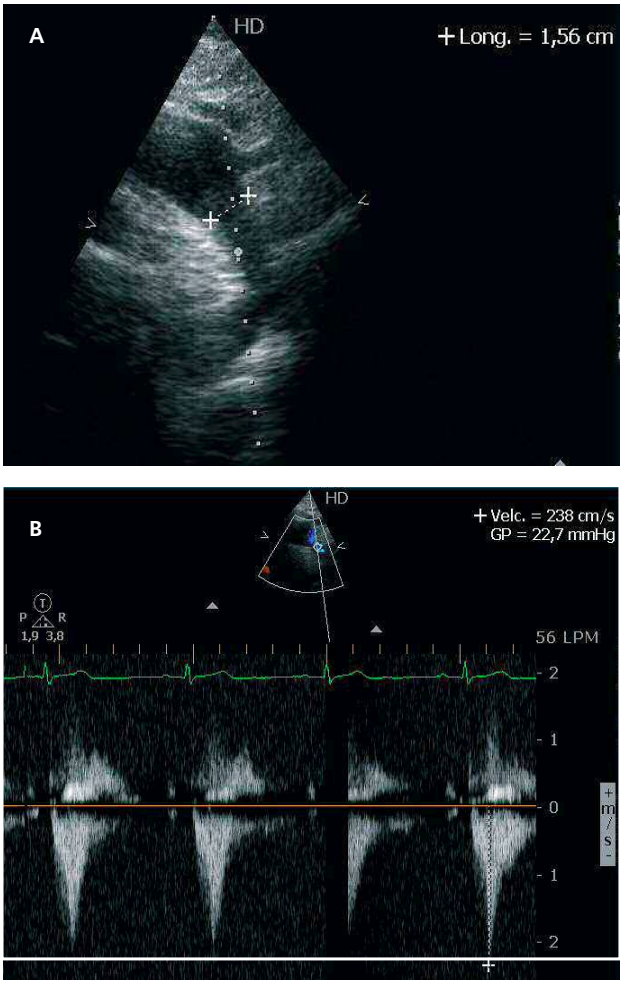

Fig. 1. Transthoracic echocardiography. **A.** Suprasternal view showing the aortic arch and the descending thoracic aorta; an evident change in diameter and narrowing just before the beginning of the descending thoracic aorta (diameter, 15.6 mm) at the level of the supra-aortic trunks are also seen. **B.** Suprasternal view with continuous Doppler at the beginning of the descending thoracic aorta, showing end-systolic acceleration of flow at that level but with no significant gradient.

Reconstructive surgery is the optimal treatment option and must be tailored depending on the characteristics of the disease. Balloon dilation has been used in the treatment of coarctation of the aorta. However, this method has not proved to be effective in patients with aortic hypoplasia affecting long segments. (6)

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Fig. 1. Contrast-enhanced chest CT scan, showing aneurysmal dilation of the ascending thoracic aorta (anteroposterior diameter, 44 mm), and a caliber change in the region of the aortic arch with diameter of 18 mm, and descending aorta of 22 mm.

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Carcinoid Syndrome

We present the case of carcinoid syndrome with cardiac involvement in a 53 year-old female patient with a history of (poorly controlled) hypertension, eclampsia, and chronic renal failure (CRF). She consults a cardiologist for palpitations and dyspnea in variable functional class. During the guided interrogation the patient referred a long-term history of flushing and a two-year history of diarrhea.

Physical examination showed grade 2/6 systolic murmur in the mesocardium, jugular ingurgitation of 4 cm, painful hepatomegaly 4 cm below the costal margin, hard, mobile, painless inguinal adenopathies, facial, chest and lower limb telangiectasias, and mild infrapatellar edema.

Doppler echocardiography revealed mild right and left atrial enlargement, mildly increased right ventricular diameter with preserved systolic function, mild posterior pericardial effusion, tricuspid valve with sclerosis, thickening and hypomobile leaflets with severe regurgitation (Figure 1), and pulmonary valve with sclerosis, high transvalvular velocity, and mild regurgitation (Figure 2).

In view of the characteristics of the heart condition, the presumptive diagnosis was carcinoid syndrome and several complementary tests were performed to identify the carcinoid tumor. The chest, abdomen and pelvis CT scan revealed round hypodense images on the right liver lobe, small amount of perihepatic fluid, isolated enlarged lymph nodes in the retroperitoneal prevertebral space, and internal iliac lymph node chains. An abdominal ultrasound and a cardiac MRI with gandolinum were performed, which were consistent with the CT scan and the echocardiography.

The patient was hospitalized due to impregnation syndrome two months after the first consultation, presenting anemia, pain in the right hypochondrium, and CRF of prerenal origin, exacerbated by diarrhea and use of diuretics. A retroperitoneal lymph node biopsy revealed sarcoid granulomas, and a wedge biopsy of the liver parenchyma marked with CK7, enolase, chromogranin, synaptophysin, and Ki67 showed total parenchyma replacement by atypical cell proliferation defining nests, cords and trabeculae of small, polygonal-shaped cells with clumped chromatin and fibroconnective and vascular pattern with ecstatic vessels without necrosis, a morphology consistent with well-differentiated neuroendocrine carcinoma.

Plasma serotonin and chromogranin and urinary 5-hydroxyindoleacetic acid were measured with positive results, confirming the diagnosis of carcinoid syndrome with unknown primary tumor.

A total-body positron emission tomography (PET) was performed to locate the primary tumor, showing mediastinal enlarged lymph nodes and heterogeneous hepatomegaly with hypodense areas. Since the patient refused to undergo upper digestive tract videoendoscopy and videocolonoscopy (VCC), a capsule endoscopy was performed, showing signs of non-erosive gastropathy without clinical relevance.

The case was presented at a multidisciplinary meeting, in which, given the absence of known primary tumor, the presence of multiple unresectable liver metastases, comorbidities and patient's poor adherence, the decision was taken to apply symptomatic therapy with octreotide. During the course of the disease, the patient had short periods of symptomatic improvement, but the progressive worsening of her

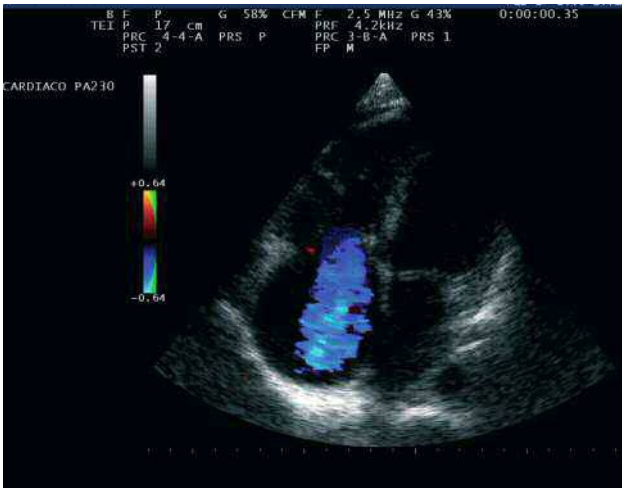

Fig. 1. Transthoracic Doppler echocardiography. Apical four chamber view of mild atrial dilation, mild right ventricular enlargement, mild posterior pericardial effusion, and tricuspid valve with sclerosis, thickening and hypomobile leaflets with severe regurgitation.

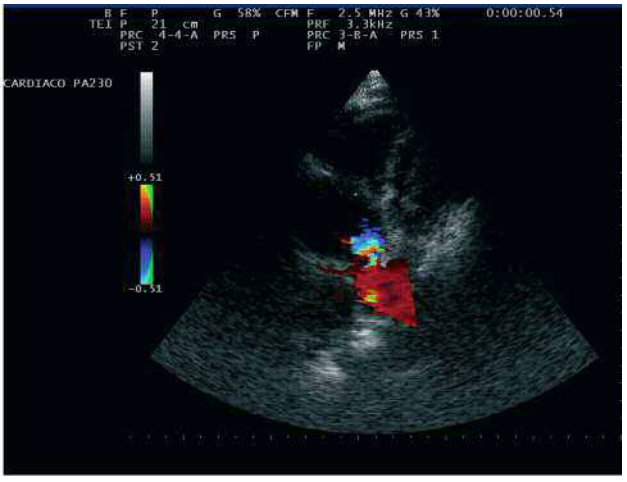

Fig. 2. Transthoracic Doppler echocardiography. Parasternal view of pulmonary artery with sclerosis, high transvalvular velocity, and mild regurgitation.

anemia, CRF, heart failure, and general condition was inevitable, as well as her recurrent hospitalizations. A VCC was performed during one of these hospitalizations, showing a low-grade tubulovillous adenoma in the ascending colon, a finding unrelated to the carcinoid syndrome.

The patient died 30 months after diagnostic confirmation of carcinoid syndrome, in anasarca and with exacerbated CRF.

Carcinoid syndrome is a group of signs and symptoms caused by the metastasis of the carcinoid tumor (rare neuroendocrine malignancy) and by the circulation of the humoral substances it secretes. Tumors of the cecal appendix and the terminal ileum account for 60-90% of the cases; the rest is due to other tumors in

the gastrointestinal system and the bronchi.

The incidence of carcinoid tumors is estimated in 1:75,000 inhabitants, 50% of whom develop carcinoid syndrome, and in turn, 50% of these develop carcinoid heart disease which typically causes abnormalities in the right heart chambers.

Cardiac manifestations are caused by the paraneoplastic effects of humoral substances released by the carcinoid tumor, such as serotonin, histamine, bradykinin, and prostaglandins. These are usually inactivated by the liver, lungs, and brain, but the presence of liver metastases may allow large quantities of these substances to reach the right heart chambers.

Vasoactive substances extend to the left heart chambers only in 5-10% of the patients with multiple liver metastases, bronchial carcinoid, or a patent foramen ovale.

Typically, symptoms are characterized by facial flushing, diarrhea, bronchospasm, and right heart failure.

Many carcinoid tumors follow a prolonged course of up to 20 years from the onset of carcinoid symptoms. However, the development of heart disease in the carcinoid patient heralds a rapid decline in the clinical condition and survival rate is much lower.

Valve regurgitation is the most common complication; about 95% of the patients display moderate to severe tricuspid regurgitation. Pulmonary regurgitation is less common. Both are responsible for right heart failure.

Diagnosis is made with Doppler echocardiography, and is confirmed with serotonin and urinary 5-hydroxyindoleacetic acid assays. Additional complementary tests are only indicative.

In the early phase of the disease, surgical resection of the carcinoid tumor tends to be curative. In some cases, resection or embolization of hepatic metastases can be an option. In patients with the carcinoid syndrome, treatment tends to be palliative; octreotide, analogous of somatostatin, is used for reduction of vasoactive substances and relief of symptoms. Once heart disease has developed, treatment is focused on heart failure.

In selected cases of tricuspid or pulmonary valve stenosis, balloon valvuloplasty produces symptomatic improvement, although recurrent symptoms have been observed after some time. Early valve surgery is the only definitive treatment for severe heart failure. Bioprosthetic valves are preferable, as anticoagulation, which would increase the risk of bleeding secondary to metastasis, can be avoided, and the life expectancy of the patient is likely to be shorter than that of the valve. Patients usually die of severe tricuspid regurgitation rather than of carcinomatosis.

Finally, our diagnostic triad for this disease consists of anamnesis, physical examination and Doppler echocardiography. However, it is even more important for cardiologists to consider carcinoid syndrome as a possible differential diagnosis in tricuspid and pulmonary valve diseases, because it is the only way to provide a better prognosis to our patients.

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Cardioembolic Ischemic Stroke Associated with Mitral Annular Calcification

Mitral annular calcification (MAC) is a chronic degenerative process of the valve fibrous skeleton. MAC has been associated with high risk of systemic embolism, among which cerebral embolism leads to ischemic stroke. We describe a case of stroke secondary to calcific embolization, possibly originated in MAC.

A 78-year-old female patient, receiving eplerenone 25 mg, atorvastatin 10 mg, losartan 50 mg and inhaled bronchodilators on a daily basis, presented with recurrent episodes of expressive aphasia lasting approximately 1 minute. She had several comorbidities, including hypertension (HT), dyslipidemia (DLP), and chronic obstructive pulmonary disease. Twenty days before hospitalization, the patient had been admitted for multiple injuries due to a car accident whereupon a computed tomography (CT) scan of the brain showed no abnormalities (Figure 1). On admission, the neurological examination and lab tests were normal. The ECG showed sinus rhythm, without significant changes. CT scan on admission (Figure 2) revealed bihemispheric subcortical microangiopathic sequelae, mild involutional changes, and punctate calcifications in the cerebellar tentorium and in the M1 segment of the left middle cerebral artery (MCA). During hospitalization, the patient repeated four similar episodes of aphasia with recovery ad integrum. The electroencephalogram revealed left temporal