Quadruple valve replacement for valve destruction in carcinoid heart disease

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We present a case of a 69-year-old female in whom all four valves were replaced for valve destruction secondary to carcinoid syndrome.

Our patient presented with small bowel obstruction due to a tumour in the terminal ileum that was subsequently resected. Histology confirmed a neuroendocrine tumour. She had hepatic metastases and raised 5-hydroxyindoleacetic acid. Her symptoms were flushing and diarrhoea but no bronchospasm. She did, however, have exertional breathlessness. On echocardiography, all four cardiac valves were markedly thickened with severe regurgitation (Panel A: mitral valve, Panel B: tricuspid valve, Panel C: aortic valve, Panel D: pulmonary valve).

She proceeded to have all four valves replaced successfully. Owing to the left-sided valve destruction, an intracardiac communication was considered and sought at the time of surgery. She was found to have a patent foramen ovale which was closed.

There are only two case reports in the literature for all four valve replacements in carcinoid disease.

Carcinoid is a neuroendocrine tumour usually found in the midgut (90%). With metastases, it is referred to as carcinoid syndrome. This is characterized by the overproduction of serotonin which classically leads to right-sided valve lesions. It is the serotonin in the venous circulation that has a direct effect on the right-sided valves. Typically, the tricuspid valve leaflets become progressively thickened, shortened, and retracted resulting in incomplete coaption and regurgitation. Similar pathogenesis occurs in the other affected valves. Ten per cent of the patients also present with true cardiac metastases.

Left-sided valve involvement is uncommon (5–10%) and patients presenting with this should be investigated for an intracardiac connection or lung involvement.