

### CARDIOMYOPATHY

### Background

Cardiomyopathy in women of childbearing age can be due to a number of conditions including:

- Peripartum cardiomyopathy/pregnancy-associated cardiomyopathy
- Idiopathic dilated cardiomyopathy some of these cardiomyopathies may be familial
- Drug-induced cardiomyopathy (for example, Adriamycin)
- Myocarditis
- Coronary artery anomalies

Women may present with deteriorating left ventricular systolic function, clinical heart failure, chest pain, atrial or ventricular arrhythmias, stroke or other embolic events, or sudden cardiac death. (1, 2, 3, 4, 5, 6, 7)

Peripartum cardiomyopathy requires special consideration. (4, 5, 6, 7) This form of cardiomyopathy has been defined as a cardiomyopathy presenting in the last month of pregnancy or within 6 months postpartum in the absence of other causes of cardiomyopathy. However, women may present earlier in pregnancy, which likely represents a variant of the same disease (the term "pregnancy-associated cardiomyopathy" has been suggested). The causes of peripartum cardiomyopathy are not known, but risk factors have been identified and include:

- a) African race
- b) Older maternal age
- c) Multiple pregnancy
- d) Multiparity
- e) Pre-eclampsia
- f) History of previous peripartum cardiomyopathy

#### Effects of Pregnancy Related Hemodynamic Changes

Pregnancy results in an increase in blood volume, heart rate and cardiac output. (see Hemodynamics in Pregnancy) Women with cardiomyopathies may not be able to adequately increase their cardiac output to meet the demands of pregnancy. Additionally, the increased blood volume may precipitate heart failure.

#### Maternal Cardiac Complications

**Dilated cardiomyopathy.** The most common maternal cardiac complications during pregnancy are heart failure and arrhythmias. In one study of pregnancies in women with idiopathic or Adriamycin-induced cardiomyopathy, 39% of the pregnancies were complicated by heart failure or arrhythmias. (1) Maternal deaths have been reported. (2)

<u>Peripartum cardiomyopathy.</u> The most common maternal cardiac complications during pregnancy are heart failure, arrhythmias and thromboembolic complications (systemic and pulmonary). Peripartum cardiomyopathy has high morbidity and mortality.

Mortality rates during the index pregnancy vary between 25-50%, (4) although they have been noted to be decreasing in recent series. (6) Maternal deaths typically occur within months after delivery. Presentation may be subclinical. While peripartum cardiomyopathy can be associated with full recovery (between 25-50% of women), a significant number of women are left with residual left ventricular systolic dysfunction. Recovery, when it occurs, usually occurs within a few months after delivery. Women should receive heart failure therapy, i.e. beta blockers, angiotensin converting enzyme inhibitors, at least until the left ventricle recovers. ACE Inhibitor therapy must be delayed until post-partum. Beta blocker therapy may be considered during pregnancy.

## **Fetal Complications**

Potential adverse fetal outcomes include premature delivery, low birth weight and intrauterine growth retardation. Fetal deaths have been reported. In one series, women with dilated cardiomyopathy had a 20% adverse fetal and/or neonatal event rate. (1) In women with peripartum cardiomyopathy and left ventricular systolic dysfunction prior to pregnancy, the risk of preterm delivery is high at 37%.

In women with dilated cardiomyopathy, fetal risks are highest in women who also have obstetric risk factors (history of premature delivery or rupture of membranes, incompetent cervix, uterine/placental abnormalities, prior cesarean delivery, intrauterine growth restriction or antepartum bleeding > 12 weeks). (1)

#### **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 or dobutamine stress echocardiography (contractive reserve) has not been defined, but may be helpful.

**Dilated cardiomyopathy**. The main determinants of adverse maternal events during pregnancy are moderate or severe left ventricular systolic dysfunction and/or NYHA functional class III or IV. (1) Asymptomatic women with dilated cardiomyopathy and only mild left ventricular systolic dysfunction, generally do well during pregnancy. Women with symptoms prior to pregnancy or moderate or severe left ventricular systolic function are at higher risk for complications during pregnancy.

**Peripartum cardiomyopathy.** Women with peripartum cardiomyopathy are at risk for adverse maternal complications with all subsequent pregnancies. Maternal cardiac complications are common with subsequent pregnancies and are dependent on the left ventricular systolic function. In women with restoration of normal left ventricular function prior to a subsequent pregnancy, the risk of clinical heart failure during pregnancy is 19%. Fourteen percent of women have a persistent decrease in left ventricular systolic function prior to a subsequent pregnancy. In women with decreased left ventricular systolic function prior to a subsequent pregnancy, the risk of clinical heart failure is 44% and the risk of maternal death during pregnancy is 19%. Twenty-five percent of women will have a persistent decrease in left ventricular ejection fraction >20% after the subsequent pregnancy. (5) These findings highlight the importance of preconception counseling in women with this condition.

Pregnancy in women with peripartum cardiomyopathy and persistent left ventricular dysfunction (EF <50%) is contraindicated.

A discussion about contraceptive methods is important in all women with cardiomyopathies. Combined oral contraceptives (estrogen/progestin) are contraindicated in women with cardiomyopathy and left ventricular ejection fractions < 30%. (see Contraception)

Women treated with angiotensin converting enzyme inhibitors or angiotensin receptor blockers will need to have these medications stopped prior to pregnancy. Assessment of ventricular function after discontinuation of therapy is useful. Medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (<u>http://www.motherisk.org</u>) 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 clinical status of the mother and the left ventricular systolic function.

Close cardiovascular surveillance, with specific attention to volume status, is important throughout pregnancy and the peripartum period.

Treatment for symptomatic heart failure with bed rest, oxygen and/or diuretic therapy may be necessary for some women. Because angiotensin-converting enzymes inhibitors and angiotension receptor blockers are contraindicated, hydralazine and nitrates may be considered for afterload reduction. The benefits of beta blockers in pregnant women with impaired systemic ventricular systolic function have not been studied, but they are likely helpful.

Cardiotoxic subfragments of prolactin are thought to play a role in the pathophysiology of peripartum cardiomyopathy, and therefore blockade of prolactin by bromocriptine has been suggested. (7)

Women treated with warfarin prior to pregnancy should be seen by a hematologist to develop an anticoagulation plan for the pregnancy. Mural thrombi in the left ventricle are relatively common in women with peripartum cardiomyopathy and full anticoagulation is indicated if one is identified.

Women with clinical heart failure often need to be managed in a coronary care unit. Some women may require intubation and ventilation and/or support with intra-aortic balloon pumps or ventricular assist devices. In these instances, it is important to have a cardiac transplant team involved for transplant evaluation.

# 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 for women in stable condition 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. 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.

The need for maternal cardiac monitoring at the time of labour and delivery is dictated by the women's clinical status and her degree of left ventricular systolic dysfunction. Oximetry or continuous electrocardiographic monitoring may be helpful in some instances. Women with cardiomyopathies and clinical heart failure may require invasive blood pressure monitoring with an arterial line.

Volume status should be carefully assessed postpartum and diuretic therapy should be initiated when necessary.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with cardiomyopathy.

# **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 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.

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