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Preconceptional evaluation of women with heart disease

Marla A. Mendelson

INTRODUCTION

Women reaching childbearing age may present with congenital or acquired cardiac disease. Cardiac complications arising during pregnancy impact morbidity and mortality of mothers and their unborn children. Anticipating cardiac problems in light of the expected hemodynamic changes of pregnancy is the basis of the preconceptional evaluation. Risks specific to each woman's particular disorder should be identified prior to conception, and intervention(s) may be required to improve cardiac status prior to pregnancy. For some women, pregnancy may worsen their cardiac condition and functional capacity, thus impacting their ability to gestate and, at a later date, to parent.

The normal hemodynamic changes of pregnancy pose potential problems for many women with pre-existing heart disease. Although the medical literature is replete with studies and case series of women who have successfully completed pregnancies despite having cardiovascular disease, it is important to anticipate those problems that may arise specific to the underlying heart disease[1–8]. The automatic dismissal of the possibility for successful pregnancy in a woman with underlying heart disease is inappropriate, although women with certain types of heart disease may not be able to sustain pregnancy without grave risk.

Overall, the objective is to provide a rational plan to encourage a pregnancy that is safe for both the mother and her developing fetus, to identify potential complications that may occur during pregnancy, and to review medications that may require change in anticipation of pregnancy. As most cardiac diseases have been reported during pregnancy, certain guiding principles have emerged for preconceptional evaluation and the potential complications that could arise during a pregnancy. Foremost, women with pre-existing heart disease require full evaluation prior to conception. Second, not only should the risk to the mother be identified but also potential risks to the developing fetus need be clarified and understood. For example, cyanotic heart disease has been associated with fetal prematurity, dysmaturity or low birth weight. Third, it is axiomatic that a woman with congenital heart disease may transmit this condition to her offspring. Finally, it is important to identify interventions that may improve the prognosis for any outcome of the pregnancy.

Recent literature has focused on outcomes in women with pre-existing heart disease and proposed and validated a risk scoring system[1,3,6,7,9,10]. Depending upon the country, the type of pre-existing cardiac disease in women of childbearing age may differ. For example, 55% of Brazilian patients had pre-existing rheumatic heart disease, whereas only 19% exhibited a congenital heart condition[6]. Distribution is different in studies based in the United States and Canada wherein 74% of

women had underlying congenital heart disease, 22% acquired heart disease and 4% isolated arrhythmias.

The adverse cardiac events common to all the studies include congestive heart failure, pulmonary edema, arrhythmias, thromboembolic events, angina, endocarditis and a decrease in the New York Heart Association functional class. Canadian researchers developed a risk score of one for each identified predictor of risk which includes: (1) prior cardiac event (heart failure, transient ischemic attack or stroke); (2) New York Heart Association class greater than two or the presence of cyanosis; (3) left heart obstruction with mitral valve area less than $2\,cm^2$, or aortic valve area less than $1.5\,cm^2$, or left ventricular gradient greater than $30\,mmHg$; and (4) ejection fraction less than 40%. In the population of women with congenital heart disease: zero risk points was associated with a 5% risk of adverse events; one risk predictor carried an 18% risk of adverse events; and two or more risk factors predicted a complication rate of 57%[10]. Predictors for neonatal complications included poor functional class of the mother or cyanosis, left heart obstruction, anticoagulation or multiple gestation.

HEMODYNAMIC RISKS OF PREGNANCY IN WOMEN WITH CARDIAC DISEASE

The overriding concern regarding maternal morbidity and mortality relates to the type of underlying heart disease and the degree of functional limitation. Women with pulmonary hypertension or systemic ventricular dysfunction cannot tolerate the hemodynamic changes of pregnancy; therefore, these problems constitute contraindications to pregnancy. Contraindications to pregnancy are listed in Table 1.

The expected hemodynamic changes of pregnancy vary with respect to the type of cardiac lesion. For example, the woman with mitral regurgitation, because of the vasodilatation and decrease in afterload occurring during pregnancy, may actually have a decrease in her valve regurgitation during pregnancy. The hemodynamic changes in pregnancy are summarized in Table 2. Simply put, the changes of pregnancy may be deleterious and not tolerated in the setting of impaired systemic ventricular function[5,9]. During pregnancy with its associated 50% increase in blood volume, the heart may become dilated, and this may lead to further dysfunction in an already compromised ventricle. Because these changes may

Table 1 Cardiac contraindications to pregnancy

Contraindication	Potential risk
Absolute: cardiac condition imperils maternal and fetal outcome	Worsening of function
Systemic ventricular dysfunction (New York Heart Association functional class III, IV)	Congestive heart failure; postpartum impairment
Pulmonary hypertension/Eisenmenger's syndrome	Increased maternal mortality; cyanosis; fetal loss
Relative contraindication: high-risk pregnancy anticipated	
Severe mitral or aortic valve stenosis	Congestive heart failure; atrial fibrillation; thromboembolism
Aortic dilatation (Marfan syndrome/ bicuspid aortic valve)	Aortic dissection or rupture
Prosthetic (mechanical) heart valve	Valve thrombosis or bleeding

Table 2 Summary of expected hemodynamic changes of pregnancy

Hemodynamic alteration	Time of peak effect	Potential risks
Cardiac output ↑30–50%	20–24 weeks	Women with limited cardiac function or reserve may develop congestive heart failure
Stroke volume ↑20%	20–24 weeks	Increased preload is a problem for obstructive lesions (mitral or aortic stenosis) or ventricular dysfunction
Heart rate ↑10–20%	Third trimester	Tachycardia causes palpitations and impairs ventricular filling
Blood volume ↑40%	20–24 weeks	'Physiologic' anemia of pregnancy caused by reduced increase in erythrocyte mass
Peripheral vasodilatation	Throughout	↓Blood pressure; ↓valvular regurgitation
↑Minute ventilation	Second trimester	Sensation of tachypnea or dyspnea

persist during the postpartum period as well, it is important that the preconceptional evaluation should attempt to anticipate whether further cardiac decompensation may occur as a result of the pregnancy.

Pulmonary hypertension constitutes a contraindication to pregnancy (Table 1). This may occur as primary pulmonary hypertension, which commonly occurs in young women; certainly any symptoms of exertional chest pain or syncope may be a manifestation of pulmonary hypertension. Pulmonary artery pressure may be estimated using non-invasive echocardiography testing. If concern is present about elevated pulmonary pressure, the patient may need to proceed to cardiac catheterization to assess the intracardiac and intravascular pressures, as pulmonary hypertension has been associated with 50% maternal mortality. Even secondary pulmonary hypertension due to the underlying congenital heart disease confers a risk of mortality. A woman having undergone surgical repair may have residual hypertension that could confer a pregnancy risk. Such risks persist into the postpartum period as well[5].

MATERNAL MEDICATIONS

It may not be prudent to abruptly stop all previously prescribed medications once it is known that a woman with a diagnosed cardiac condition is pregnant[11,12]. Medication choices are an important aspect of preconceptional counseling. Alterations in medications should be part of the preconceptional evaluation; normally this process involves changing the patient to medications that may be better tolerated and safer during pregnancy. Indeed, medications to improve cardiac status may be necessary for a successful pregnancy outcome. Certain of these have been classified as contraindicated during pregnancy and have received a Food and Drug Administration (FDA) classification of 'X'. The common cardiac medications and their potential problems for pregnancy are summarized in Table 3.

Unfortunately, research on medication during pregnancy is limited; even the FDA classification is based on isolated case reports and animal studies. Medication risks must always be outweighed by their potential benefits, and,

Table 3 Cardiac medication during pregnancy

Medication	FDA class	Risks in pregnancy	Recommendations
Warfarin	X	Teratogenic; bleeding	Avoid in first trimester and prior to delivery; dose <5 mg
Aspirin	C/D	Bleeding; early ductal closure	Low dose if possible
Digoxin	C		Continue
Diuretics	B/C/D	Hypovolemia; electrolyte disorder	Use only for congestive heart failure or pulmonary edema
Atenolol	D	Hypotension	Change to metoprolol
Metoprolol	C	Hypotension	Continue if indicated
Calcium channel blockers	C	Hypertension; uterine effects	Continue if indicated
Hydralazine	C	Hypotension; fetal thrombocytopenia	May be continued if necessary
ACE inhibitors	D	Fetal abnormalities	Discontinue; change to hydralazine
Amiodarone	D	Fetal thyroid disorder	Continue if necessary

ACE, angiotensin converting enzyme

in some populations as discussed below, women definitely need to continue to take their medications.

CONGENITAL HEART DISEASE

The population of women of childbearing age who were born with heart disease is increasing because of the advances in cardiac surgery, diagnosis and intervention during childhood. Each type of problem, whether it be a shunt, valvular or complex congenital heart lesion, needs to be examined uniquely with regard to potential risks of pregnancy.

Shunt lesions

Atrial septal defect

In an adult an unrepaired atrial septal defect (ASD) carries a theoretic risk of paradoxical embolus. This is of particular concern during pregnancy, because as the pregnancy progresses a woman becomes more hypercoaguable. Although earlier literature suggested such individuals be prophylacticly anticoagulated, this is not warranted in an ambulatory patient. The potential risk of arrhythmia, particularly supraventricular arrhythmia, is also present. If the defect is particularly large, bidirectional shunting may be present. In the setting of chronic right to left shunting, increases in the pulmonary pressure may result in Eisenmenger's syndrome, which results from progressive increases in pulmonary vascular flow causing changes within the pulmonary vasculature. The shunt eventually reverses from right to left, and the patient becomes cyanotic. In this setting, pregnancy is contra-indicated because the patient cannot tolerate the hemodynamic changes of pregnancy. Also, cyanotic mothers have a high risk of having small for gestational age infants, which increases their risks. Echocardiography with Doppler may quantify the pulmonary artery pressure. If there is a concern regarding hypertension, a right-heart catheterization may be indicated for a definitive diagnosis. The overall

risk to the offspring is estimated at 8–10% if a maternal ASD is present[13]. Closure of the septal defect prior to conception may improve outcome, although it is best to wait 6 months after surgery or device placement for endothelization of the interatrial septum to occur before pregnancy is attempted.

Women with a repaired ASD may exhibit residual shunting which then would confer the risk of paradoxical embolus. The repair may consist of a surgically placed patch or catheter-based device, and the risk of arrhythmias may persist. Symptoms suggestive of arrhythmias should be investigated prior to pregnancy. Women born with a primum ASD may have also had a cleft mitral valve. Despite repair of the mitral valve in childhood, significant mitral regurgitation may be present in the adult. Although unrepaired, there is still an increased risk of congenital heart disease in the fetus. In one report women with shunt lesions had the highest prevalence of obstetric and cardiac complications[1].

Ventricular septal defect

An unrepaired ventricular septal defect (VSD) confers a risk of endocarditis, pulmonary hypertension and aortic valve insufficiency. Usually there is a high pressure left to right shunting, and endocarditis may occur. A high velocity left to right shunt over time increases pulmonary arterial flow causing permanent vascular changes within the lung and, subsequently, pulmonary artery pressure increase. This may also result in an Eisenmenger's syndrome (see above). Due to the severity of the shunt and proximity to the aortic valve of a membranous VSD, there may be progressive aortic insufficiency. Progressive aortic insufficiency, increasing pulmonary pressures or a history of endocarditis are indications to close a VSD. Echocardiography with Doppler should help identify complications of a VSD in the preconceptional evaluation. It is important to remember in a woman with the Eisenmenger's physiology syndrome that her risks

persist after delivery when she is undergoing fluid shifts; they are also present with termination of pregnancy when there may also be fluid shifts and a drop in systemic pressure due to the effects of anesthesia. Such a decline in systemic pressure in a woman with Eisenmenger's physiology who already has a right to left shunt may worsen the right and left shunting thereby worsening the cyanosis.

After repair of a VSD, a residual shunt may put the patient at risk for endocarditis. A bundle branch block may be seen on the electrocardiogram, and, depending on the age of the repair, persistent elevation of pulmonary artery pressure may be noted by Doppler echocardiography or further defined by right heart catheterization.

Patent ductus arteriosus

An unrepaired patent ductus arteriosus (PDA) presents a theoretic risk of endarteritis and pulmonary hypertension. This risk is related to the persistence of the connection between the pulmonary artery and the aorta. In adult life the shunt would be from left to right, and, therefore, there would be increasing flow within the pulmonary vasculature. Theoretically there may also be associated Eisenmenger's physiology.

This lesion can be repaired during childhood when the PDA is ligated; pregnancy is well tolerated in patients who have had a repair. However, their offspring still carry a risk of congenital heart disease. The patient may also have had a closure using a coil in a catheter-based intervention; theoretically pregnancy should be tolerated well as long as there is no residual pulmonary hypertension.

Complex congenital heart disease

Coarctation of the aorta

The adult form of coarctation of the aorta represents a narrowing of the aorta distal to the

left subclavian artery. This condition often is diagnosed in childhood but may only present in adulthood. If this is the case, mild coarctation of the aorta is present, and patients develop an extensive network of collateral flow to supply their lower body. Associated bicuspid aortic valve with aortic valve pathology or progressive ascending aorta dilatation may also be present and dissection is possible at the site of coarctation[13].

A coarctation is either repaired or bypassed during childhood, or a stent may have been placed within the aorta. The women should be assessed prior to pregnancy because restenosis may have occurred. If so, stenting may be considered prior to pregnancy. After the stent is placed, pregnancy should be delayed approximately 6 months. Even in the absence of restenosis, the adult may experience persistent hypertension particularly with exercise. Medications for hypertension may need to be altered in anticipation of pregnancy, and the patient should be monitored closely for elevations in blood pressure and aortic root enlargement during pregnancy.

Tetralogy of Fallot

Women of childbearing age with an unrepaired tetralogy of Fallot are cyanotic and pregnancy is not advised. Most women of this age will have already undergone a repair and preconceptional evaluation to look for residual abnormal hemodynamics, pulmonary valve disease, ventricular dysfunction and arrhythmia or heart block[14,15]. Risk of sudden death is present in this population. Severe pulmonary insufficiency after repair has been associated with right ventricular enlargement or dysfunction during pregnancy[2,15]. There also may be a risk of sudden death if the QRS is particularly prolonged (over 160 m/s). The overall risk of fetal loss and congenital heart disease is increased, and other associated problems may be present, for example a 22q11 deletion, which is associated

with the DiGeorge's syndrome. Such patients may have tetralogy of Fallot or interrupted aortic arch.

Ebstein's anomaly

Women with mild forms of the Ebstein's anomaly may not have required repair prior to reaching the childbearing years but should nonetheless undergo complete evaluation. Assessment should focus on the extent of tricuspid regurgitation, the presence of cyanosis and existence of residual right ventricular dysfunction. In Ebstein's anomaly the tricuspid valve is set low into the right ventricle, so that the right ventricle becomes atrialized. Severe forms are diagnosed during childhood and repaired. After repair it is important to assess the patient for residual hemodynamic abnormalities such as tricuspid insufficiency and residual cyanosis. The tricuspid insufficiency may increase and be accompanied by right ventricular failure during pregnancy. Heart block and arrhythmias also may occur. The association of Wolff-Parkinson-White syndrome (accessory bypass tract) confers the risk of atrial arrhythmias in these patients. Fetal risks have been described, including low birth weight and fetal loss, both of which seem to be associated with maternal cyanosis. The risk of congenital heart disease in the fetus has been estimated at 6%[13].

Single ventricle

Women born with a variant single ventricle variant may have undergone a variety of surgical interventions. Single ventricle variants include tricuspid artresia or double outlet right ventricle. Such individuals may have had a Glenn procedure, in which the superior vena cava flow is directed to the pulmonary artery, and a Fontan procedure, in which in the inferior vena cava flow is directed to the

pulmonary artery either outside the heart or incorporating some of the right atrium. Recent approaches have placed external conduits to the pulmonary artery, but variations of the Fontan repair are reported. All women who have had a Fontan procedure should have preconceptional evaluation[13]. There have been many case reports of successful pregnancies; however, there are multiple variations of the Fontan conduit[15–17]. Revision of the Fontan may be recommended prior to conception. Anticoagulation should be continued. Maternal risks include atrial arrhythmias and heart block. Women who have underlying ventricular dysfunction may not be able to sustain a pregnancy. They may exhibit edema and ascites if there is failure of the Fontan circuit, and both should be identified prior to pregnancy. Fetal risks include spontaneous abortion, premature birth and embryopathy, because these patients are often continued on warfarin, in addition to other medications[15–17].

D-transposition of the great vessels

In women born with d-transposition of the great vessels, the aorta arises from the right ventricle and the pulmonary artery from the left ventricle. These women will have undergone some type of repair prior to reaching childbearing age and require full evaluation including a clinical, functional and echocardiographic evaluation to look specifically for atrioventricular (AV) valve regurgitation, atrial arrhythmia and, importantly, systemic ventricular function[18]. After an atrial switch procedure (Mustard/Senning), the right ventricle serves as the systemic ventricle and over time may dilate in response to the volume load[19]. There may be an overall decrease in systemic ventricular ejection fraction. An atrial switch (Mustard/Senning procedure) diverts the incoming flow to the appropriate ventricles: the incoming deoxygenated vena caval flow is directed by a baffle to the mitral valve of the

left ventricle and thence to the pulmonary artery. At the same time, the pulmonary arterial flow is directed by another baffle to the tricuspid valve, right ventricle and aorta. Women who have had the atrial switch procedure need to be assessed for right ventricular size and function. The right ventricle is a systemic ventricle and needs to sustain the increase volume load of pregnancy. The patient may have had a Rastelli procedure in which there is a left ventricle to aorta baffle with closure of the VSD. Such individuals need to be assessed for left and right ventricular obstruction; in addition their systemic ventricular function should be assessed, although they do have a left ventricle for their systemic flow. Pregnancies after atrial switch have been described[18,19]. Reported risks include systemic ventricular failure and arrhythmias. Obstetric complications include premature delivery, growth retardation and thromboembolic complications[19].

The more up-to-date repair is an arterial switch (Jatene procedure), in which the great vessels are actually switched and the coronary arteries reimplanted. Case reports of pregnancy after the arterial switch procedure are available. Because the coronary arteries are reimplanted, the arteries and the aorta need to be assessed. The same may be said for functional capacity prior to pregnancy.

L-transposition of the great vessels

L-transposition of the great vessels may not be diagnosed until adulthood due to the associated pulmonic stenosis, VSD or congenital heart block. The right ventricle is on the left side and connected to the aorta. Having to serve as the systemic ventricle, it may fail over time. Preconceptional evaluation should assess systemic ventricular function, and the extent of AV valvular regurgitation which may worsen as pregnancy progresses[20]. Echocardiography is essential to assess anatomy and AV valve function. A pacemaker may have been placed

and its status requires substantiation. Magnetic resonance imaging (MRI) may be useful to evaluate anatomy. With stress testing aerobic capacity is paramount, and pregnancy is not well tolerated if aerobic capacity is less than 75% of that predicted. In a study of 60 pregnancies in 22 women, there was a 16% miscarriage rate, but no congenital heart disease was identified in the offspring. The maternal risks included congestive heart failure which was related to systemic AV valve regurgitation[20].

Valvular lesions

Women of childbearing age may have been born with valve disease or acquired it after a bout of rheumatic fever[21]. Rheumatic heart disease is less common today in the United States and Europe, but remains a problem for women of childbearing age in the Philippines, India and the Middle East. Despite its etiology, valvular heart disease may be complicated by the expected hemodynamic changes of pregnancy with increased preload and decreased afterload[22]. Endocarditis may recur during pregnancy resulting in further valve deterioration. Most patients with valvular heart disease should undergo preconceptional evaluation which may include stress testing to detect dynamic changes in valve function after exercise.

The aortic valve

The woman born with a bicuspid aortic valve does not have significant hemodynamic changes within the valve or may have a combination of aortic stenosis and aortic insufficiency. When severe aortic stenosis is present, pregnancy should be discouraged if the aortic valve area is less than $1.0\,cm^2$. The concern is that the volume load of pregnancy may be difficult to tolerate and increase the incidence of congestive heart failure during pregnancy[23,24]. Pregnancy should be avoided and valve repair

considered if the woman develops dyspnea, syncope or angina. It may be best to proceed with valvular repair in anticipation of pregnancy.

Aortic insufficiency

Aortic insufficiency is often well tolerated in the adult patient. It is important, however, to assess the degree of ventricular dilatation; if the ventricle is quite dilated and/or the patient is symptomatic, intervention prior to pregnancy is advisable. It is interesting to note that valve regurgitation may actually decrease during the course of a pregnancy, because of the natural afterload reduction that occurs. If ventricular function and dimensions are normal, on the other hand, pregnancy is usually well tolerated[22].

A bicuspid aortic valve, may be associated with the aortic enlargement which needs to be assessed prior to and during pregnancy. If enlargement is present, beta blockade may protect the aorta from enlargement as seen in the setting of Marfan syndrome. During pregnancy changes may occur within the media of the vessel wall that promote aortic enlargement[25]. Pregnancy should be discouraged in the woman with Marfan syndrome who has an aortic root dimension greater than 4 cm. If a woman with aortic dilatation becomes pregnant she should be maintained on beta blockers. The aortic root dimension should be monitored by echocardiography during the course of the pregnancy because of a theoretic risk of aortic enlargement. In addition, patients may not be able to tolerate the prolonged Valsalva maneuver which may occur during the pushing of the second stage of labor, as there may be strain on the aortic wall.

Supraventricular aortic stenosis

Supraventricular aortic stenosis is often associated with William's syndrome and may be

identified if there is a family history of hypertension, coronary artery disease or stroke. Pregnancy should be discouraged if significant obstructive coronary involvement or aortic disease is present[13].

Pulmonic valve

The pulmonary valve is often involved in complex congenital heart disease. The patient may also present with isolated pulmonic valve stenosis or insufficiency. Pregnancy is usually well tolerated if pulmonic valve stenosis is mild to moderate. If severe, on the other hand, a valvotomy should be considered prior to pregnancy[22]. This catheter-based procedure relieves significant gradients across the valve and thereby makes the patient a better candidate for pregnancy. It must to be remembered, however, that even with right ventricular outflow obstruction the right ventricle is going to increase in volume during the pregnancy; this may not be well tolerated and theoretically could result in tricuspid regurgitation.

Pulmonic insufficiency

Pulmonic insufficiency is an isolated finding; it is often well tolerated but becomes more complicated if it appears following complex congenital heart surgery such as that for tetralogy of Fallot. The outcome of pregnancy may be determined by right ventricular size and function. Right ventricular function may be identified by stress testing to assess right ventricular reserve with exercise. With severe pulmonary insufficiency, the right ventricle is impaired either at rest and/or with exercise, so much so that there may be an indication to replace the valve with a homograft to permit a less potentially complicated pregnancy.

Mitral valve

Mitral valve stenosis is usually rheumatic in origin. The major risks of mitral stenosis include pulmonary edema, atrial thrombus and embolic atrial fibrillation. In the presence of atrial fibrillation, congestive heart failure may result with the loss of atrial 'kick'. This circumstance may be accentuated during pregnancy or if the patient is already volume overloaded. It is therefore important to identify mitral stenosis prior to pregnancy. As a woman with mitral stenosis develops atrial fibrillation or becomes hypercoaguable, she may develop thrombi in the left atrium and be at risk for thromboembolic complications[21,26]. This patient needs to be followed closely during pregnancy for changes in rhythm and/or fluid overload. Valve repair may be indicated prior to pregnancy. Percutaneous balloon valvotomy has been used during pregnancy when congestive heart failure develops. Prior valve replacement with a mechanical prosthesis creates potential problems with regard to anticoagulation. Several strategies are available for heparin and warfarin anticoagulation[27–30].

Mitral insufficiency

This problem is usually well tolerated during pregnancy; with the natural afterload that occurs with vasodilatation, it may actually decrease. Mitral insufficiency may be seen after ASD repair, because primum ASD may be associated with a cleft mitral valve. In view of the fact that a cleft mitral valve may have been repaired during childhood but, as an adult, residual mitral regurgitation may be present, it is important to look for this prior to pregnancy. If mitral regurgitation is severe, the patient may benefit from valve repair prior to pregnancy.

Tricuspid valve

Tricuspid stenosis is most often identified during childhood, and patients undergo repair (discussed below). Tricuspid valve insufficiency is often well tolerated during

pregnancy, although it may increase with the hemodynamic changes of increased volume.

AORTIC DISEASE

Marfan syndrome

Marfan syndrome is characterized by cardiac involvement, including progressive aortic dilatation and mitral valve regurgitation. In women with aortic dilatation, aortic dissection and rupture may occur in pregnancy, particularly if the aorta is 40 mm or greater in diameter[31–36]. Aortic dissection/rupture can occur at anytime during the third trimester when there also may be a worsening of mitral regurgitation. The autosomal dominance of Marfan syndrome itself presents a risk to the fetus. The patient needs to be followed during pregnancy with echocardiography to assess the valves and aortic root. Beta blockade should be continued, but only metoprolol is recommended during pregnancy (Table 3). Women on losartan should be changed to a beta blocker prior to conception.

ARRHYTHMIAS

Women of childbearing age may have a history of a prior arrhythmia[37,38]. This may occur in the absence of structural heart disease but is common in women with congenital heart disease[39]. Recurrence risks in pregnancy vary greatly depending on the arrhythmia. Recurrence risks for supraventricular tachycardias have been estimated at 50%, atrial fibrillation or flutter 52% and ventricular tachycardia 27%[37]. Adverse fetal events have been associated with antipartum arrhythmias.

Superventricular tachycardia may occur in the absence of structural heart disease, but in one study 53% of patients had underlying heart disease[37,40]. Tachycardia may be atrial or superventricular, and patients may already

be on medications during pregnancy. If the patient has been relatively asymptomatic, a trial off mediation could be implemented during the first trimester; however, if the superventricular episodes have been at rather fast rates, ablation prior to pregnancy should be a consideration. It is possible that the superventricular tachycardia could recur with pregnancy with rapid rates and require additional medical therapy which could be problematic[11,12,41]. Wolff-Parkinson-White syndrome is associated with superventricular tachycardia, and it has been recommended that those patients should either continue their medications or consider ablation prior to pregnancy[38]. Atrial fibrillation and flutter is usually associated with underlying structural heart disease. Patients may require anticoagulation that needs to be continued at low dose throughout the pregnancy. During the first trimester patients should be changed to heparin. In the setting of atrial fibrillation during pregnancy, 96% of patients present with underlying structural heart disease[37].

Ventricular arrhythmias may also have occurred prior to pregnancy. This may occur in the setting of a structurally normal heart; although in one study in women with pre-existing ventricular tachycardia, 55% had underlying structural heart disease[37,42]. This condition may require continued therapy throughout the pregnancy. If an automatic implantable defibrillator is present, antiarrhythmic medication is necessary to prevent the device from firing[11,12,41]. In a recent study treatment of prolonged QT syndrome with beta blockade was found to decrease cardiac events (ventricular arrhythmias) in the postpartum period for up to 9 months[43].

MYOCARDIAL DISEASE

Women of childbearing age may have acquired myocardial disease. Depending upon the type of myocardial disease, pregnancy may be

contraindicated. For example, if systolic dysfunction is present and the ejection fraction is less than 40%, pregnancy is contraindicated[44]. All women who have underlying myocardial dysfunction require a full evaluation prior to pregnancy. This may include stress testing to assess functional ventricular reserve. Evaluation may be performed off medication, as the medication would optimally be best withheld in the first trimester in pregnancy. Medication could then be resumed later in pregnancy, but certain medications often used to treat women with myocardial disease may be contraindicated during pregnancy.

CORONARY ARTERY DISEASE

Atherosclerotic coronary artery disease is rare in women of childbearing age, except perhaps for female diabetics or those with familial hypercholesterolemia. Little current information is available regarding the preconceptional evaluation of women with coronary disease. Regardless, it is important to rule out active or potentially active coronary lesions and to ensure that the woman has normal ventricular function both at rest and with exercise[45]. It is also important to document the absence of ischemia during exercise. With these conditions met, pregnancy can take place with very careful monitoring throughout. For women who have had prior stenting, aspirin may be continued during pregnancy, although there is a risk of early ductal closure. Plavix has not been widely used during pregnancy and may be problematic.

DILATED CARDIOMYOPATHY

A woman of childbearing age may have a dilated cardiomyopathy due to a prior viral illness or to a prior pregnancy. In the setting of a dilated cardiomyopathy with reduced ejection fraction, pregnancy is contraindicated. However, it becomes more controversial if a woman has

recovered from pregnancy followed by a peripartum cardiomyopathy[44]. If her ejection fraction remains reduced, she has a risk of clinical deterioration and death[44]. It is important to observe cardiac reserve, preferably off medication, with stress echocardiography. If a patient has not recovered from a peripartum cardiomyopathy, she remains at risk for recurrence and further deterioration[46]. Women who have received anthrocyclin therapy and/or radiation to the chest for childhood malignancies may have a cardiomyopathy as an adult. In a woman with this history, it is important to evaluate ventricular function and reserve very carefully prior to pregnancy using stress echocardiography. There may also be an underlying radiation induced vasculitis. There have been case reports of pregnancy after cardiac transplantation, at times occurring after peripartum cardiomyopathy. In addition to ventricular function, immunosuppressive medications and their potentially harmful effects on the fetus need to be considered[47,48].

HYPERTROPHIC CARDIOMYOPATHY

It is rare that a woman with hypertrophic cardiomyopathy develops heart failure with pregnancy. Symptoms occurring during pregnancy usually occur in women who have experienced symptoms prior to pregnancy, and pregnancy is often well tolerated[49]. In one study there were no increased arrhythmia rates in pregnancy compared to a non-pregnant control with hypertrophic cardiomyopathy. On the other hand, congestive heart failure has been described in women with hypertrophic cardiomyopathy, particularly in women who have a family history of hypertrophic cardiomyopathy[50].

RISKS TO FETUS

Risks to the fetus include prematurity, low birth weight and an increased risk of congenital

heart disease. The overall risk of congenital heart disease probably ranges between 4 and 10%[10]. The risk of congenital heart disease is higher if there is maternal or paternal outflow tract lesions such as aortic stenosis or coarctation of the aorta.

The preconceptional evaluation should begin with a very careful history to identify current symptoms or problems which could denote cardiac dysfunction. A complete history of prior surgeries must be obtained and the sequelae of such surgeries documented in detail. Next, a complete medication history is required to identify medications that need to be changed in anticipation of conception or to be withheld for the first 13 weeks when organ development is occurring. The issue of coumadin therapy and risk of embryopathy in the first trimester is complex and described elsewhere.

Family history is important to identify members of the family with congenital heart disease or a history of sudden death. It is important to understand the patient's level of activity in the course of her normal day. The ability to exercise and the type of exercise performed should be documented to provide an overall understanding of the functional capacity and how pregnancy may be tolerated. Medical problems that would affect the patient's cardiac status during pregnancy should be identified and treated, including as anemia, thyroid disease, smoking, alcohol abuse and illicit drug use[4]. Advanced maternal age may play a role with respect to coronary artery and cardiomyopathy risk.

Appropriate diagnostic testing is necessary as part of the preconceptional evaluation, and recommendations are summarized in Table 4. Electrocardiographic testing includes a 12-lead electrocardiogram or arrhythmia monitoring with a Holter monitor for brief periods of time or an event monitor. Echocardiography is important to identify the structure of the heart, valvular disease and shunt lesions, which can be further identified by contrast bubble studies. Echocardiography combined with stress testing is useful to assess functional reserve. Cardiopulmonary testing may be performed to assess aerobic threshold. Cardiac catheterization may be indicated to better define pulmonary pressures prior to pregnancy

Table 4 The preconceptional evaluation for common cardiac lesions

Lesion	Exam	CXR	ECG	Echo	MRI	Stress test	Holter
ASD	✓			✓			
VSD	✓		✓	✓			
Aortic stenosis	✓		✓	✓		✓	
Coarctation of aorta	✓		✓	✓	✓		
Tetralogy of Fallot	✓	✓	✓	✓		✓	✓
TGA – arterial switch	✓	✓	✓	✓		✓	✓
TGA – atrial switch	✓	✓	✓	✓	✓	✓	✓
Fontan	✓	✓	✓	✓	✓	✓	✓
Ebstein's anomaly	✓	✓	✓	✓		✓	✓
Dilated cardiomyopathy	✓	✓	✓	✓		✓	✓
Hypertrophic cardiomyopathy	✓		✓	✓	✓	✓	✓

CXR, chest X ray; ECG, electrocardiogram; Echo, echocardiogram; MRI, magnetic resonance imaging; ASD, atrial septal defect; VSD, ventricular septal defect; TGA, transposition of great arteries

and to identify a woman who may benefit from a catheter-based intervention.

Counseling the patient and her family

A total risk assessment needs to be formulated, depending on the nature of the cardiac lesion. This risk assessment may include a complete preconceptional evaluation to look for potential problems that may require attention prior to becoming pregnant. Medications may need to be altered in anticipation of pregnancy and interventions may need to be performed.

The potential risks of pregnancy, labor and delivery should be discussed with the patient and her family in the preconceptional evaluation. The frequency and monitoring of the tests that will be used to follow the patient should also be discussed. For example, that the patient may require hemodynamic monitoring for 24–28 hours after delivery in an intensive care setting to assess volume shifts should be discussed. The patient should understand that she may be physically separated from her child during that time. Other potential considerations at the time of labor and delivery can also be discussed, but not all complications can be fully anticipated.

In higher risk women, subsequent pregnancies also require full evaluation. It cannot be assumed that because a woman has had a successful pregnancy that her next will be equally uncomplicated. Full and complete evaluation must be undertaken prior to every pregnancy; and this discussion also should include the instruction to prevent pregnancy during the time of evaluation and intervention.

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