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at 70 bpm, QTc of 440 ms and was followed-up in the outpatient clinic with favorable outcome. In conclusion, TDP amiodarone-induced LQTS is an infrequent but severe condition requiring timely electrocardiographic diagnosis and specific treatment.

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Non-compaction peripartum cardiomyopathy: just a coincidence?

The prevalence of non-compaction cardiomyopathy (NCC) is low; yet, its diagnosis is progressively increasing as a consequence of advances and use of the different diagnostic imaging techniques. It is characterized by a thin, compacted external layer and an extensive non-compacted internal layer, with prominent trabeculation. Although its pathogenesis is still controversial, the American Heart Association includes this condition as a primary genetic cardiomyopathy. The interruption of the myocardial compaction process which normally occurs during intrauterine development may play a role in its pathogenesis. The clinical presentation of NCC is variable; subjects may be completely free of symptoms and the diagnosis is incidentally made or may present heart failure, arrhythmias, thromboembolism or sudden death. (1)

Peripartum caripomyopathy (PPCM) is defined as the development of heart failure in the last month of pregnancy or within 5 months of delivery in the absence of another etiology and in the absence of demonstrable previous heart disease. Its prevalence is very low and varies in different regions from 1 per 100 live births in Zambia and Nigeria to 1 per 3000 live births in the United States. Although the etiology of PPCM is unknown, many potential causes have been proposed, including malnutrition, hormonal abnormalities, infections, abnormal immune response and abnormal response to increased hemodynamic burden of pregnancy. It is more frequent in black women, and in those with hypertension, diabetes, multifetal pregnancies and >30 years. (2)

We report the case of a patient who developed signs and symptoms of heart failure in the immediate peripartum period with complementary tests and morphologic criteria of NCC.

A 30-year-old woman complained of progressive dyspnea four months after delivery, with edema of the lower extremities, fever and bloody sputum. She reported a family history of sudden death in a 15-year-old brother and a 3-year old nephew. She had had five pregnancies and five uneventful deliveries, the last one in September 2011. Two months before consultation (on the second month after delivery) she began presenting edema of the lower extremities and progressive dyspnea in FC III. A few days before seeking medical assistance, she complained of fever and bloody sputum. At physical examination, she looked severely ill and presented tachypnea, tachycardia and fever. Blood pressure was 120/80; 2+ pitting edema in the lower extremities and 2/3 jugular engorgement were observed. The apex beat was laterally displaced and was palpable in early diastole. The first heart sound and the second heart sound were normal and a gallop third sound was heard. Crackles were heard at both lung bases; vesicular breath sounds were decreased and percussion produced a dull tone at the right lung base. The electrocardiogram showed sinus tachycardia and signs of left ventricular hypertrophy. The hematocrit was 32%. A right pulmonary effusion was observed in contrast-enhanced computed tomography scan, with areas of consolidation of the right middle lobe and anterior segment of the right lower lobe, and filling defects in the segmental branches of the pulmonary artery. A diagnosis of heart failure and pulmonary embolism was made. Oxygen therapy was administered, anticoagulant drugs were started and negative fluid balance was indicated, with favorable response. Transthoracic echocardiography showed spheroid left ventricular dilatation with areas of non-compacted myocadium in the mid inferior, apical inferior, mid lateral and apical lateral segments (end-systolic noncompacted-to-compacted ratio: 2.8) with severe left ventricular dysfunction (ejection fraction = 29%). The right ventricle was moderately dilated but with normal systolic function. Diastolic filling had a restrictive pattern and moderate mitral regurgitation and tricuspid regurgitation were observed (Figure 1). Gadolinium-enhanced cardiac magnetic resonance imaging confirmed the echocardiographic diagnosis of NCC, with absence of myocardial edema and late enhancement (a patchy pattern at the level of the septum suggestive of non-ischemic fibrosis). Therapy

was started with bisoprolol, enalapril, spironolactone, ivabradine and vitamin K antagonists. In the months following hospital discharge, the patient remained in FC III and was referred to other center (year 2012) to evaluate the possibility of heart transplantation. Her immediate family was evaluated and seven new cases of NCC were detected by echocardiography. By mid 2014, the patient returned to our outpatient clinic with marked improvement of her general status, without edemas and with dyspnea in FC I-II. A new echocardiogram performed at that time showed left ventricular dilatation with moderate left ventricular dysfunction (ejection fraction 44%, two-dimensional global longitudinal strain: -15%), moderate diastolic dysfunction (pseudonormal mitral filling pattern), and mild mitral and tricuspid regurgitation (Figure 2).

Non-compaction cardiomyopathy is characterized by its genetic heterogeneity and lack of correlation between genotype and phenotype. This suggests that, beyond genetic abnormalities, other not completely known factors are involved. Sporadic and familial forms have been described, with different patterns of inheritance: autosomal dominant, autosomal recessive or X-linked. Despite the increasing number of series published since 1990, the real prevalence of NCC and its mid and long-term outcome are little known. (3) On the contrary, PPCM is recognized as a non-genetic cardiomyopathy in most cases; yet, some cases have been reported in association with familial dilated cardiomyopathy. A recent pathophysiological mechanism has been described. A genetic abnormality decreases the antioxidant capacity and increases cathepsin D expression (proteolytic activity) and prolactin cleavage into its antiangiogenic and proapoptotic forms. A small study has demonstrated beneficial effects of bromocriptine, a prolactin inhibitor. (4)

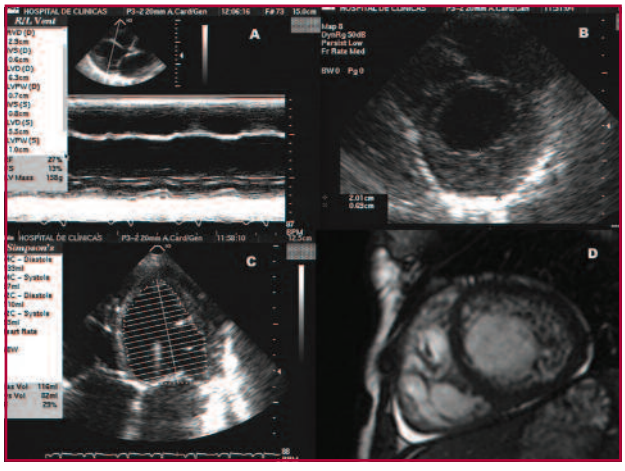

Fig. 1. Echocardiographic image showing left ventricular dilatation (A) and severe left ventricular dysfunction (B). Short-axis echocardiographic views C) and magnetic resonance imaging (D) showing non-compacted myocardium.

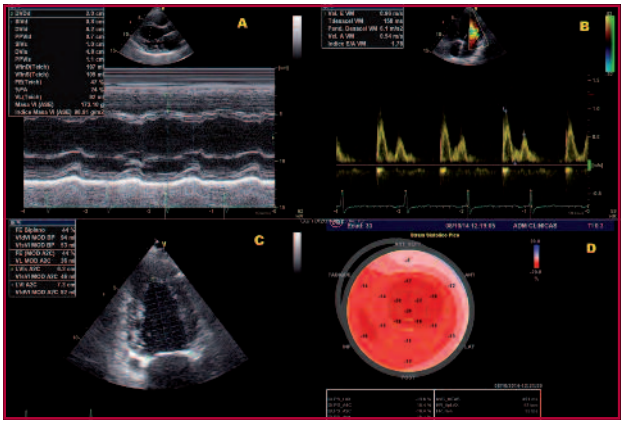

Fig. 2. Echocardiographic image showing left ventricular dilatation (A) with moderate diastolic dysfunction (B) and moderate left ventricular dysfunction (C)

It seems clear that our patient had an asymptomatic form of NCC according to the morphological criteria described and the family history. However, there are some topics of interest to mention. Firstly, the patient had never complained of symptoms during her five previous pregnancies despite the hemodynamic burden associated. Symptoms (dyspnea and edema of the lower extremities) developed during the second month after delivery, when the hemodynamic overload had resolved, and, finally, systolic function and functional class improved in the following months. Although this may happen in patients with NCC, it is more common in those with PPCM. Since 2007, seven case reports of NCC in which the diagnosis was made during the peripartum period have been published. (5) Van Spaendonck-Zwarts et al. have recently described a higher prevalence of PPCM in women with history of familial dilated cardiomyopathy. (6) The interaction of metabolic phenomena characteristic of the pregnancy-delivery-puerperium period (probably due to imbalance between oxidative stress and antioxidant mechanisms) on a genetically predisposed myocardium might explain the occurrence of this condition in our patient.

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Late surgical correction of anomalous origin of the left pulmonary artery and ventricular septal defect

Hemitruncus is defined as anomalous origin of one branch of the pulmonary artery from the ascending aorta with the other branch arising normally from the pulmonary trunk. (1) This acyanotic heart defect with increased pulmonary blood flow with manifestations of severe pulmonary artery hypertension (PAH) and heart failure should be corrected at early stages of life. We describe the case of a 9-month-old infant with late diagnosis of anomalous origin of the left pulmonary artery emerging from the aorta, associated with ventricular septal defect, who underwent successful corrective surgery.

A 9-month-old infant was admitted with heart failure. She had a history of hospitalizations due to recurrent pneumonias. She had signs of protein-energy malnutrition, marked hypernea, subcostal and low intercostal retractions, harsh vesicular breath sounds and rhonchi. Cardiovascular auscultation revealed loud P2, ventricular gallop, and an early and midsystolic murmur grade III/VI heard at Erb's point radiated to the left and right paraesternal borders. An early diastolic murmur of pulmonary regurgitation was heard at the pulmonic area (Graham Steell murmur). The amplitude of peripheral pulses was low. Chest X ray showed a cardiothoracic ratio of 0.68, bilateral hilar congestion, increased pulmonary blood flow specially in the right pulmonary field and peripheral vessel oligemia in the left pulmonary field. The electrocardiogram showed sinus rhythm, right axis deviation (+120°), increased R wave amplitude in V1-V2 and deep S waves in V5-V6 (right Sokolow-Lyon index). The echocardiogram defined the anomalous origin of the left pulmonary artery emerging from the ascending aorta, with severe PAH with suprasystemic pulmonary artery pressures, biventricular dilatation and moderate left ventricular dysfunction with ejection fraction of 43%. A diagnostic cardiac catheterization was performed which confirmed the severity of PAH estimated by transthoracic echocardiography (Figures 1 A and B). Vasoreactivity testing demonstrated a discrete reduction of pulmonary artery pressures and pulmonary resistance index. The patient underwent corrective surgery considering the presence of refractory heart failure and severe but potentially reversible PAH. The procedure consisted of the reinsertion of the left pulmonary artery in the main pulmonary artery and closure of the ventricular septal defect with a fenestrated patch. During the postoperative period, the institutional protocol for PAH was initiated, which includes intravenous high-dose sildenafil and epoprostenol associated with furosemide, spironolactone, captopril and carvedilol for left ventricular dysfunction. The patient evolved with favorable outcome and is followed-up at the outpatient clinic. A computed tomography angiography performed after 6 months of surgery confirmed the effective surgical correction (Figure 2 A and B) and serial echocardiograms show normalization of ventricular dimensions, pulmonary artery pressures and left ventricular function.

The anomalous origin of one of the pulmonary arteries from the aorta was initially described by Fraentzel in 1868, and is commonly associated with other congenital heart defects as tetralogy of Fallot, ventricular septal defect, transposition of the great vessels and subvalvular aortic stenosis. (2)

The largest case series published on this heart disease is attributed to Kutsche and Van Mierop in 1988, who described 108 patients, 89 with anomalous origin of the right pulmonary artery and 19 of the left pulmonary artery emerging from the aorta. (3) Other authors report that the anomalous origin of the right pulmonary artery is 4 to 8 times more frequent than the involvement of the left pulmonary artery. (1)

The embryological development of these varieties of hemitroncus is different: anomalous origin of the right pulmonary artery is thought to be secondary to incomplete or delayed leftward migration of the right sixth aortic arch, while the defect of the left pulmonary artery is considered as failure in the development of the left sixth aortic arch and persistence of the fifth aortic arch. (4)

The pathophysiology of PAH in this congenital heart disease is due to two mechanisms: one of the pulmonary artery branches is subjected to systemic pressure from the ascending aorta and the remaining artery receives the entire output of the right ventricle. (1)

Estimating pulmonary artery pressures and pulmonary vascular resistance index is crucial to define the possibility of surgical correction. In the presence of ventricular septal defect, the traditional calculations by the Fick method may fail and other variables should be analyzed, as manifestations of heart failure, pulmonary venous flow velocity entering the left atrium and the presence of cardiomegaly in the chest X-ray. In these cases, the estimation of pulmonary artery flow and pulmonary vascular resistance index should be calculated for each pulmonary artery with cardiac