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Carcinoid heart disease

A 79-year-old patient with no underlying disease presented with a history of severe weight loss, nausea, diffuse upper abdominal pain and exertional dyspnoea. Physical examination

showed malar telangiectasia, signs of severe right heart failure and of tricuspid regurgitation including hepatomegaly with systolic pulsations. The results of laboratory tests were as follows: haematocrit 48%, white-cell count 8800 per cubic millimetre, platelet count 567000 per cubic millimetre, sodium 136 mmol per litre; potassium 4.4 mmol per litre, creatinine 82 μ mol per litre, serum total bilirubin 34 μ mol per litre, aspartate aminotransferase 28 U per litre, alanine aminotransferase 17 U per litre, and serum alkaline phosphatase 118 U per litre. ECG showed a normocard atrial flutter with complete right bundle branch block.

Abdominal ultrasound and computed tomography revealed multiple hepatic masses (fig. 1). Echocardiography showed a severe tricuspid regurgitation secondary to valvular destruction (fig. 2), with systolic flow reversal into the vena cava inferior and the hepatic veins (fig. 3), severe dilatation of the right atrium and ventricle, a severely depressed right ventricular systolic function, and a D-shaped left ventricle, consistent with right ventricular pressure overload. The pulmonary valve appeared functionally normal and both the aortic and mitral were slightly thickened, with moderate aortic and mild mitral regurgitation. Left ventricular size and systolic function was normal and a slight pericardial effusion was noted.

Guided percutaneous needle biopsy of a

Figure 1

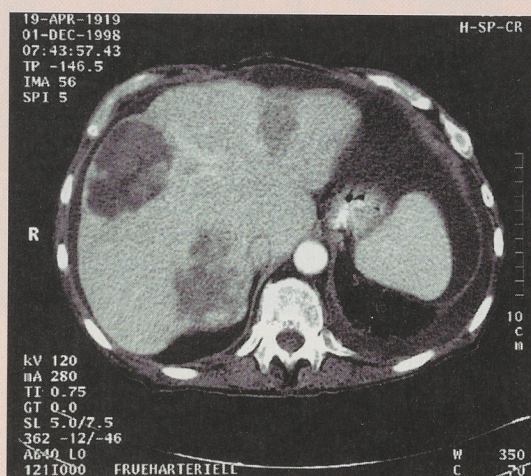
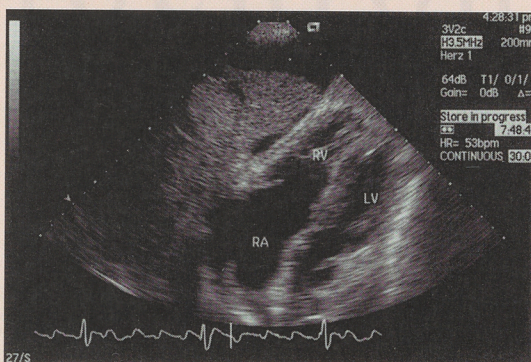


Figure 2



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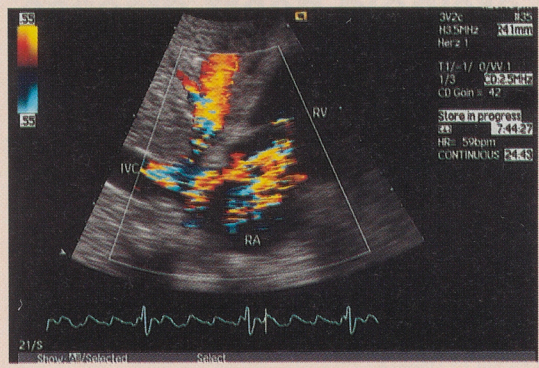
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Figure 3



hepatic mass and markedly elevated 24 h-urinary excretion of 5-hydroxyindoleacetic acid (1245 $\mu\text{mol}/24\text{ h}$) confirmed the suspected diagnosis of carcinoid heart disease. Oesophago-gastroduodenoscopy, colonoscopy and enteroclysis failed to locate the primary tumour, but octreotide scintigraphy revealed increased

uptake in the caecal region. After more than 4 months of follow-up the patient is free of symptoms on digitalis, diuretics and long-acting octreotide.

Carcinoid syndrome leads to focal or diffuse deposits of fibrous tissue on the endocardium of the valvular cusps and of the cardiac chambers, occurring primarily in the right side of the heart. These plaques are usually deposited on the ventricular surfaces of the tricuspid valve, causing the leaflets to adhere to the underlying right ventricular wall, thereby producing tricuspid regurgitation. Carcinoid plaques are also often present in the outflow tract of the right ventricle, where they result in retraction and fusion of the valve cusps, leading to pulmonary stenosis. The left side of the heart may be involved to a minimal degree. Established valvular heart disease cannot be reserved by medical therapy.