HEART DISEASE & PREGNANCY

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GENERAL CONSIDERATIONS

Every woman with acquired or congenital heart disease considering pregnancy should undergo preconceptual cardiac counseling by a cardiologist with expertise in pregnancy and heart disease. If pregnancy has already occurred, a comprehensive assessment by a cardiologist with experience in pregnancy should be organized early in pregnancy.

Questions to Consider During Preconceptual Counseling of Women with Heart Disease

1. What are the risks of pregnancy for the mother?
2. What the risks of contraception for the mother? (see Contraception)
3. Which medications are safe during pregnancy?
4. What are the late effects of pregnancy on the heart?
5. What is the overall prognosis for the mother?
6. What are the risks for the fetus/neonate?
7. For mothers with congenital heart disease, what is the probability of transmission of congenital heart disease to offspring?

What are the risks of pregnancy for the mother?

In a large prospective study of women with heart disease, adverse maternal cardiac events occurred in 13% of pregnancies. (1) The probability of an adverse event in women with congenital heart disease varies from 7.6% (3) to 19.4%. (2)

The most common complications that occur during pregnancy in women with heart disease are:

1. Arrhythmias
2. Heart failure
3. Stroke
4. Other embolic complications
5. Endocarditis
6. Myocardial infarction
7. Death –cardiac and noncardiac

Probability of a maternal complication varies according to the underlying cardiac lesion. (see lesion specific sections on website) (4)

In addition, there are general (or global) cardiac factors that are predictors of increased risk of cardiac complications during pregnancy. These include: (1,2,3)

1. Poor functional status (NYHA functional class > II)
2. Cyanosis (oxygen saturation at rest < 90%)
3. Left ventricular systolic dysfunction (ejection fraction < 40%)
4. Right ventricular systolic dysfunction and/or severe pulmonary regurgitation
5. Moderate or severe systemic atrioventricular valve regurgitation
6. Moderate or severe pulmonary atrioventricular valve regurgitation
7. Left heart obstruction
8. Mechanical valve prosthesis
9. History of cardiac events prior to pregnancy (arrhythmia, pulmonary edema or stroke)

Risk scores have been proposed to quantitate global cardiac risk of pregnancy. The original risk score from a Canadian consortium (CARPREG risk score) (1) is based on 4 risk predictors: 1) poor functional status (NYHA class > II) or cyanosis, 2) systemic (left) ventricular systolic dysfunction, 3) left heart obstruction, and 4) history of prior cardiac events (arrhythmia, stroke, heart failure). Each predictor is assigned one point. Patients with 0 predictors were at low risk (5%), patients with 1 predictor were at intermediate risk (25%) and those with >1 predictor were at high risk (75%) of adverse cardiac events during pregnancy. The risk score by Khairy et at (3) incorporated right ventricular systolic dysfunction and/or severe pulmonary regurgitation into the CARPREG risk score. The most recent risk score from the ZAHARA investigators, (3) is a weighted scoring system and incorporates a number of new variables into the risk prediction model including: a) cardiac medications before pregnancy, b) systemic atrioventricular regurgitation, and c) mechanical valve prosthesis.

In addition to the above global risk predictors, lesion specific risks should be incorporated into risk assessment. Some conditions known to have high pregnancy risk may not have been adequately represented in the large cohort studies from which the global risk predictors were derived. Women at high risk for complications but not identified by the risk predictors above include those with Marfan syndrome and dilated aortic roots, Fontan operations, or peripartum cardiomyopathy.

Which medications are safe during pregnancy?

Many medications are not safe during pregnancy. Medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. Some commonly used cardiac medications that are not safe in pregnancy are:

1. Warfarin
2. Angiotensin converting enzyme inhibitors
3. Angiotensin receptor blockers
4. Bosentan
5. Amiodarone
6. HMG CoA reductase inhibitors

The MOTHERISK website is an excellent resource. (http://www.motherisk.org)

What are the late effects of pregnancy on the heart?

In general, we do not fully understand the impact of pregnancy on the natural history of any cardiac condition. This remains an important unanswered question. However, there are a small number of studies that have suggested pregnancy may have late adverse effects on the heart. In women with peripartum cardiomyopathy, subsequent pregnancies may result in permanent ventricular dysfunction. (5) Worsening systemic right ventricular dilation, systolic dysfunction and tricuspid regurgitation are described in women with transposition of the great arteries and Mustard or Senning operations. In some cases these changes are irreversible. (6) Increase in the subpulmonary ventricular size late after pregnancy has been reported in women with tetralogy of Fallot. (7) Women with congenital aortic stenosis who have undergone pregnancy have increased rates of late cardiac events compared to those who have never been pregnant. (8)

What is the overall prognosis for the mother?

Long-term prognosis can be a difficult topic, but needs to be addressed in spite of its sensitivity. Long-term prognosis is based on the underlying cardiac condition, the medical history and the associated

residual lesions. This should be addressed by a physician with a good understanding of the mother's underlying heart condition.

What are the risks for the fetus/neonate?

Miscarriage rates are increased in women with cyanotic heart disease. In women with pre-pregnancy maternal oxygen saturation ≤ 85%, the chance of a live birth is only 12%. (9)

In a large prospective cohort study of women with heart disease, fetal and neonatal adverse events occurred in 20% of pregnancies. The most common adverse events were premature birth and small-for-gestational-age birth weight babies. Respiratory distress syndrome and intraventricular hemorrhage were often associated with prematurity. Rarely, pregnancies were complicated by fetal or neonatal deaths. (1)

Predictors of poor fetal/neonatal outcomes include: (1,2)

1. Poor maternal functional class
2. Cyanosis
3. Left heart obstruction
4. Mechanical heart valves
5. Anticoagulation
6. Smoking during pregnancy
7. Twin or multiple gestation

For mothers with congenital heart disease, what are the risks of transmission of congenital heart disease to offspring?

Transmission of congenital heart disease to offspring should be discussed with all women and men with congenital heart disease planning a pregnancy. The risk of transmission of congenital heart disease if a parent has congenital heart disease is between 5-50%, compared to a background risk of 1%. Risk of transmission varies depending on the underlying cardiac condition of the mother or father. For lesions with autosomal dominant inheritance (such as DiGeorge, Marfan, or Noonan syndrome), the risk of recurrence of congenital heart disease is 50%. Women can be offered a fetal echocardiogram at 20 weeks gestation.

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