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Successful Emergency Cesarean Section in A Hypertrophic Cardiomyopathy Patient Presenting with Progressive Heart Failure and Non-sustained Ventricular Tachycardia: A Case Report and Literature Review

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Abstract

Physiological status in pregnancy varies with the gestational age and postpartum period. Although most pregnancies in patients with hypertrophic cardiomyopathy are uneventful, the underlying conditions pose additional risks for maternal cardiac events. A 42-year-old woman with a 14-year history of hypertrophic cardiomyopathy presented with sudden-onset intermittent chest tightness since 2 days prior, along with dyspnea and near-syncope, at a gestational age of 36 weeks and 5 days. Echocardiography revealed that the mid-left ventricular wall thickness was increased to 31.2 mm without systolic anterior motion of the mitral leaflet or significant mid-left ventricular and outflow tract pressure gradient. The multidisciplinary team decided to deliver at a gestational age of 37 weeks due to an episode of non-sustained ventricular tachycardia. Cesarean section was performed, with a cardiologist, a cardiothoracic surgeon, and an extracorporeal membrane oxygenation team on standby. A male baby was delivered uneventfully. No further non-sustained ventricular tachycardia was observed postpartum. However, worsening heart failure symptoms were observed on the third day postpartum, which were resolved via fluid restriction and diuretics. A single-chamber implantable cardioverter defibrillator was placed 6 months after delivery. With risk stratification and adequate corresponding levels of care, pregnancies in patients with hypertrophic cardiomyopathy can be managed uneventfully. (J Intern Med Taiwan 2021; 32: 457-464)

Key Words: Hypertrophic cardiomyopathy, Heart failure, Non-sustained ventricular tachycardia, Pregnancy

Case presentation

A 42-year-old pregnant woman presented to our hospital at 36 weeks and 5 days of gestation with a two-day history of intermittent sudden-onset chest tightness, dyspnea, and near-syncope.

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Fourteen years prior to the consultation, the patient had experienced intermittent retro-sternal chest tightness that was more prominent at times of exertion and stress. This was associated with palpitations, dizziness, nausea, and cold sweats. Each episode had a duration of 2 hours and was partially alleviated with rest. These episodes progressed in severity and frequency. She also experienced exercise intolerance, and could only climb three flights of stairs. She visited a local clinic and was diagnosed with hypertrophic cardiomyopathy (HCM) on electrocardiography (ECG) and echocardiography. She was advised follow-up; however, she returned to our hospital only a year ago.

Physical examination at that time revealed a laterally displaced maximal apical impulse with an apical heave. An ECG revealed left ventricular hypertrophy (Figure 1). Echocardiography yielded findings compatible with apical hypertrophy with a wall thickness of 24.3 mm at the mid-left ventricular level (Figure 2). There was no systolic anterior motion of the mitral leaflet and no significant mid-left ventricular and outflow tract pressure gradient. A Holter monitor identified frequent ventricular ectopics.

Upon detailed inquiry, a significant family history of premature sudden cardiac death (SCD) was noted. Her father, uncle, and aunt had been diagnosed with HCM (Figure 3). Her father, who had experienced syncope, received an implantable cardioverter defibrillator (ICD) but died due to heart failure at 59 years of age. Her aunt had died suddenly at the age of 38, while her uncle had died of ischemic stroke at 54 years old. Despite planned genetic study and advised follow-up, the patient was lost to follow-up again.

Five months prior to the consultation, she returned to our obstetric clinic, then pregnant at a gestational age (GA) of 19 weeks. Echocardiography later revealed that wall thickness at the mid-left ventricular level was slowly increasing from 23.8 mm to 26.3 mm (measured at GA 19 and 31 weeks, respectively) (Figure 2). The other prenatal examinations were within normal limits.

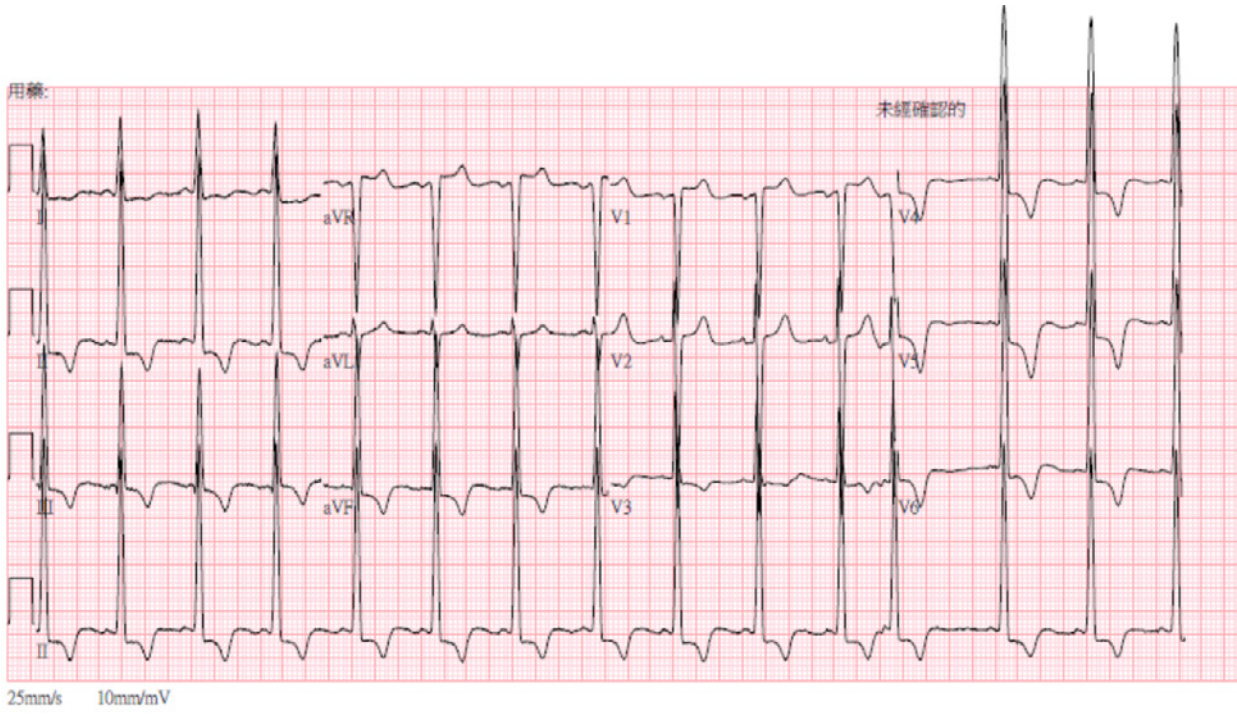

Figure 1. Electrocardiography demonstrating sinus rhythm with left ventricular hypertrophy pattern.

The pregnancy was relatively uneventful until sudden-onset chest tightness along with dyspnea, cold sweat, and near-syncope occurred at GA 36 weeks and 5 days. The symptoms lasted for one to two minutes during rest. She was brought to our emergency department whereupon occasional pre-

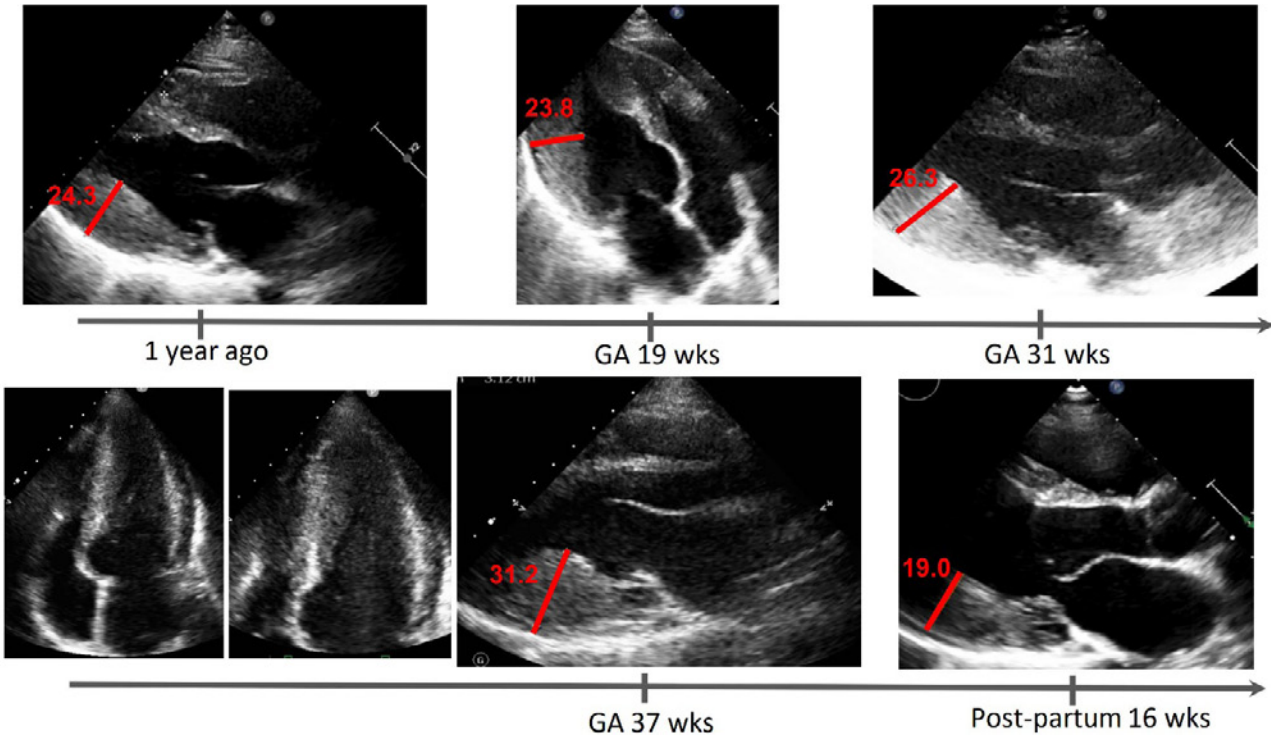

Figure 2. Transthoracic echocardiography demonstrating the temporal change of the mid-left ventricular thickness associated with gestational status. The measurements are in millimeters.

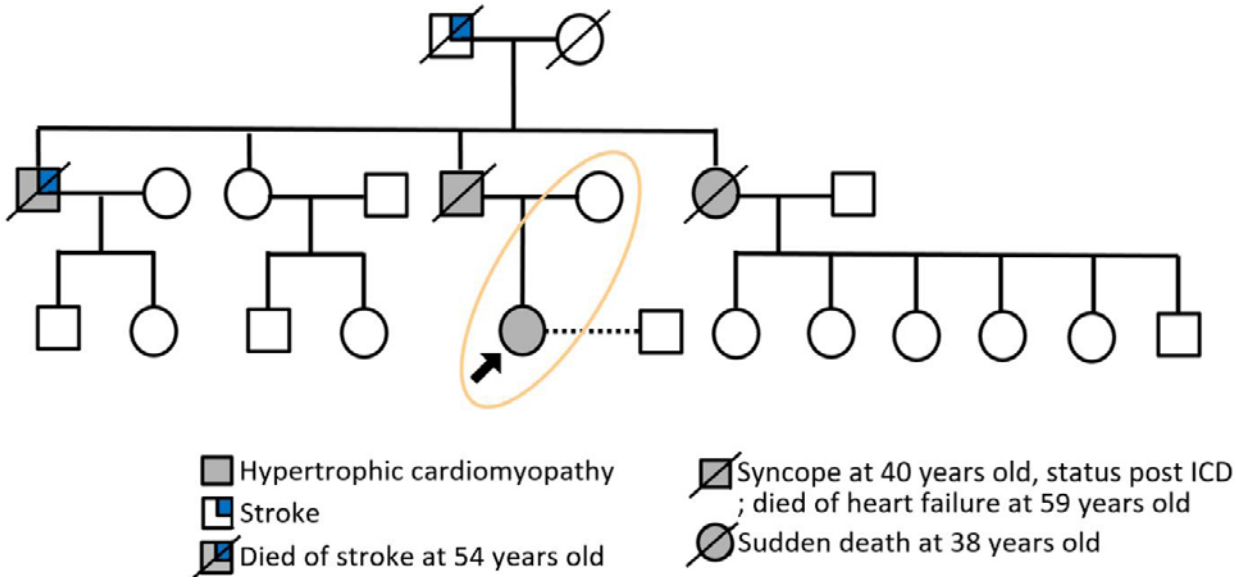

Figure 3. The paternal pedigree illustrates a significant family history of sudden cardiac death.

mature ventricular complexes were observed on the ECG monitor. The wall thickness at the mid-left ventricular level increased to 31.2 mm as measured on echocardiography (Figure 2). There was still no systolic anterior motion of the mitral leaflet or significant mid-left ventricular and outflow tract pressure gradient.

The patient was admitted to the obstetrics ward. A team led by an obstetrician and a cardiologist initially decided to deliver at term to reduce further maternal risks, and hence intramuscular betamethasone (12 mg) was administered. Two days later, telemetry recorded non-sustained ventricular tachycardia for 1.8 seconds (Figure 4) while the patient was asleep. She was transferred to our cardiac care unit and the daily administration of bisoprolol (1.25 mg) was initiated. A multidisciplinary care conference involving the original care team, an anesthesiologist, a cardiothoracic surgeon, the patient, and her family was urgently held on the same day. It was decided that cesarean section would be performed in the afternoon of the same day with a cardiologist, a cardiothoracic surgeon, and an extracorporeal membrane oxygenation (ECMO) team on standby. A mature male baby was delivered. The birth weight was 2600 g, and the Apgar scores were 7 and 9 at 1 and 5 minutes, respectively.

Postoperatively, the patient did not experience palpitations or chest tightness. She was transferred to the general ward two days later. However, on the third day postpartum, the patient complained of exertional dyspnea; bilateral pleural effusion was noted on ultrasound. Water restriction and diuretics were initiated, and the symptoms gradually reduced. Bisoprolol was replaced with metoprolol due to breastfeeding-associated safety concerns. Due to the patient's multiple risk factors for sudden cardiac death (SCD), such as a family history of SCD, massive left ventricular hypertrophy, and non-sustained ventricular tachycardia (NSVT), an ICD was considered for primary prevention. Six months after delivery, the patient finally agreed to undergo a single-chamber ICD implantation. The male infant was placed on regular follow-up with our pediatrician, and the developmental milestones were met for his age. Genetic counseling was recommended for the infant.

Discussion

Risk stratification of pregnant women with cardiovascular disease

The physiological state is altered significantly and dynamically during pregnancy, throughout different gestational ages and in the postpartum period. Cardiac output increases primarily due to plasma volume expansion during early pregnancy and accelerated heart rate in late pregnancy. The cardiac chambers also enlarge and the left ventricu-

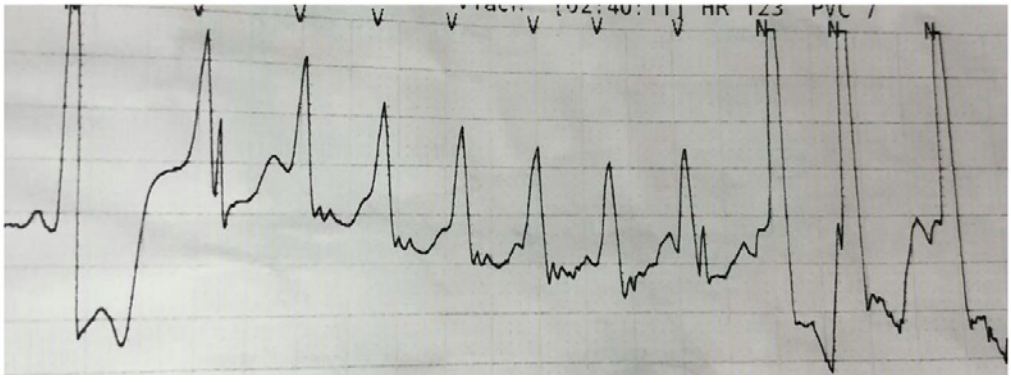

Figure 4. The telemetry recording the non-sustained ventricular tachycardia event.

lar wall mass increases. In addition to physiological changes, other factors that influence cardiac function are uterine contraction, pain, blood loss, and anesthesia[1]. The modified World Health Organization (mWHO) Classification of Maternal Cardiovascular Risk stratifies pregnancy-related risk according to different underlying maternal cardiovascular conditions. There are four risk categories, with class IV indicating extremely high risk for maternal mortality or severe morbidity, so that pregnancy becomes a contraindication. Class IV conditions include pulmonary arterial hypertension, severe systemic ventricular dysfunction (left ventricular ejection fraction (LVEF) < 30% or New York Heart Association (NYHA) functional class III-IV), previous peripartum cardiomyopathy with residual left ventricular impairment, severe symptomatic mitral or aortic stenosis, severe aortic dilatation, vascular Ehlers-Danlos syndrome, and severe (re)coarctation[1].

HCM is classified under mWHO class II-III, indicating intermediate increased risk of maternal mortality or moderate to severe increase in morbidity and a 10-19% maternal cardiac event rate during pregnancy. Therefore, trimestral to bimonthly follow-up visits during pregnancy are recommended for patients with HCM. These women should also have their delivery in a tertiary hospital[1]. The Registry of Pregnancy and Cardiac disease (ROPAC) provided a prospective cohort for pregnant women with structural heart disease. Using the database, Goland *et al*. analyzed 60 pregnancies in women with HCM. The study demonstrated no maternal mortality, but 23.3% of the pregnancies were complicated by major adverse cardiovascular events (MACE). The most frequently observed MACE were heart failure (15%) and ventricular tachycardia (10%). The first MACE is most frequently encountered during the third trimester, followed by the early postpartum period. NYHA functional classification ≥ II and signs of heart failure prior to pregnancy were highly correlated with the development of MACE. For patients with MACE, the duration of pregnancy was shorter and the rate of emergency cesarean section was higher[2]. Our patient had an increased risk of MACE according to these criteria. She developed non-sustained ventricular tachycardia in the third trimester, leading to an emergency cesarean section. The clinical course was complicated with progressive heart failure symptoms on the third day postpartum.

Tachyarrhythmia management during pregnancy

According to a nationwide database of pregnancy-related hospitalizations in the United States, the most frequent arrhythmias in pregnancy during the years 2000 to 2012 were atrial fibrillation (27/100,000), followed by supraventricular tachyarrhythmia (22/100,000) and ventricular tachycardia (VT, 16/100,000). The frequency of arrhythmia was greater overall in women aged 41–50 years. Furthermore, for mothers admitted with any arrhythmia, both the frequency of in-hospital death (5.9%) and maternal or fetal complications (36.5%) were greater than those in all women (0% and 21.8%, respectively). When more specifically targeting VT, the risk of mortality is elevated with an odds ratio of 40.89 (95% confidence interval 26.08–64.1; $P < 0.0001$)[3]. Narrowed down to women with HCM, the ROPAC registry reported that the prevalence of VT is as high as 10% but with zero mortality[2]. Our patient belongs to the medium risk classification of the European Society of Cardiology (ESC)[1]. Thus, the recommendations are consultation with a multidisciplinary team at a specialized center, cardiac rhythm monitoring, preparation for intravenous administration of antiarrhythmic drugs, and availability of an external cardioverter defibrillator on site[1]. If sustained VT occurs and causes hemodynamic instability, immediate electrical cardioversion[1] and ICD implantation is recommended[4]. Cardioversion has been found to be safe in all phases

of pregnancy without compromising fetal hemodynamics[5]. However, transient fetal arrhythmia has been reported; hence, fetal heart rate monitoring during cardioversion is advised[6]. For hemodynamically stable patients, beta-blockers are the preferred agents while monitoring fetal growth and cardiac rhythm[4].

Management of patients at risk of sudden cardiac death and ventricular ectopics

Two other patient groups worth discussing are women with an increased risk of sudden cardiac death (SCD) and those with a high burden of premature ventricular contraction (PVC). The 2014 ESC guidelines adopted the HCM-Risk SCD Score takes the following factors into consideration: age, family history of SCD, unexplained syncope, LV outflow gradient, maximum LV wall thickness, left atrial diameter, and NSVT. The 5-year risk of SCD was categorized as < 4%, 4%–6%, and > 6%. Our patient, with a calculated risk of 6.75%, should be considered for an ICD implantation[4,7]. None the less, the 2020 American Heart Association (AHA)/American College of Cardiology (ACC) guidelines adopted a policy for ICD implantation with a lower threshold. Prophylactic ICD is justified for patients with at least one of the following risks: family history of SCD, massive LVH (wall thickness ≥ 30 mm), unexplained syncope, apical aneurysm, LVEF ≤ 50%, NSVT, and extensive late gadolinium enhancement on cardiovascular magnetic resonance imaging[8]. The AHA/ACC strategy is more sensitive for predicting events that could terminate potentially lethal ventricular tachyarrhythmias, while the ESC model is associated with higher specificity for recognizing low risk patients[9].

For the influence of PVC burden, a prospective case-control study demonstrated that 11% of the pregnancies with high PVC burden (greater than 1% per day) developed maternal cardiac events, including heart failure, VT, and NSVT; to be specific, they all had a PVC burden greater than 5%. Fortunately, all cardiac events were successfully managed with medical therapy during pregnancy[10]. Relating this to our patient's case, her Holter examination prior to pregnancy revealed a high PVC burden (6.54%). Reflecting on her pregnancy course, which was complicated by NSVT, earlier beta-blocker use could have been beneficial.

Delivery modalities in patients with cardiovascular disease

From the obstetric point of view, vaginal delivery is preferred for patients with cardiovascular disease due to the lower associated blood loss, infection, and thromboembolic risks[11]. A cesarean section should be considered when the patient is receiving oral anticoagulation, and in cases of Marfan syndrome wherein an ascending aorta diameter > 45 mm, acute or chronic dissection, and acute heart failure are observed[1]. Our patient developed non-sustained ventricular tachycardia at 37 weeks of gestation without labor signs. The guidelines did not specify instructions on the delivery modality in patients with this condition. After a multi-disciplinary team discussion, a cesarean section was conducted in an adequately equipped operating theatre that allowed continuous monitoring with direct supervision from anesthesiologists, cardiologists, and cardiothoracic surgeons, with the ECMO team on standby.

Summary

Most women with HCM tolerate pregnancy well; however, there is still a significant risk of MACE. Physicians should be aware of the effects of the physiological changes during pregnancy on the variable underlying cardiac conditions of the patients. Genetic counseling may clarify the risks associated with the transmission of HCM and thus be used to lay out screening plans. Preconception assessment should be performed based on the

mWHO classification and risk for arrhythmia with hemodynamic compromise at delivery. The risk of SCD should be evaluated, and ICD implantation should be undertaken in appropriate patients. It is important to take into account the complex physiologic changes of pregnancy and their impact on any underlying cardiovascular pathology. Ventricular tachycardia occurs most frequently during the third trimester. For patients with a high PVC burden, prophylactic beta-blockers could be considered. In the event that VT does occur, electrical cardioversion is appropriate for patients with hemodynamic instability, while beta-blockers may be given to those who are stable. On the other hand, heart failure develops most often in the early postpartum period, and thus the fluid status should be cautiously managed. In selecting medications to administer, any placental transfer to the fetus and presence in the breast milk should be taken into consideration. A delivery plan should be formed at the end of the second trimester by a multidisciplinary team[4]. Vaginal delivery is generally preferred, but cesarean section could be considered in certain conditions such as severe LV outflow tract obstruction or severe heart failure. Careful monitoring of heart rate and rhythm during delivery is recommended[4]. Finally, genetic and clinical screening for the offspring should be performed early in order to arrange follow-up, lifestyle plans, and timely intervention to prevent adverse events[8,12,13].

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肥厚型心肌病變孕婦伴隨心衰竭惡化及非持續性心室過速緊急剖腹產成功：案例分析與文獻回顧

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摘 要

孕婦生理隨著孕程有相應變化，大部分肥厚性心肌病變的孕婦能夠適應並生產，但仍有一定風險。病人是位有14年肥厚性心肌病變病史的42歲女性，在懷孕36周5天時感到胸悶，伴隨喘、近昏厥；心臟超音波顯示左心室中部厚度達31.2毫米，無心室中、出口的壓力差；37周時觀察到非持續性心室心搏過速，在心臟內外科及葉克膜團隊陪同下順利進行剖腹產。產後第3天出現心衰竭症狀，經限水和利尿劑使用後恢復，未再出現心律不整，病人在6個月後置放植入式心臟去顫器。根據風險分級給予適當照顧，能降低患肥厚型心肌孕婦的生產風險。