



## HYPERTROPHIC CARDIOMYOPATHY

### Background

Hypertrophic cardiomyopathy (HCM) is characterized by left ventricular hypertrophy (typically asymmetric and septal in distribution, although almost any pattern of hypertrophy can be present) with a maximal wall thickness usually exceeding 15 mm in the absence of another explanation for the hypertrophy such as aortic stenosis or hypertension. The left ventricular hypertrophy is generally associated with a nondilated and hyperdynamic chamber. Left ventricular outflow tract obstruction can be a feature of the disease present under resting conditions although more commonly the obstruction is dynamic occurring or exacerbating with exercise and/or provocative maneuvers. When left ventricular outflow tract obstruction is present, the condition can be called hypertrophic obstructive cardiomyopathy (HOCM). The obstruction can be either subaortic or at the mid cavity level. The former is caused by systolic anterior motion of the anterior mitral valve leaflet and mid-systolic contact with the ventricular septum resulting in mid to late systolic outflow tract obstruction and concomitant mitral regurgitation. Left ventricular systolic dysfunction can also occur in a minority of cases as a result of myocardial disarray and fibrosis that occurs as a result of abnormal sarcomere replication. This can also create an arrhythmogenic substrate predisposing patients to ventricular tachyarrhythmias.

The majority of cases of hypertrophic cardiomyopathy (HCM) are familial and are inherited in an autosomal dominant fashion. The condition is attributable to a mutation in one of a number of genes that encode for sarcomere proteins.

Clinical presentation can occur at any phase of life with up to 25% of individuals with HCM achieving normal longevity (1). Many women with HCM encountered in pregnancy are asymptomatic. Cardiac complications include:

- a) sudden cardiac death
- b) presyncope/syncope
- c) embolic stroke
- d) arrhythmias including atrial fibrillation
- e) progressive heart failure
- f) angina

The overall risk of disease-related complications is 1-2% per year (2).

Risk factors for sudden cardiac death include:

- a) prior cardiac arrest
- b) family history of sudden cardiac death
- c) unexplained syncope,
- d) ventricular tachycardia
- e) abnormal blood pressure response to exercise
- f) significant left ventricular hypertrophy (echocardiographic wall thickness > 30 mm).

### Effects of pregnancy-related hemodynamic changes

The pregnancy-related decrease in systemic vascular resistance and increase in myocardial contractility can both worsen and/or unmask left ventricular outflow tract obstruction and associated

symptoms. (see Cardiovascular Changes During Pregnancy) However, the concomitant increase in intravascular volume may attenuate any increase in outflow tract obstruction. Women with significant left ventricular systolic and/or diastolic dysfunction may decompensate as a result of the increase in intravascular volume and cardiac output. Moreover, the increasing hemodynamic burden, in association with the hormonal changes of pregnancy, may precipitate arrhythmias such as atrial fibrillation.

### **Maternal cardiac complications**

Several contemporary series of pregnancies among women with HCM have been reported (3, 4, 5). Women with HCM who are asymptomatic prior to pregnancy and have preserved ventricular function, usually tolerate pregnancy well and few develop significant functional decline. In contrast, women with symptoms prior to pregnancy are at substantial risk of having a significant decline in functional capacity or developing heart failure. Pregnant women are at risk for atrial fibrillation, although this is relatively rare. Women may also develop syncope in the setting of arrhythmias. The development of significant symptoms during pregnancy is more common in women with HCM and left ventricular outflow tract obstruction. Maternal sudden death has been reported in women with high risk features described above. (5)

In one study, women with symptoms prior to pregnancy were at risk of preterm delivery (18%). (4)

### **Fetal complications**

In an uncomplicated pregnancy, the main risk to the fetus is the 50% recurrence rate. HCM is not usually apparent in childhood and offspring should continue to be screened well into adulthood, unless there is a known genetic marker that can exclude the inheritance. Fetal deaths have been reported. (3)

### **Management strategies**

#### **Preconception counseling/Contraceptive methods**

Ideally, a comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination, an echocardiogram and an electrocardiogram. The additional prognostic benefit of cardiopulmonary exercise testing has not been defined, but may be helpful in some cases.

Asymptomatic women generally do well through pregnancy. Women with symptoms prior to pregnancy are at higher risk for complications (decline to NYHA functional class III-IV, pulmonary edema or arrhythmias) during pregnancy. Women with clinical heart failure should be advised about the high risk of pregnancy, as they may be unable to tolerate the increased hemodynamic load.

Decisions regarding internal cardioverter defibrillator (ICD) placement for prevention of sudden cardiac death and invasive therapies to decrease symptoms (i.e. surgical myectomy, alcohol ablation and biventricular pacing) should be made prior to pregnancy.

Transmission of heart disease to offspring should be discussed. The risk of transmission of HCM is approximately 50%, compared to a background risk of any congenital heart disease of 1%.

A discussion about contraceptive methods is appropriate in all women with HCM. Combined oral contraceptives (estrogen/progestin) are not advised in women with cardiomyopathy and left ventricular ejection fractions < 30%. (see Contraception)

Medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (<http://www.motherisk.org>) is an excellent resource.

### **Ante-partum care**

Coordinated care with a heart specialist and a high-risk obstetrician should be implemented. The frequency of assessments (clinical and echocardiographic) during pregnancy should be determined on the basis of the maternal functional status, the family history of the women, the systolic and diastolic ventricular function, the degree of outflow tract obstruction and the severity of mitral regurgitation.

Close cardiovascular monitoring, with specific attention to volume status, is important throughout pregnancy and the peripartum period. Volume depletion should be avoided as it can precipitate left ventricular outflow tract obstruction in women with hypertrophic obstructive cardiomyopathy. In contrast, volume overload should be avoided among women with heart failure and/or systolic/diastolic left ventricular dysfunction. Treatment for symptomatic heart failure may be necessary in some women.

Atrial fibrillation can be treated medically or with DC cardioversion when women are unstable or unresponsive to medical therapy. (see Arrhythmias) Women treated with warfarin prior to pregnancy should be seen by a hematologist to develop an anticoagulation plan for the pregnancy.

Women should be offered fetal echocardiography at approximately 20 weeks gestation.

### **Labour and delivery**

Labour and delivery should be planned carefully with a multidisciplinary team well in advance. It is important to communicate the delivery plan to the woman and to other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Generally, vaginal deliveries are recommended unless there are obstetric indications for a cesarean delivery. Good pain management for labour and delivery is very important in order to minimize maternal cardiac stress. Regional anesthesia should be carefully administered to avoid hypotension as vasodilation is poorly tolerated. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. To decrease potential harmful complications from difficult mid cavity-assisted delivery, uterine contractions are often utilized to facilitate the initial descent of the presenting part. Oxytocin can induce vasodilation and arterial hypotension and should be administered with great care if truly needed. Hypovolemia or blood loss should be aggressively corrected.

The need for maternal cardiac monitoring at the time of labour and delivery is dictated the women's functional status, the degree of ventricular dysfunction, and the degree of left ventricular outflow tract obstruction. Most women with HCM do not require invasive monitoring. To detect potential arrhythmias early, continuous electrocardiographic monitoring may be helpful in some instances.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with HCM. However, some experts continue to administer antibiotics because they feel that the risks of adverse reactions to antibiotics are small and the risk of developing endocarditis has major health consequences.

## **Post-partum care**

The hemodynamic changes of pregnancy may take up to six months to normalize. Women should be seen early after pregnancy (usually within 6-8 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.

## **References**

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