

Acute heart failure in an adult with unrecognized congenital heart disease

Scompenso cardiaco acuto in un adulto con cardiopatia congenita non diagnosticata

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ABSTRACT: *Acute heart failure in an adult with unrecognized congenital heart disease. M. Correale, A. Lopizzo, F. Santoro, A. Ruggero, A. Cuculo, R. Leva, M. Di Biase.*

We report a case of 46 year-old man, admitted to our Department for a possible massive pulmonary embolism. Instead, diagnosis of Tetralogy of Fallot was es-

tablished by echocardiography and cardiac catheterization.

Keywords: *right heart failure, congenital heart disease, tetralogy of Fallot, diagnosis.*

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Case Report

A 46 year-old man, with no risk factors and permanent atrial fibrillation, had been admitted in the cardiology department of a close hospital, due to a worsening dyspnea, palpitation, and lower limbs edema. The echocardiogram has shown that in diastole the left ventricle takes on a D-shaped configuration with normal systolic function (left ventricle ejection fraction LVEF 60%), furthermore a dilation of right chambers and pulmonary artery trunk with severe pulmonary arterial hypertension and poor inspiratory collapsibility of the inferior vena cava. A massive pulmonary embolism diagnosis had been established by the hypotension and the hypoxemia. The patient has been transferred to our hospital for a possible thromboembolism. Nevertheless, during the hospitalization, a new echocardiogram has showed: besides dilation of right chambers (figure 1) and D-shaped configuration of LV, the right ventricular hypertrophy, overriding of the aorta (figure 2), subaortically, a small structural irregularity is evident, as a small subaortic VSD (figure 3), shunt not evident by Color for a probable bidirectional shunt, furthermore, the presence of a muscular spur in pulmonic subvalvular zone that produced an severe subvalvular pulmonic stenosis (the continuous-wave Doppler showed a peak velocity which is consistent with a systolic gradient of 120 mmHg), the pulmonary artery right and left branch had a standard dimension. The patient has then undergone to cardiac catheterization. So, the catheterization showed increased right ventricle volume with diffused accentuation of the muscular trabeculation, right ventricle outflow obstruction as for infundibular stenosis (subvalvular

gradient of 110 mmHg); instead, left ventricle with standard volume and contractility; in the membranous portion of the ventricular septum a slight opacification of the right ventricle; pulmonary trunk presents a slightly increased diameter. Hemodynamic parameters of cardiac catheterization were: pulmonary artery peak systolic: 23 mmHg; pulmonary artery end-diastolic 14 mmHg; pulmonary artery mean: 18 mmHg; right ventricle peak systolic: 133 mmHg, right ventricle end-diastolic 13 mmHg; average right atrial pressure 14 mmHg; therefore a diagnosis of Fallot Tetralogy has been established, and the patient, has been furthermore transferred to a cardiac surgery center to solve the problem.

Discussion

The reported case has shown how the diagnosis of a congenital heart disease in an adulthood could be difficult to recognize and could be underestimated by cardiologists dedicated to adult cardiology, without being trained or with poor experience in the diagnosis of the less recurring congenital cardiopathies.

The right ventricle has been historically less studied by echocardiography, compared to the left one and cardiac MRI, cardiac catheterization, thoracic pulmonary scintigraphy, high resolution chest CT to exclude extracardiac pathologies, could be used in the differential diagnosis with other pathologies that can lead to the dilated and dysfunction right ventricle (such as enlarging cardiomyopathy interesting the right ventricle, pulmonary embolism, pulmonary arterial hypertension, right ventricle infarction usually associated to left ventricle inferior wall infarction).

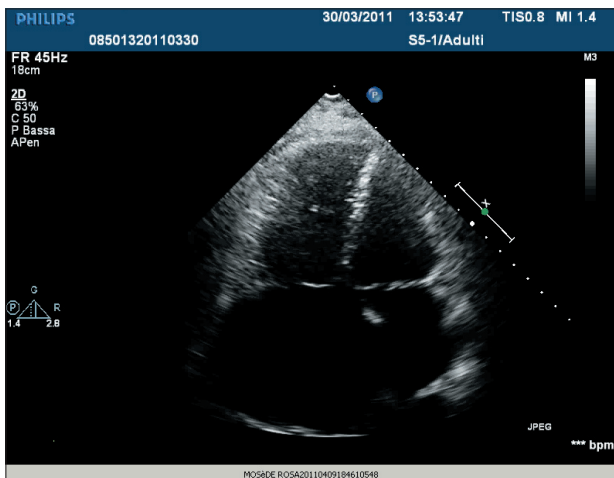


Figure 1. - Two-dimensional echocardiogram: apical four-chamber view of the heart revealing right chambers dilatation.

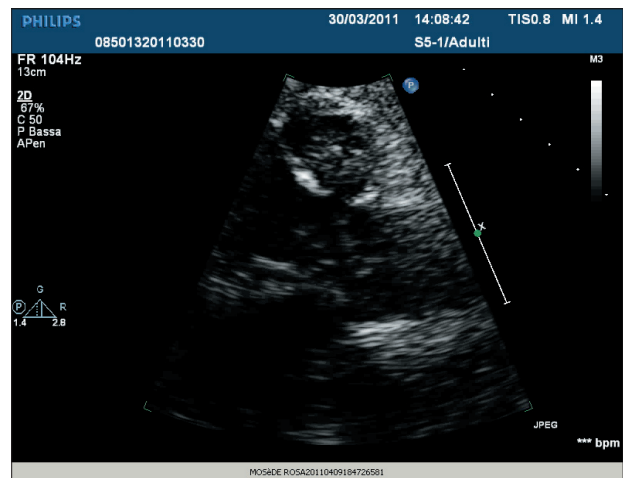


Figure 2. - Two-dimensional echocardiogram: detail of parasternal long-axis view with visualization of overriding of the aorta.

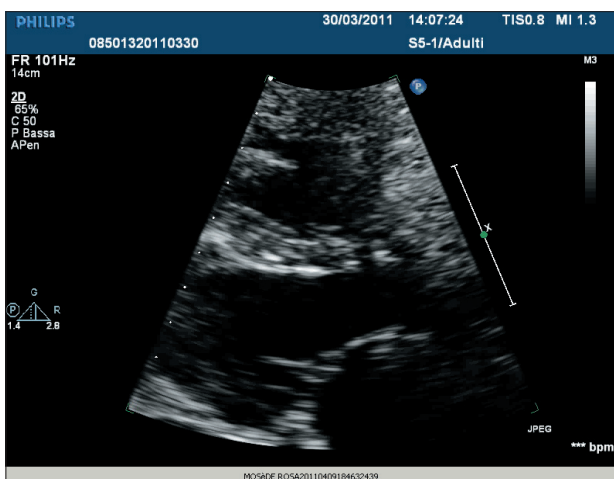


Figure 3. - Two-dimensional echocardiogram: detail of parasternal long-axis view with visualization of a small subaortic VSD.

Among the causes of acute heart failure in adult patients, the congenital heart diseases should be also considered, above all, in those patients with right chambers dilatation associated to ventricular dysfunction. Congestive heart failure is not common in adult congenital heart practice, the adult patients with congenital heart disease develops heart failure in the presence of a substrate (myocardial dysfunction, valvular regurgitation) and a precipitant (such as pregnancy, sustained arrhythmia or hyperthyroidism) [1], as in this case where the not tolerated atrial fibrillation of the patient might have been the outbreking factor.

Particularly, in the adult affected by Fallot tetralogy some typical clinical signs of the pediatric age (such as asphyxial crisis, digital hippocratism) can be absent.

The adult age survival rate in patients affected by Fallot tetralogy is rare, only the 3% of these patients can go over forty without palliatives or definitive adjustment [2].

Factors contributing to longevity included small pulmonary arteries and presumed slow development of subpulmonary obstruction together with moderate concentric left ventricular hypertrophy or systemic-pulmonary artery collaterals for pulmonary blood flow -features previously seen in long term survivors [3].

Moreover, the surgical correction for the Fallot patient who has not been operated, is recommended, since the result are satisfying and the operating risk is comparable to the pediatric one [4], as long as severe concomitant pathologies are absent.

Riassunto

Riportiamo un caso di un uomo di 46 anni, trasferito da altro nosocomio nel sospetto di un' embolia polmonare massiva. Una diagnosi definitiva di Tetralogia di Fallot fu posta dalla nostra equipe dopo nuovo esame ecocardiografico e cateterismo cardiaco.

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