

Idiopathic dilated cardiomyopathy in an asymptomatic pregnant patient

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Abbreviations

LVEF: left ventricular ejection fraction

LV: left ventricle

ABSTRACT

Dilated cardiomyopathy in pregnancy is generally of unknown origin, but in 20-35% it is hereditary. Here is presented the case of a 22-year-old patient, 34.6 weeks of pregnancy, primipara, with a history of slight bronchial asthma, who was asymptomatic from the cardiovascular point of view, until a heart rate greater than 130 beats per minute and edema in lower limbs were found, which were the causes that bring her to the Department of Cardiology. The transthoracic echocardiogram revealed dilated cardiomyopathy with severely depressed left ventricular function, diastolic dysfunction, severe tricuspid and moderate mitral regurgitations, as well as slight pulmonary hypertension. It was considered a very high obstetric risk (group IV of the World Health Organization Classification) and, after coordinating with the national department of heart disease and pregnancy, the patient was sent to the reference center, where a cesarean section was performed without complications.

Keywords: Pregnant women, Dilated cardiomyopathy, Heart failure

Miocardiopatía dilatada idiopática en paciente embarazada asintomática

RESUMEN

La miocardiopatía dilatada en el embarazo es, generalmente, de origen desconocido, pero en un 20-35% es hereditaria. Se presenta el caso de una paciente de 22 años de edad, con 34,6 semanas de gestación, primípara, con antecedente de asma bronquial leve, que se encontraba asintomática desde el punto de vista cardiovascular, hasta que se encontró una frecuencia cardíaca mayor de 130 latidos por minuto y edema en miembros inferiores que fue la causa de la consulta de Cardiología. El ecocardiograma transtorácico reveló una miocardiopatía dilatada con función ventricular izquierda gravemente deprimida, disfunción diastólica, insuficiencia tricuspidea grave y mitral moderada, e hipertensión pulmonar leve. Se consideró muy alto riesgo obstétrico (grupo IV de la clasificación de la Organización Mundial de la Salud) y, previa coordinación con el servicio nacional de cardiopatía y embarazo, se envió a la paciente al centro de referencia donde se realizó cesárea sin complicaciones.

Palabras clave: Mujeres embarazadas, miocardiopatía dilatada, Insuficiencia cardíaca

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INTRODUCTION

Dilated cardiomyopathy is characterized by a dilated left ventricle (LV) with systolic dysfunction that is not caused by an ischemic or heart valve disease¹. There is often a latent period of asymptomatic LV systolic dysfunction, before clinical symptoms appear. Patients with this disease are at risk of ventricular arrhythmias and can sometimes be initially treated for aborted sudden cardiac death¹.

Despite a thorough evaluation, in a significant proportion of patients with this disease, there is no obvious cause of cardiomyopathy, thus, they are assigned the diagnosis of idiopathic dilated cardiomyopathy. Extensive family-based studies have shown that, if clinical screening with electrocardiogram and echocardiogram is carried out on the first-degree relatives of these patients, evidence of cardiomyopathy will be found in at least 20-35% of them, which establishes therefore, the diagnosis of familial dilated cardiomyopathy^{1,2}.

A 50% of cases with dilated cardiomyopathies in pregnant women are of unknown origin, and 20-35% of them are hereditary. A 40% of the genetic causes of dilated cardiomyopathy have been identified, with more than 50 described genetic mutations. Predictive factors of maternal mortality are: functional class III-IV of the New York Heart Association (NYHA) and left ventricular ejection fraction (LVEF) < 40%. Very adverse prognosis is related with LVEF < 20%, mitral regurgitation, right ventricular dysfunction, atrial fibrillation and low blood pressure³.

Electrocardiography frequently reveals hypertrophy of the left ventricle, nonspecific changes of the ST-segment and the T wave, or a branch block, but it may not be specific for cardiovascular disease⁴.

Echocardiography reveals ventricular dilatation, which can be in a wide range (from mild to severe), as well as LV systolic dysfunction. Left ventricular wall thickness is usually in a normal range, but the LV mass is almost invariably increased. Most commonly, there is global hypokinesia of the left ventricle, but regional abnormalities of wall movement, particularly dyskinesia of the septum, can also be seen in those with left bundle branch block. There is often mitral and tricuspid regurgitation, which can be serious, due to annulus dilation, even though the physical examination does not reveal an intense murmur. Apart from impaired valve coaptation, the mitral and tricuspid valves appear structurally normal⁵.

Pre-pregnancy treatment includes modifying heart failure medications to avoid fetal harm. The evaluation and treatment of pregnant patients with dilated cardiomyopathy depend on the clinical situation. In all cases, joint obstetric and cardiologic actions, serial echocardiography, determination of natriuretic peptide as well as fetal ultrasound are required. These patients should be treated by a multidisciplinary team, with experiences in cardiovascular diseases in the pregnant woman, and in a center designated for this⁶.

CASE REPORT

A 22-year-old woman, 34.6 weeks of pregnancy, primipara, with a history of slight bronchial asthma, without medical treatment and without crises in several years, is presented. The patient reported that her pregnancy had passed without difficulty, with tolerance to decubitus position and without cardiovascular symptoms; but for 10 days prior to the consultation, she had been feeling slight dyspnea at night, especially when she had to sleep, where she remained in a semi-fowler position.

She was evaluated in her health area, where a sustained regular tachycardia was observed, as well as edema in both lower limbs to the knees; despite this, the patient reported feeling good. Upon arrival at the hospital, she was evaluated by the on-call cardiology specialist. At physical examination, the signs described were observed, and cardiovascular auscultation revealed rhythmic heart sounds, of good intensity, with midsystolic II/VI murmur in tricuspid and mitral areas, no third or fourth sound or pericardial rubbing. Her heart rate was 145 beats per minute and her blood pressure was 100/60 mmHg. The examination of the respiratory tract was normal.

An electrocardiogram was performed, where sinus tachycardia of 125 beats per minute was observed, without other data of interest (**Figure 1**), thus, an echocardiogram was performed (**Figure 2**), which showed the presence of a dilated LV with severe systolic dysfunction (LVEF < 25%), dilation of the rest of the cardiac chambers, pseudonormalized diastolic filling pattern and mitral (moderate) and tricuspid (severe) regurgitations. The details of the measured values are shown in the **table**. The obstetrical ultrasound, performed to determine the fetal well-being, was normal.



Figure 1. 12-lead electrocardiogram performed on the patient.

Treatment with low molecular weight heparin and digoxin 0.125 mg/day was imposed and, after coordination with the national department of heart disease and pregnancy, she was referred to the reference center in Havana, where she had a cesarean section, without complications for the mother or the fetus. The patient, 45 days after delivery, maintained the same echocardiographic characteristics and presented clinical improvement.

COMMENT

In dilated cardiomyopathy, there are several possible situations that cause LV dilatation and dysfunction, such as certain viral infections, previous toxicity and ischemia; but there is a significant percentage where the cause is unknown. Other possibilities are hereditary diseases and, currently, those caused by different genetic mutations have a crucial importance, which respond to a significant group of dilated cardiomyopathies described as idiopathic^{6,7}.

Patients with severely depressed left or right ventricular function do not tolerate pregnancy well due to the cardiovascular changes that occur in pregnancy, therefore, the risk of maternal and fetal complications exceeds 50%; moreover, the presence of marked dyspnea, especially with small efforts, sustained palpitation, symptoms of low cardiac output or pulmonary edema, presyncope, syncope, among others. In any patient having a left ventricular dysfunction with a LVEF < 40%, pregnancy should be discouraged. Similarly, stress tests may be of interest, since women with LVEF of 40-50% may not tolerate

Table. Values of the measurements obtained on the echocardiogram.

Echocardiographic variable	Value
Aortic root	24 mm
Left atrium diameter	43 mm
Left atrium area	24 cm ²
Basal right ventricle	45 mm
Medial right ventricle	33 mm
Interventricular septum	9 mm
LV posterior wall	8 mm
Right atrium area	25 cm ²
LV end-diastolic diameter	56 mm
LV end-systolic diameter	48 mm
LVEF (area-length)	23,5 %
LVEF (modified Simpson)	20 %
TAPSE	17 mm
Pressure in left atrium	8 mmHg
LV end-diastolic volume	162 ml
LV end-systolic volume	116 ml
E wave	86 cm/seg
A wave	61 cm/seg
E/A ratio	1,41
Jet area of mitral regurgitation	6,97 cm ²
Jet area of tricuspid regurgitation	12 cm ²
PA systolic pressure	35 mmHg
Estimated mean pressure in PA	27 mmHg
Inferior vena cava diameter	19 mm
Inferior vena cava collapse	<50 %

LV, left ventricular; LVEF, left ventricular ejection fraction, PA, pulmonary artery; TAPSE; tricuspid annular plane systolic excursion

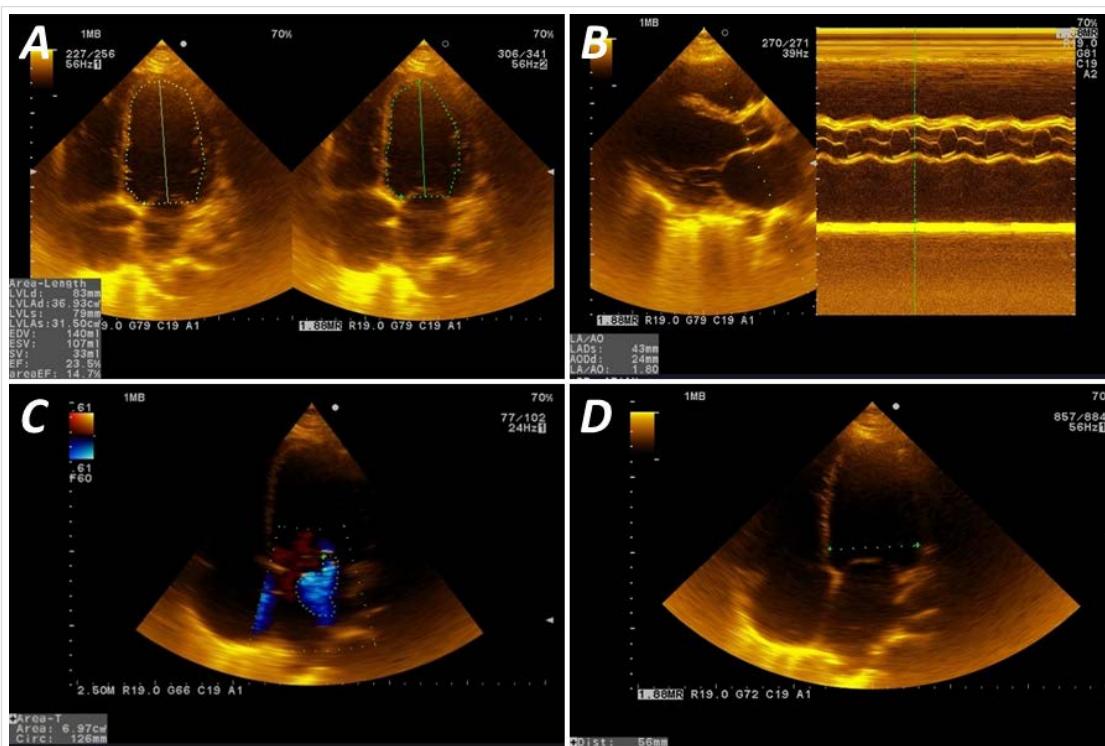


Figure 2. **A.** Calculation of the left ventricle ejection fraction by area-length in a 4-chamber apical view. **B.** Parasternal long axis view, where the aortic root and the left atrium diameter are determined. **C.** Mitral regurgitation by color Doppler. **D.** Left ventricle diameter in 4-chamber view.

ate pregnancy well when their functional aerobic capacity is low⁸.

All patients with dilated cardiomyopathy who are planning a pregnancy should receive multidisciplinary advice and care, since there is a high risk of irreversible deterioration of the ventricular function, maternal death and fetal loss^{9,10}; but in the case of the presented patient, this antecedent was not known and the diagnosis was made at an advanced stage of gestation.

In pregnant patients, the diagnosis of a dilated cardiomyopathy with severely depressed ventricular function represents a contraindication to continuing gestation, and it is classified in group IV (high risk) of the World Health Organization for pregnant patients with cardiovascular diseases, by which she should continue her care in an institution (in-hospital setting) and with a multidisciplinary team with experience in this type of patient. Regarding delivery, the most recommended is the planned cesarean^{9,10}.

In these patients, some medications that are commonly used in heart failure are contraindicated: angiotensin-converting enzyme inhibitors, angioten-

sin receptor blockers, angiotensin receptor and neprilysin inhibitors, mineral receptor antagonists or corticosteroids, and ivabradine. The administration of any drug in the pregnant woman must be individualized, although the first thing is to end the pregnancy in order to favor the most appropriate treatment for the patient; and in the case that the fetus is viable, a lung maturation must be performed and proceed to the interruption of pregnancy¹⁰.

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