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CASE REPORTS

Dilated cardiomyopathy in pregnancy: Beyond obstetric outcomes

Cardiomiopatia dilatada na gravidez: Para lá dos resultados obstétricos

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ABSTRACT

Introduction: Pregnancy is associated with relevant hemodynamic changes that can lead to clinical decompensation in women with previous cardiomyopathy. Given this risk, the literature generally advises against pregnancy in these cases, but this recommendation is controversial.

Case description: Herein is described the case of a woman with dilated cardiomyopathy and left ventricular ejection fraction of 28% who decided to proceed with pregnancy. The baby was born by cesarean section at 32 gestational weeks, with good neonatal outcomes. However, significant cardiac deterioration after birth determined the need for cardiac transplant.

Discussion: The physiological changes that characterize pregnancy can be too demanding for women with previous cardiomyopathy and low left ventricular ejection fraction. In these cases, pregnancy should be monitored by a multidisciplinary team, with cardiac disease control. The timing and mode of delivery is still controversial, given the lack of studies in the area. In addition, the long-term impact of pregnancy and delivery on cardiac function should not be neglected.

Keywords: cardiovascular pregnancy complication; dilated cardiomyopathy; implanted cardioverter-defibrillator; left ventricular dysfunction; pregnancy

RESUMO

Introdução: A gravidez está associada a importantes alterações hemodinâmicas que podem conduzir a descompensação clínica em mulheres com cardiomiopatia de base. Por esse motivo, a literatura desaconselha a gravidez nestes casos, mas esta recomendação não é consensual.

Descrição do caso clínico: É descrito o caso de uma mulher com cardiomiopatia dilatada e fração de ejeção ventricular esquerda de 28%, que optou por prosseguir com a gravidez. O bebé nasceu por cesariana às 32 semanas de gestação, com bons resultados neonatais. Contudo, subsequente deterioração cardíaca materna determinou a necessidade de realização de transplante cardíaco.

Conclusão: As alterações fisiológicas características da gravidez podem não ser bem toleradas em mulheres com cardiomiopatia prévia e baixa fração de ejeção do ventrículo esquerdo. Nestes casos, a gravidez deve ser monitorizada por uma equipa multidisciplinar, com controlo da patologia cardíaca. O momento e tipo de parto são ainda controversos, dada a falta de dados nesta área. Para além disso, o impacto a longo prazo da gravidez e parto na função cardíaca não deve ser negligenciado.

Palavras-chave: cardiomiopatia dilatada; cardioversor-desfibrilhador implantável; complicação cardiovascular de gravidez; disfunção ventricular esquerda; gravidez

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INTRODUCTION

Pregnancy is characterized by relevant hemodynamic changes, including a significant increase in cardiac output, stroke volume, and heart rate. These physiological changes in women with known cardiomyopathy can lead to clinical decompensation. Since the risk of maternal and/or neonatal mortality is very high in women with dilated cardiomyopathy with reduced left ventricular ejection fraction (LVEF), the literature generally advises against pregnancy in these cases. However, as there are no clinical trials addressing this issue, there is a lack of robust data supporting this recommendation.[1,2]

CASE DESCRIPTION

The present case refers to a twenty-nine-year-old woman with dilated cardiomyopathy and LVEF of 28%, sinus bradycardia, and ventricular dysrhythmia, already carrying an implantable cardioverter-defibrillator (ICD). Her usual medication included ramipril, furosemide, and bisoprolol.

As relevant family history, the woman had a sister who died at the age of 30 years due to heart failure and an uncle who died of sudden death at the age of 33 years.

The woman got pregnant against medical recommendation and, after being assessed in maternal-fetal consultation and explained the risks, decided to proceed with the pregnancy.

She suspended ramipril but maintained furosemide and bisoprolol and was kept under close surveillance by the medical team, without major complications.

At 24 weeks of gestation, she was admitted for surveillance and induction of fetal lung maturation with betamethasone. The pregnancy proceeded with maternal clinical stability and no signs of fetal distress. At 32 weeks of gestation, a cesarean section was performed, and a healthy boy with 1650 g was born. The woman was admitted to the Intensive Care Unit and, given the favorable clinical course, discharged home one week after birth.

One month after delivery, she returned to the hospital due to an ICD shock. ICD analysis revealed sustained ventricular tachycardia, with a cycle length within the ventricular fibrillation zone, and the bisoprolol dose was increased. Four days later, the woman again returned to the hospital due to an episode of lipothymia. ICD analysis revealed another episode of ventricular tachycardia, and she was admitted to the hospital with the diagnosis of arrhythmic storm, starting amiodarone perfusion. The transthoracic echocardiography was repeated, showing mild mitral and tricuspid insufficiency, severe dilation of the left ventricle (end-diastolic volume of 93mL/m^2), and LVEF of 28%. The patient was discharged home three days later with oral amiodarone.

Seven months later, the patient's ICD was upgraded to a cardiac resynchronization therapy defibrillator. However, her clinical status continued to deteriorate, reaching a LVEF <20%, and two years after delivery, she was submitted to cardiac transplant.

DISCUSSION

Dilated cardiomyopathy refers to a heterogeneous group of myocardial disorders characterized by decreased left ventricular contractility and left ventricular dilation in absence of other left ventricular dysfunction causes.[3] Around 50% of cases are idiopathic, of which 20–35% are hereditary.[4]

LVEF <40% has been shown to be a predictor of adverse maternal cardiac outcomes in pregnancy and maternal mortality.[2,4] In fact, the American College of Obstetricians and Gynecologists and the European Society of Cardiology recommend against pregnancy in women in modified World Health Organization (WHO) pregnancy risk category IV, which includes those with ejection fraction <30%.[4,5] Most maternal cardiac complications occur in late pregnancy, corresponding to the cardiac output peak. On the other hand, pregnancy seems to have a negative impact on the clinical course of women with dilated cardiomyopathy, with an important increase in the occurrence of cardiac events after birth compared to nulliparous women.[1]

The lack of clinical trial data remains a significant problem in this area.[1,2]

The management of women with dilated cardiomyopathy with reduced LVEF should always include patient counseling about the expected prognosis and potential maternal and neonatal risks, preferentially in preconception setting.

During pregnancy, there is an increase in circulating blood volume, stroke volume, and maternal heart rate in response to increased estrogen and progesterone levels and activation of the renin-angiotensin-aldosterone system. The increase in cardiac output is driven by the increase in stroke volume during the beginning of pregnancy and in heart rate in the last pregnancy trimester.[5]

The development of placental circulation leads to a substantial reduction in systemic vascular resistance and, in the third trimester, compression of the inferior vena cava by the gravid uterus can lead to preload reduction.[6,7]

Furthermore, there is a catecholamine-induced increase in heart rate and stroke volume due to pain and anxiety during labor, and an up to 50% increase in cardiac output may be seen in the second stage of labor. Also, abrupt changes in fluid balance can occur, particularly during uterine contractions.[6] Increased susceptibility to pulmonary edema at the time of delivery and immediate postpartum derives from increased hydrostatic pressure and decreased colloid osmotic pressure.[5] The heart ventricle mass increases as a response to plasma volume increase during pregnancy. However, ejection fraction variations during pregnancy are inconsistent: some women show no changes, while others show a decrease in ejection fraction. The structural changes in the maternal heart return to baseline until one year postpartum.[5]

These physiological hemodynamic challenges during pregnancy, labor, and delivery can pose important risks to women with preexisting cardiomyopathy, due to their limited cardiovascular

reserve. Therefore, the management of these patients during pregnancy and puerperium should be assured by a multidisciplinary team.

Pharmacological therapy is important to maintain the disease stable and often must be adjusted to pregnancy requirements.[2,6] Angiotensin-converting enzyme inhibitors (ACEI) and angiotensin receptor blockers (ARB) are known for their fetotoxicity and should be stopped during pregnancy. Spironolactone is also not recommended during pregnancy due to potential anti-androgenic effects on male fetuses. Beta-blockers are an option. While atenolol should be avoided for its association with intrauterine growth restriction, no adverse effects have been reported with the use of bisoprolol or carvedilol during pregnancy.[2] Furosemide, hydralazine, long-acting nitrates, and digoxin may be safely used.

The use of ICD can prevent sudden death during pregnancy in women with cardiomyopathy. Although little is known about the outcomes of women carrying ICD during pregnancy, there is no evidence that these patients are at higher risk of adverse complications. On the other hand, the physiological changes in pregnancy seem to have no influence on device operation/activity. Nevertheless, it should always be switched off for surgical interventions where diathermy is used, as inappropriate shocks may be delivered.[8,9]

The decision regarding the timing and mode of delivery is controversial and should always be made on an individual basis and according to multidisciplinary decision.[6] Deterioration of left ventricular systolic function tends to occur in late stages of pregnancy, and its prediction is important to plan the best delivery timing.[8] The main goal of intrapartum care is to ensure sufficient oxygen delivery to meet maternal and fetal demands.

The option of interrupting pregnancy around 32 weeks of gestation has the main purpose of preventing one of the most critical stages of pregnancy regarding hemodynamic changes, thus preventing maternal decompensation. However, if left ventricular function significantly deteriorates or heart failure symptoms ensue, the patient may require labor induction irrespective of fetal gestational age.[8]

After delivery, secretion of endogenous oxytocin makes the uterus contract in order to control the bleeding. Uterine massage and intravenous oxytocin administration should be considered as external maneuvers to help uterus contractions, maintaining a slow oxytocin perfusion to reduce its hypotensive effects. Volume redistribution after delivery usually occurs over 12 to 24 hours. Women with severely reduced left ventricular systolic function are at increased risk of developing heart failure symptoms and pulmonary edema during this period.[3]

Pregnancy cardiovascular complications are not limited to the immediate postpartum. In fact, cardiac events frequently occur in the days, weeks, or months after delivery in women with known cardiovascular disease.[5]

The physiological changes that characterize pregnancy can be too demanding for women with previous cardiomyopathy with low LVEF. For this reason, pregnancy is usually not recommended in these women. However, even with full knowledge of the associated risks, some women decide to proceed with pregnancy. In these cases, a multidisciplinary team should be ready for pregnancy surveillance and cardiac disease control. On the other hand, the timing and mode of delivery remain controversial, given the lack of studies in the area. In the present case, pregnancy interruption was decided at the beginning of the third trimester (at 32 gestational weeks) due to the potential risks posed by hemodynamic changes occurring during this period. This case is also a reminder that pregnancy may lead to an increase in cardiac events after birth, stressing the need for close long-term follow-up in the postpartum.

AUTHORSHIP

Joana Portela Dias – Conceptualization; Writing - original draft
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