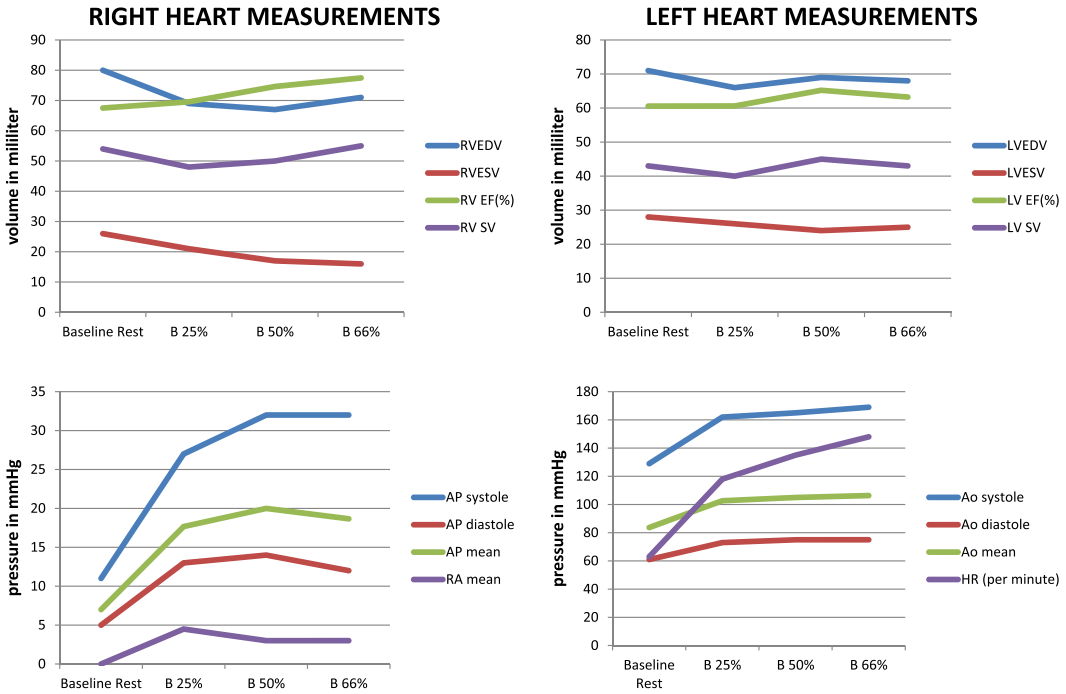
Figure 2 Volume and pressure measurements on invasive exercise cardiovascular magnetic resonance. Left: AP, pulmonary artery; RA, right atrium; RV EF, right ventricle ejection fraction; RV SV, right ventricle stroke volume; RVEDV, right ventricle end-diastolic volume; RVESV, right ventricle end-systolic volume. Right: Ao, aorta; HR, heart rate; LV EF, left ventricle ejection fraction; LV SV, left ventricle stroke volume; LVEDV, left ventricle end-diastolic volume; LVESV, left ventricle end-systolic volume. Baseline: measurements in rest; B 25%, 50%, and 66%: measurements on 25%, 50%, and 66% of maximum exercise capacity, as measured on conventional cardiopulmonary exercise testing beforehand.



including exercise cardiac MRI, possibly without invasive pressure measurement, can identify the patients best suitable for valve surgery. This case is changing our current practice. Together with the digestive oncology department, we are currently setting up a comprehensive clinical pathway with focus on early detection and surgery to improve outcomes for patients with this rare cardio-oncological syndrome.

References

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