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Is it Possible to Bring 4 Pregnancies to Term with Obstructive HCM without Maternal or Fetal Complications? (A Case Report and Literature Review)

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ABSTRACT

Obstructive hypertrophic cardiomyopathy (HCM) during pregnancy is associated with significant maternal and perinatal morbidity. Physiological changes during pregnancy can induce complications: sudden death, heart failure and arrhythmia involving the maternal and fetal prognosis. Women with HCM generally tolerate pregnancy well. The risk is however higher in women who are symptomatic before pregnancy or in those with severe left ventricular outflow tract obstruction. The incidence of arrhythmias does not appear to be increased during pregnancy and maternal mortality is low. Prior to conception, women with HCM should have a risk assessment as well as genetic counseling. A vaginal delivery with regional anesthesia is usually appropriate. Women should be managed by a specialist multidisciplinary team.

KEYWORDS: Hypertrophic cardiomyopathy; pregnancy; anterior systolic movement; LV gradient.

INTRODUCTION

HCM is increasingly diagnosed in women of childbearing age due to more widespread use of echocardiography and familial screening programs. The majority of young women with heart disease, including HCM, wish to consider pregnancy and therefore obstetric admission is a common cause for hospitalization in this patient population. However, to date pregnancy outcome data for these patients are scarce. HCM has recently been defined by the European Society of Cardiology (ESC) as increased ventricular wall thickness or mass in the absence of loading conditions (hypertension, valve disease) sufficient to cause the observed abnormality [1]. Around 70% of HCM patients have at least 1 affected family member, but sporadic mutations are also known to occur [2]. Myocardial disarray and fibrosis are the histological hallmarks of the disease. The diagnosis is primarily made by transthoracic echocardiography, although cardiac magnetic resonance imaging has an additional role, particularly if the hypertrophy is isolated to the LV apex, and it can also diagnose the presence and severity of myocardial fibrosis using gadolinium injection. The phenotypic expression of the disease is very heterogeneous [3]. Diastolic left ventricular dysfunction is invariably present and some patients also develop systolic dysfunction later. There is an increased risk of sudden death, particularly in those with a family history of sudden death, symptoms of syncope, ventricular tachycardia, blunted blood pressure response on exercise and severe hypertrophy. Pharmacological therapy is indicated in symptomatic patients.

CASE REPORT

We report the case of a 39-year-old patient with no known family history of HCM or any notion of sudden death in the family. G4P5 (the second pregnancy is twin). All 4 deliveries are completed and were vaginally without maternal or fetal complications. Our patient was pregnant at 38 weeks of amenorrhea, and she was referred to the gynecology department for urgent exploration of palpitations with a predominantly right heart failure picture. The clinical examination showed a conscious patient, dyspneic, blood pressure at 100 / 60mmHg, a regular heartbeat, a heart rate at 100 bat/min, a systolic murmur in left parasternal, edema of the lower limbs, reaching the thighs taking the cup, and discreet crackling grooves at the bases. The electrocardiogram showed a regular sinus rhythm with paroxysmal passage to atrial tachycardia, left anterior hemiblock, left atrial hypertrophy, left ventricular hypertrophy. III de Maron, with the presence of an anterior systolic movement of the mitral valve (ASM) occupying half of the systole **(Figure 2).** The left ventricle (LV) is not dilated with a septum at 31 mm and a posterior wall at 17 mm **(Figure 1),** and an ejection fraction at 45%. The right

ventricle (RV) is also hypertrophic with a preserved systolic function. Biauricular dilation was demonstrated. We note on doppler a significant left intraventricular gradient at 75mmHg with a saber-blade appearance **(Figure 3).** There is also a type II diastolic dysfunction. An intermediate probability of pulmonary hypertension was objective with a tricuspid leak. It is mentioned that during our examination a supraventricular hyperexcitability is observed in the trace of the ECG of the paroxysmal passages in atrial tachycardia. Our patient was put on betablockers, with an indication of anticoagulation which was asked but not received immediately given her hemorrhagic risk, she was transferred to the intensive care unit for monitoring, and she gave birth at term 2 days later via the vaginal route without incidents. With an MRI planned **(Figure 4),** and a follow-up in cardiology to evaluate the thromboembolic and rhythmic risk, specifically that of sudden death, and also for therapeutic management. The patient had a coronavirus, thus forcing her containment and the management of her disease, which delayed the realization of the etiological assessment of HCM.

DISCUSSION

HCM was defined by the presence of increased left ventricular wall thickness that is not solely explained by abnormal loading conditions. The diagnosis HCM was confirmed if one or more left ventricular (LV) myocardial segments showed a thickness of> 15 mm on echocardiography, or cardiac resonance imaging. Obstructive HCM was defined as 'an instantaneous peak Doppler LV outflow tract pressure gradient> 30 mm Hg at rest or during physiological provocation such as Valsalva maneuver, standing and exercise [3].

During pregnancy, plasma volume and cardiac output increase. The increase in cardiac output in the first and second trimesters is achieved by a larger stroke volume, while later in pregnancy there is an increase in heart rate. The additional volume load of pregnancy causes enlargement of the ventricular cavity, which theoretically might reduce the left ventricular outflow tract (LVOT) obstruction; however, the increased cardiac output tends to counteract this effect and the LVOT gradient will increase with advancing gestation [4]. The same volume loading increases distension of the left atrium and thereby risk of atrial fibrillation. In the context of diastolic disease, the volume changes and increased heart rate are not well tolerated, aggravating symptoms of dyspnea and lowering the threshold for developing left heart failure. At the time of delivery, cardiac output increases further secondary to auto-transfusion of blood from the contracting uterus and increased catecholamine levels. There is also an increase in heart rate secondary to blood loss, pain and stress, while the expulsive efforts during delivery tend to diminish venous return. All of these physiological changes lead to an increase in LVOT gradient and shorten the diastolic filling period, therefore increasing the risk of pulmonary oedema. Which is the case of our patient. All of these physiological changes lead to an increase in LVOT gradient and shorten the diastolic filling period, therefore increasing the risk of pulmonary oedema. Which is the case of our patient. All of these physiological changes lead to an increase in LVOT gradient and shorten the diastolic filling period, therefore increasing the risk of pulmonary oedema. Which is the case of our patient [5].

Only a limited number of pregnancies in women with HCM have been described in the literature [6]. It was demonstrated that pregnancy is generally well tolerated [7]; however, few patients have significant complications [8]. To identify patients at increased risk a pre-conceptual review is recommended [9]. The ESC guidelines on the management of cardiovascular disease during pregnancy advise that a risk assessment is performed using the modified World Health Organization (WHO) classification. HCM is considered a WHO class II or III risk lesion, implying there is a moderate risk of morbidity in most women and potentially a significant risk for some [10]. Therefore, pregnancy is contraindicated in a minority of women, if there is significant impairment of systolic function.

The transthoracic echocardiography is the key of diagnosis. The measures of diastolic function, systolic function, localization and severity of hypertrophy, outflow tract gradients at rest and with provocation, cavity dimensions and gradients, SAM and degree of mitral regurgitation and left atrial dimensions. Some studies have identified several predictors for cardiac complications during pregnancy. Amongst these are New York Heart Association class III or IV, left ventricular outflow tract obstruction and pre-pregnancy arrhythmias [11].

Thaman and al. [12] reported on 271 pregnancies in 127 women. They found a relationship between symptoms before and during pregnancy, whereby of the 28.3% reporting cardiac symptoms during pregnancy, 90% had been symptomatic prior to pregnancy.

Autore and al. described pregnancy outcome in 49 women with HCM, 12 women were symptomatic and in 5 of these there was worsening of their clinical status (42%) (dyspnea or pulmonary congestion). Of the 28 previously asymptomatic women only 1 developed symptom (4%) [13]. These studies, report however a high percentage of heart failure (30.3%), particularly during the third trimester, with low LV function, but LVOT obstruction severity are not given [14].

Several studies showed that clinical deterioration occurred more often in women with LVOT obstruction (peak gradient> 30 mmHg) than in women without obstruction (25% versus 11%); however, the difference was not statistically significant [15].

One retrospective study found that LVOT obstruction was not a predictor of complications [16]. Maternal mortality is low in the few prospective studies that are available [17].

Combining these series there were a total of 71 women and 2 died during pregnancy; however, both were considered at very high risk and had been counseled against pregnancy.

Pregnancy does not appear to aggravate arrhythmias; however, arrhythmia pre-pregnancy is probably predictive of arrhythmia during pregnancy.

In conclusion, available data suggest that the risk of pregnancy is very much dependent on pre-pregnancy clinical status. The risk would appear to be higher in symtomatic patients, those with a history of arrhythmias, significant LVOT obstruction, or impairment of left ventricular systolic function. Severe complications are, however, rare. When discussing maternal risk with the future mother, the risk of disease recurrence in the offspring should be included and genetic counseling should be offered [14].

Other side, concerning the Obstetric and fetal outcome, the most frequently observed fetal morbidity was premature birth (26%), followed by being small for gestational age (8%) and developing fetal bradycardia (3%) [15]. In a review of a large number of published cases Schinkel reported a 25% Caesarean section rate for predominantly obstetric indications and a 15% rate of spontaneous abortion 5% therapeutic abortions and a 2% stillbirth rate [17].

As per the ESC guidelines for the management of cardiovascular diseases during pregnancy [11], women with HCM who are in WHO class II should be reviewed by a cardiologist with clinical examination and echo assessment each trimester. For women in WHO class III more frequent follow-up, monthly or bimonthly, is warranted. These women should be managed in a specialist center by a multidisciplinary team [11]. Follow-up during pregnancy should focus on the development of symptoms especially dyspnoa, arrhythmias, aggravation of outflow obstruction, and diastolic and systolic ventricular function.

The ESC guidelines also recommend that a beta-blocker should be considered if there is more than mild LVOT obstruction or a septal thickness of> 15 mm, in order to reduce the risk of pulmonary congestion [11]. Beta-blockers are also helpful for controlling atrial fibrillation and reducing the risk of ventricular arrhythmias. Metoprolol is the preferred beta-blocker since there is ample experience of its use in pregnancy; however, bisoprolol may also be used. Following birth, the infant should be monitored for bradycardia and hypoglycemia.

Verapamil can be used during pregnancy, but with caution due to the risk of fetal atrioventricular block. Amiodarone should not be used unless absolutely necessary, as it can induce fetal thyroid disorders. Sotalol can be used during pregnancy.

In the end of the second trimester a delivery plan should be in place with a joint discussion of the multidisciplinary team. Vaginal delivery is usually preferred and caesarean delivery is reserved for obstetric indications [11]. Epidural and spinal anaesthesia must be administered cautiously in women with severe LVOT obstruction due to the consequent vasodilatation and hypotension and single-shot spinal anaesthesia should be avoided. However, when an epidural or spinal anaesthetic is administered judiciously, the benefits of pain reduction and reduced sympathetic stimulation may outweigh the disadvantages of these regional techniques [16]. Oxytocin should be given only as a slow intravenous infusion. The fluid shifts associated with delivery and immediately afterwards increase the risk of pulmonary oedema.

CONCLUSION

HCM is usually well tolerated in pregnancy but those with prior symptoms or arrhythmias tend to have a worsening of symptoms during pregnancy. The majority of these patients, however, can be managed medically with good effect. A small subset of women with severe LVOT obstruction are at increased risk but with expert management by a specialist multidisciplinary team, a good outcome can be expected for the most. Functional status and signs of heart failure prior to pregnancy are important risk factors for cardiac complications in pregnant women with HCM. Pre-pregnancy counseling, close monitoring, and optimal care are mandatory to prevent complications in women with HCM.

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FIGURES

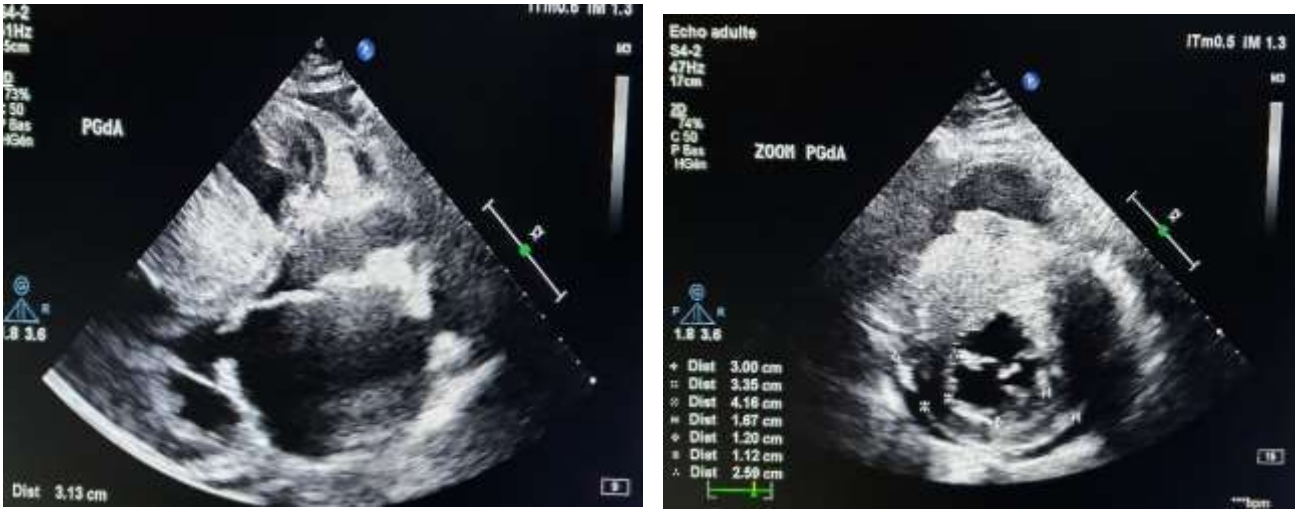

Figure 1: Transthoracic echocardiography (parasternal long- axis view and parasternal short- axis view) showing an important and asymmetric hypertrophy of LV with measures.

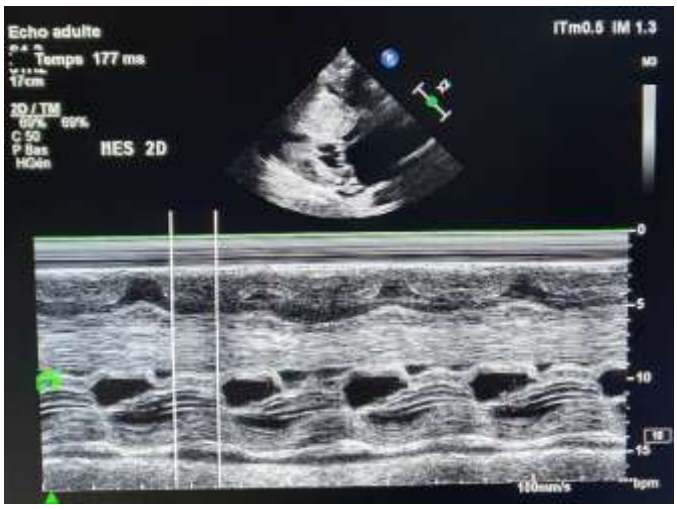

Figure 2: Echocardiography (time-motion) showing the anterior systolic movement of the mitral valve occupying half of the systole

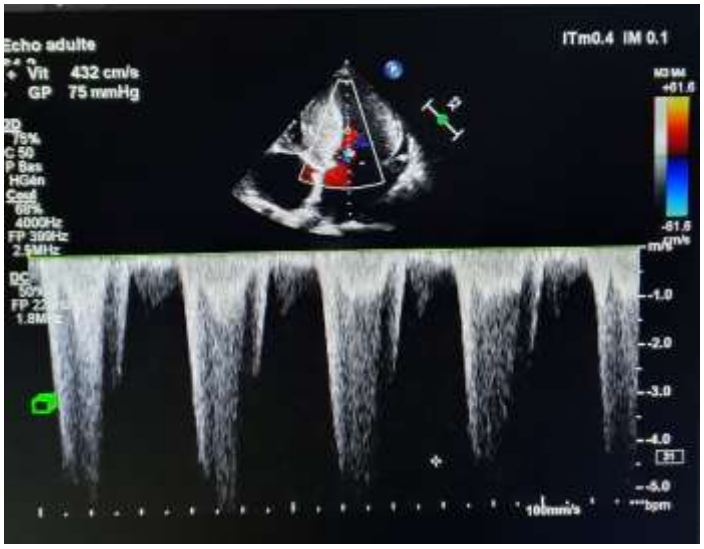

Figure 3: Doppler a significant left intraventricular gradient at 75mmHg with a saber-blade appearance

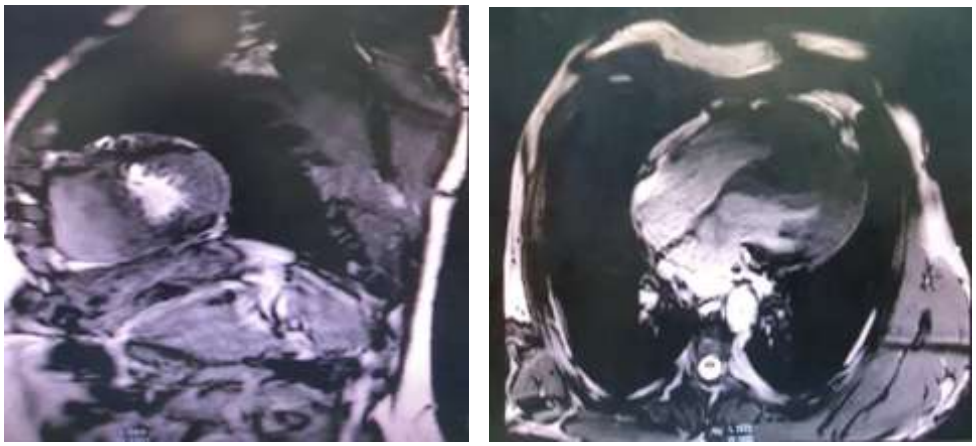

Figure 4: MR showing hypertrophic cardiomyopathy