Case 5/2009 - Eight-month old Infant with Mitral Insufficiency due to Hammock Mitral Valve with Frank Heart Failure

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Clinical data

The patient, who had presented dyspnea from birth, showed a clear worsening after the onset of bronchopneumonia at three months of age, with the dyspnea becoming progressively more intense in spite of the appropriate use of specific medication.

At the physical examination, the patient was acyanotic and presented malnutrition, intense dyspnea and decreased pulses. Her weight was 6.3 Kg, heart rate was 166 bpm and the respiratory rate was 80 rpm. The aorta could not be palpated at the suprasternal notch. There were precordial impulses and a clear concave in the left sternal border, with a diffusely palpable *ictus cordi*. The heart sounds were accentuated at all auscultation sites. An intense holosystolic murmur, +++, rough, and a diastolic murmur, +/++, after the third sound, were audible in the mitral area and in the axillary region. The liver was palpable at 4 cm from the right costal border and at 6 cm from the xiphoid appendix.

The electrocardiogram (ECG) (Figure 1) showed signs of biventricular overload and left anterosuperior division block. The ventricular complexes were very broad, with R=35 mm in V1 and 26 mm in V6. There were diffuse alterations of ventricular repolarization. ÂQRS was at -60º and ÂP at +60º.

Radiographic image

The image shows a markedly enlarged cardiac area, due to the dilatation of the four cardiac chambers and a congested pulmonary vascular network. The middle arch is concave (Figure 1).

Diagnostic impression

The radiographic image is compatible with an acyanogenic congenital cardiopathy, with marked volume overload, accompanied by pulmonary venocapillary congestion. Considering the higher enlargement of the left cavities, it suggests a diagnosis of left atrioventricular valve failure.

Differential diagnosis

Acyanogenic cardiopathies with marked left-to-right blood flow shunt of the interventricular communication (IVC)/patent ductus arteriosus (PDA) type must be considered. Additionally, one must consider the acyanogenic cardiopathies with blood flow obstruction on the left side of the heart, such as aortic stenosis and aortic coarctation, when in the presence of left ventricular failure.

Diagnostic confirmation

The clinical elements with marked features of pulmonary venocapillary hypertension (intense dyspnea) and signs of low systemic output (decreased pulses) led to the diagnosis of marked mitral insufficiency, with pulmonary artery hypertension exteriorizing biventricular overload at the ECG. The echocardiogram (Figure 2) confirmed the marked mitral valve insufficiency with increase mainly of the left heart chambers and preserved ventricular function (left atrium: 31; Aorta: 15; left ventricular diastolic diameter : 43; left ventricular systolic diameter: 26 mm; left ventricular ejection fraction: 71%).

Management

The extreme clinical consequences required an emergency cardiac surgery. At the surgery with an ECC of 60', the marked enlargement of the left heart cavities, anomalous mitral valve with cusp borders that were retracted and fused with the papillary muscles, with short and thick *chordae tendineae* were observed, in addition to the mitral ring dilatation. A plastic surgery of the valve apparatus was performed, with an anterior and posterior papillarotomy as well as an anterior and posterior annuloplasty. The patient presented a good initial evolution, but symptom recurrence and the marked mitral valve insufficiency required a new intervention, three months later, with the valve being substituted by a 21-mm St Jude prosthesis, with resolution of the anatomofunctional picture.

However, even through the patient was adequately treated with warfarin and remained clinically stable, she died due to unexplained sudden death three months after the second surgery.

Comment

In congenital cardiopathies, radiographic details can be of great diagnostic help, as in the present case, in which the marked enlargement of the left cavities can guide the diagnosis of mitral valve insufficiency and, specifically in infants, of more striking congenital anomalies of this valve, such as the hammock mitral valve. It is an emergency situation and the valve plasty has little practical effectiveness.

Key words

Infant; heart defects,congenital; mitral valve, insufficiency; heart failure

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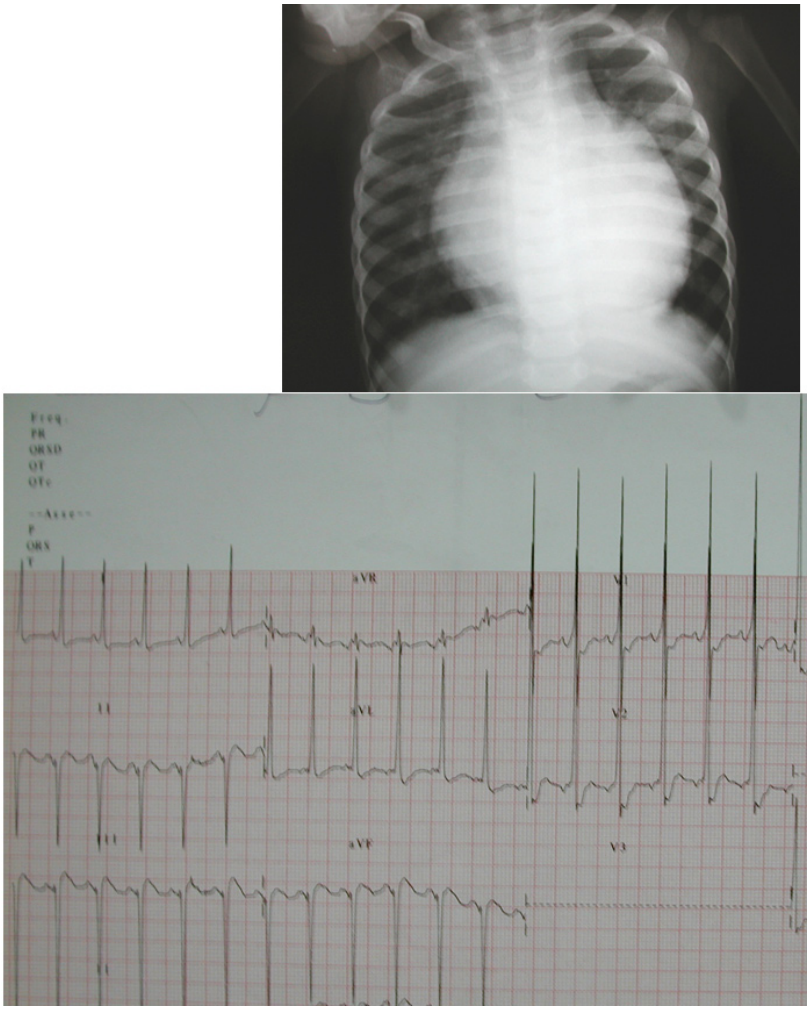

Figure 1 - Radiographic image showing the markedly enlarged cardiac area, mainly the left cavities, with bulging of the middle arch and a congested pulmonary vascular network. The ECG demonstrates the marked increase in the biventricular electrical potentials, given the left ventricular volumetric overload and the right ventricle pressure overload.

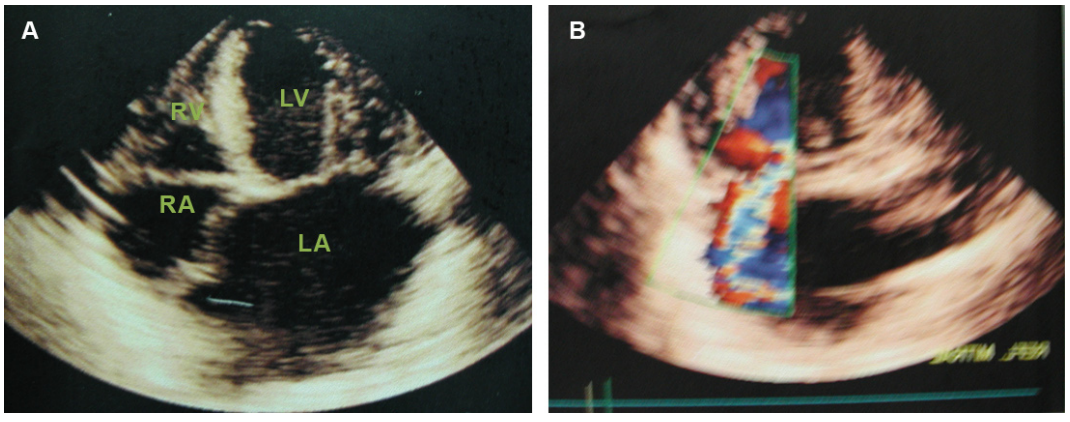

Figure 2 - The echocardiogram shows the marked enlargement of the left cardiac cavities, with the atrial and ventricular septa dislocated to the right in the parasternal 4-chamber view, in A. Additionally, in the subcostal view, the clear mitral valve insufficiency, in Color Doppler, in B; RA – right atrium; LA – left atrium; RV – right ventricle; LV – left ventricle.