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Obstructive Hypertrophic Cardiomyopathy: A Potential Threat during Pregnancy

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Hypertrophic cardiomyopathy (HCM) is a disease that can be both clinically and genetically diverse and is a significant factor behind unexpected death and heart failure. Typically, it occurs in the left ventricular ejection pathway, leading to obstruction in the ejection tract.
We present two cases of obstructive hypertrophic cardiomyopathy occurring in pregnant women, with a different evolution, monitoring, and management between the two cases.
Pregnant women with this disease require special attention because of potential complications. Close monitoring is necessary to minimize the risks.

Keywords: Hypertrophic cardiomyopathy; pregnancy; ejection tract obstruction.

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ABBREVIATIONS

HCM : Hypertrophic Cardiomyopathy
LV : Left Ventricle
SAM : Systolic Motion of the Mitral Valve
LVP : Left Ventricular Filling Pressures
GFR : Glomerular Filtration Flow Rate
Hb : Haemoglobin
ECG : Electrocardiogram

1. INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is one of the most common inherited cardiomyopathies, with an estimated prevalence of at least 1 in 500, with equal incidence in males and females in an autosomal pattern. Sexual inheritance [1].

Hypertrophic cardiomyopathy (HCM) is characterized by myocardial hypertrophy, which can be systemic or localized, often limited to the left ventricular ejection tract, which can lead to obstruction of the ejection tract.

Pregnancy can be challenging due to pregnancy-related drug restrictions and difficulty managing the condition during pregnancy [2].

Few studies have systematically assessed the clinical course and maternal and fetal outcomes in pregnant patients with HCM. Due to the inherent complexity of the topic, all currently available studies have sources of bias and/or limitations [3].

This article aims to accentuate how pregnancy affects individuals with hypertrophic cardiomyopathy, emphasizing prognostic factors that contribute to the emergence of complications during gestation. We will accomplish this objective through a comprehensive literature review and two observations.

2. CASES PRESENTATION

2.1 Case 1

A 20-year-old female patient, primigravida with an active pregnancy at 13 months of age, without any cardiovascular risk factor, without any particular pathological history, and any notion of sudden death in the family, was admitted to our structure for obstructive hypertrophic cardiomyopathy, revealed 10 days before her admission by the onset of exertional dyspnea associated with episodes of palpitations without any other associated sign.

Her clinical examination found an asymptomatic patient at rest, with a blood pressure of 105/62mmHg and a heart rate of 88 bpm, without signs of heart failure. The rest of the clinical examination was unremarkable.

Her electrocardiogram showed a regular sinus rhythm with pseudo necrosis Q waves in the anterior extended and negative T waves in the inferior.

Her echocardiogram showed a non-dilated left ventricle with severe concentric left ventricular hypertrophy with a septal predominance (SIV: 16mm, PP: 12mm) and an intra LV gradient at rest at 133mmHg, high LVP (AP-AM > 30mmHg) with the presence of a moderate mitral insufficiency and a complete MAS, with a preserved ejection fraction at 57% (SB), a right ventricle of good function and a PH at 46mmHg.

Biological workup: Hb: 12g/dl, platelets at 250,000 elements/mm³, K+: 4mmol/L, good renal function with a GFR at 117 ml/min/1.73m². An elevated BNP level at 3414 pg/ml (N < 100pg/ml), inflammatory workup was negative with hypersensitive C-reactive protein level at 3.1 ng/L, (N 0-10 ng/L), white blood cells at 7500 (N (4~10) × 10 3 /L)), and normal thyroid workup.

A 48-hour ECG Holter was performed on our patient showing paroxysmal sinus tachycardia at 130bpm without rhythm or conduction disturbances.

The patient was put on carvedilol 40 mg x3/D and low-dose diuretics 20mg per day.

Echocardiograms were performed for the patient's parents and brothers objectifying incidental non-obstructive HCM in the brother, genetic counseling was also done.

The case of our patient was discussed with the gynecologists in a multidisciplinary consultation meeting, and the medical interruption of the pregnancy was indicated given the important risk of maternal complications, in particular arrhythmias and heart failure.

An echocardiographic check-up was performed on our patient after the termination of her pregnancy, showing a clear decrease of the atrioventricular gradient to 70mmHg VS 133mmHg.

Then the patient was referred for a possible myomectomy.

2.2 Case 2

Patient of 33 years old, pregnant with evolutive pregnancy at 8SA, 3rd gesture 2nd pare with the notion of 2 neonatal deaths, the 2nd one had a global CMH at the echocardiography made at birth, the notion of consanguineous marriage and sudden death in her cousin, followed for CMH since 2021 under beta-blockers, without notable cardiovascular risk factor. She was admitted to our facility for obstructive hypertrophic cardiomyopathy, whose history dates back to one month before her admission with the aggravation of her exertional dyspnea associated with episodes of syncope.

Her clinical examination found an asymptomatic patient at rest, with a blood pressure of 120/73 mmHg and a heart rate of 70 bpm, with no signs of heart failure; cardiac auscultation did not find a systolic murmur. The rest of the clinical examination was unremarkable.

Her electrocardiogram showed a regular sinus rhythm with a significant electrical left ventricular hypertrophy (LVH) and secondary repolarization disorders such as negative T waves in the anterior region.

Her echocardiography showed a non-dilated left ventricle with severe concentric left ventricular hypertrophy with a septal predominance (SIV: 20mm, PP: 10mm) and a media-ventricular gradient at rest of 35mmHg, non-elevated left ventricular filling pressure (LVP) with a preserved ejection fraction of 62% (SB), no valvulopathy, no systolic anterior movement (SAM) and a good right ventricle.

Biological workup: Hb 13g/dl, platelet count 170,000 elements/mm³, K+: 4mmol/L, good renal function with a GFR of 102 ml/min/1.73m². BNP level was 360 pg/ml (N < 100pg/ml), inflammatory workup was negative with hypersensitive C-reactive protein level at 6 ng/L, (N 0-10 ng/L), white blood cells at 7630 (N (4~10) × 10 3 /L)), and normal thyroid workup.

A Holter ECG was performed on our patient and did not reveal any rhythm or conduction disorders.

The patient was also put on carvedilol 40 mg x3/D.

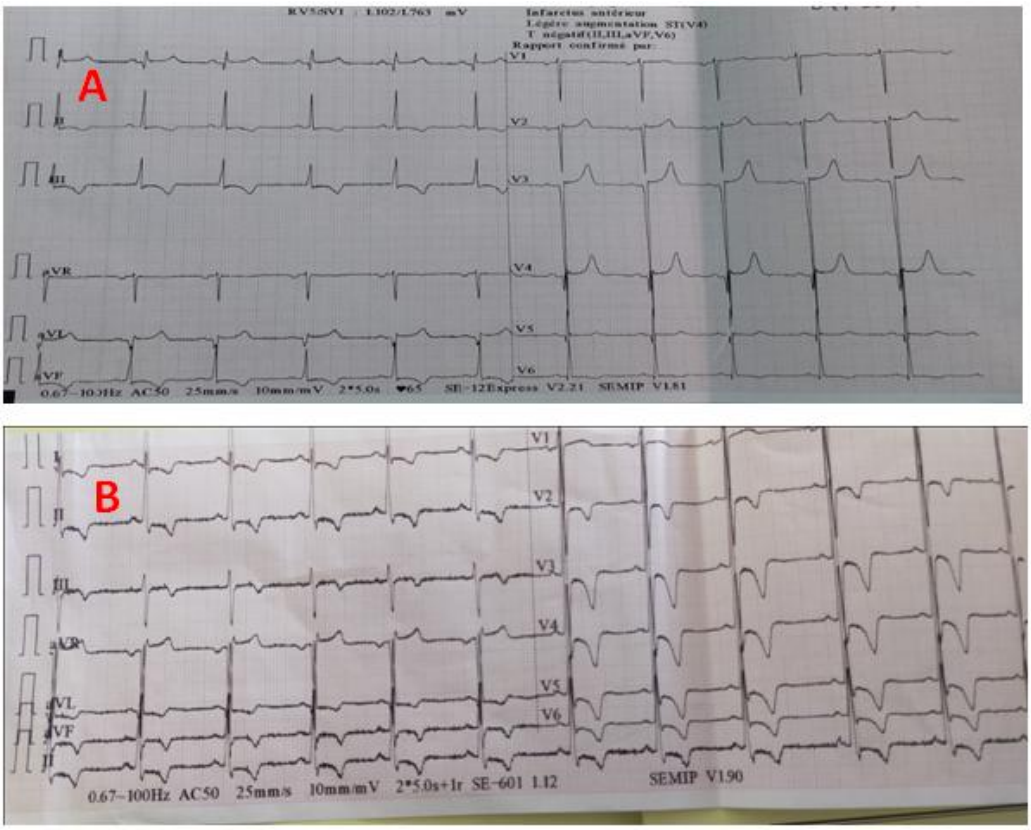

Fig. 1. Electrocardiograms of the two patients; A) CASE 1; B) CASE 2

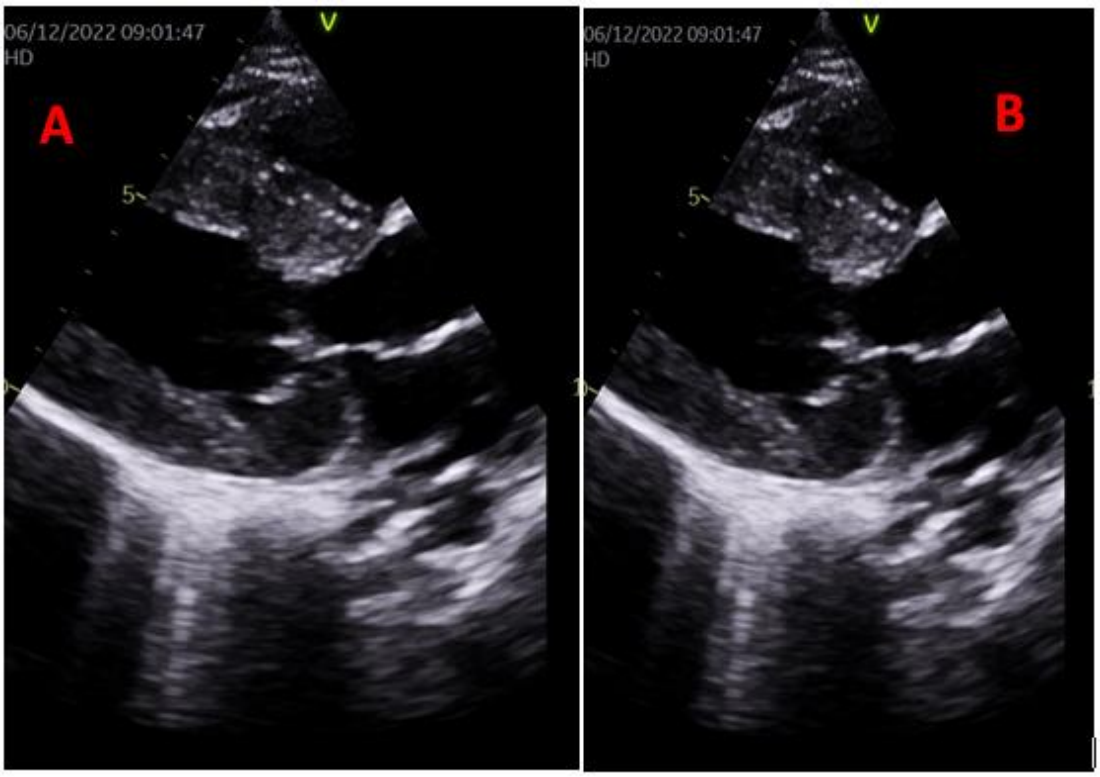

Fig. 2. Echocardiographic images showing concentric left ventricular hypertrophy with septal predominance. A) CASE 1, 2) CASE 2

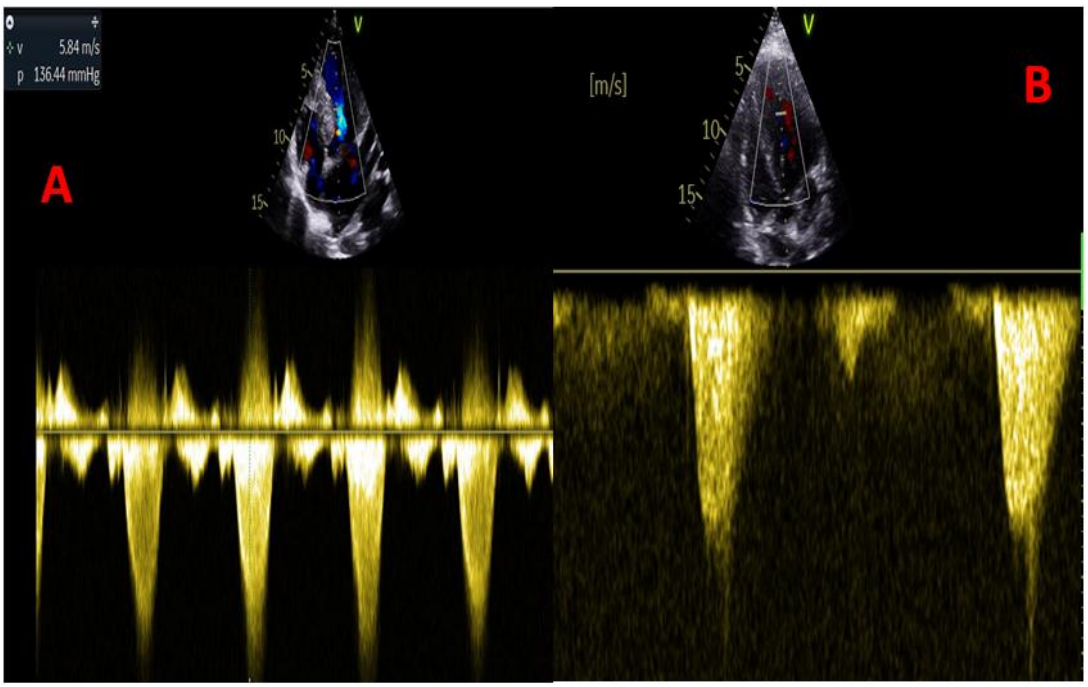

Fig. 3. Echocardiographic images showing a midventricular gradient. A) CASE 1 with a large gradient at 136mmHg, B) CASE 2 with a gradient at 35mmHg

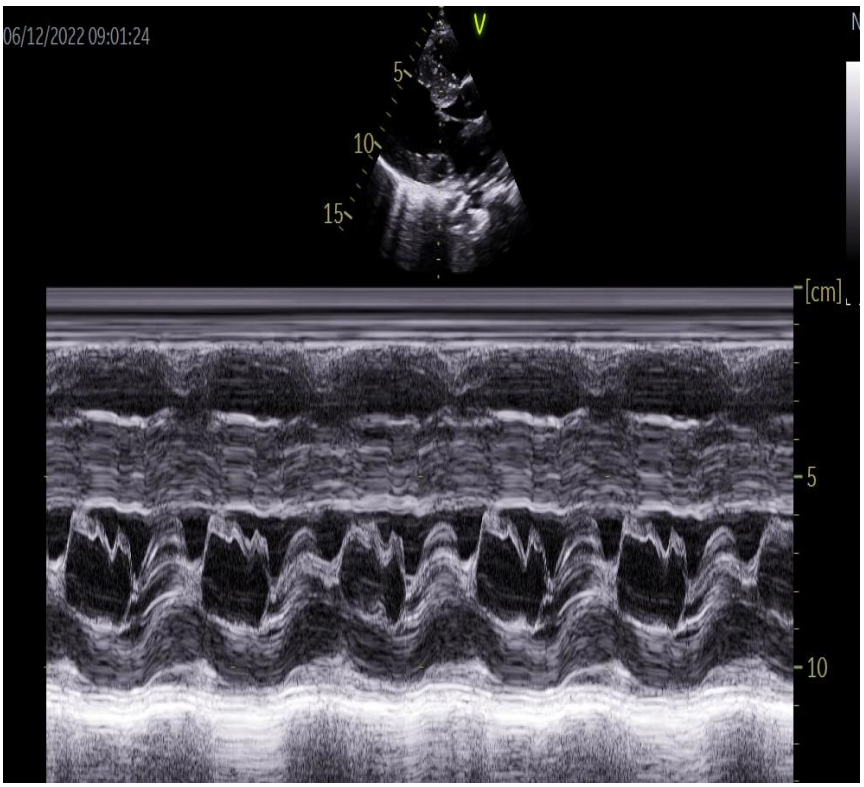

Fig. 4. Echocardiographic image objectifying a complete MAS in the first patient

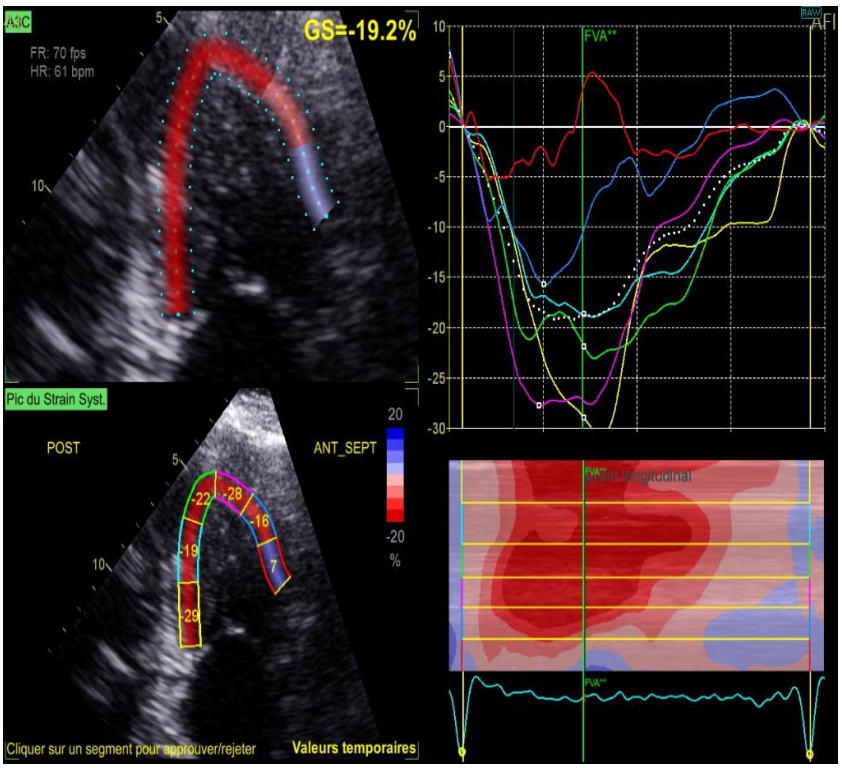

Fig. 5. Image showing the global longitudinal strain in the 2nd patient

After discussion with the gynecologists, medical termination of the pregnancy was not indicated for this patient, but close follow-up with monthly echocardiographic monitoring was necessary until delivery.

The patient is currently at 34 SA of pregnancy, followed monthly in our structure, she is clinically asymptomatic, with a stable gradient at echocardiography.

3. DISCUSSION

Hypertrophic cardiomyopathy (HCM) is the most common inherited heart disease. It is characterized by abnormal thickening of the heart wall, primarily the left ventricle, most often at the level of the interventricular septum, with no apparent clinical cause. This disorder is one of the leading causes of sudden death in young people [4].

HCM is a worldwide disease and is considered to be one of the most common inherited heart diseases, with reported prevalence varying widely between 1/500 (0.2%) and 1/3000 (0.03%), reflecting Different study designs and population characteristics were used [5]. Therefore, the incidence among pregnant women may be similar.

Left ventricular midline obstruction is a rare mechanism of obstruction caused by resistance to blood flow in the left ventricular midline lumen, occurring in approximately 10% of patients [6].

The detection of midventricular obstruction is based on an estimated 30 mmHg midventricular gradient. This blockage is attributed to a pronounced septal hypertrophy, leading to contact with the hypercontractile free wall of the left ventricle, according to the source [7].

Studies show that patients suffering from midventricular obstruction exhibit multiple symptoms, with dyspnea being the most prevalent. Moreover, these patients are at a higher risk of developing progressive heart failure and succumbing to sudden death or arrhythmic events [6,7].

Expectant mothers may experience heightened hemodynamic demands from the cardiovascular system during pregnancy, especially if pre-existing cardiomyopathy is present [8]. While the body adapts during pregnancy, the maternal physiological response may not meet increased demands. During pregnancy, plasma volume can surge up to 40% and cardiac output by 30-50%, primarily due to an increase in stroke volume and a small uptick in heart rate. The increase in stroke volume is secondary to an increase in blood volume and a decrease in systemic vascular resistance [9].

Most patients belonged to group II of the World Health Organization (WHO) revised maternal cardiovascular risk scheme. However, a subset of women with HCM may be at higher risk of adverse pregnancy outcomes, including women with severe left ventricular outflow tract obstruction, symptomatic arrhythmias, and moderate left ventricular systolic dysfunction, who are classified as high-risk class III modified WHO program [10].

Pregnancy is well tolerated by most patients with obstructive HCM, as increased volume and subsequent enlargement of the left ventricular cavity reduces left ventricular obstruction. However, this is usually offset by an increase in cardiac output. If these effects are unbalanced, patients with obstructive HCM may experience more symptoms during pregnancy [2].

Increased preload during pregnancy may decrease the left ventricular ejection tract gradient, whereas increased inotropy and decreased afterload increase the gradient [11].

Numerous complications have been described in women with HCM during pregnancy, such as heart failure, arrhythmias, syncope, and thromboembolic complications. This rate varied between 15% and 48%, depending on the study design and patient population [12,13].

A retrospective cohort study of pregnant women diagnosed with HCM and followed up at a single tertiary center between 1995 and 2019 included 18 women, 27 of whom had been pregnant for at least 20 weeks: 12 with obstructive Women with hypertrophic cardiomyopathy became pregnant and 15 women became pregnant with non-obstructive hypertrophic cardiomyopathy.

Left ventricular ejection obstruction is associated with increased cardiac events, including arrhythmias and heart failure. A patient with obstructive hypertrophic cardiomyopathy had her pregnancy terminated at 21 weeks because of uncontrolled arrhythmias. There is no maternal mortality ratio. In more than 50% of cases, preterm birth is due to maternal (40%) or fetal

(60%) causes. Most births are preterm between 34 and 36 weeks [14].

Analysis of data from nine cohorts (237 women and 408 pregnant women) showed that pregnancies in most women with HCM were uneventful. However, pregnancy in a woman with HCM carries risks for both mother and child. Maternal mortality was 0.5%, and complications or worsening of symptoms occurred in 29% of patients. Fetal mortality was caused by spontaneous abortion (15%), therapeutic abortion (5%), and stillbirth (2%). Preterm birth was observed in 26% [15].

The functional status and signs of heart failure before pregnancy are important risk factors for cardiac complications in pregnant women with HCM [16].

Preconception counseling, close monitoring, and optimal care are essential to avoid complications in women with HCM. In addition to monitoring of fetal growth by ultrasound in the third trimester and assessment of uteroplacental perfusion by Doppler studies in the second and third trimesters, it is warranted in all patients with hypertrophic cardiomyopathy, regardless of its severity [14].

4. CONCLUSIONS

Most women with HCM usually tolerate pregnancy well. However, left ventricular ejection obstruction has been associated with adverse cardiac events, including arrhythmias or heart failure.

This requires careful consultation by cardiologists and gynecologists before pregnancy, as well as close, professional treatment and monitoring to avoid complications between mother and fetus.

CONSENT

As per international standard or university standard, patient (s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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