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# Cardiovascular problems and pregnancy: An approach to management

## ABSTRACT

Women with some congenital or acquired heart lesions are at increased risk for a number of maternal and neonatal complications during pregnancy. Knowing what constitutes high risk and when to refer to a specialty clinic are key to successfully managing such patients.

## KEY POINTS

Cardiac output increases 50% in pregnancy while peripheral vascular resistance and blood pressure decrease. These changes can exacerbate certain pre-existing cardiac conditions.

Pregnant women with congenital heart lesions are at risk for heart failure, arrhythmia, stroke, neonatal complications, and even death in some conditions.

Women of reproductive age with congenital heart lesions should be counseled as to whether pregnancy is advisable and whether their problem can be corrected or palliated before pregnancy.

Women at intermediate or high risk for maternal or fetal complications should be referred for specialty care.

**I**S IT SAFE for a woman with a heart problem to have a baby? What should we advise her and how should her pregnancy be managed?

The answers depend on the problem and on the woman's heart status. With some types of heart disease, outcomes are excellent without any special management in women with good function. With other types, pregnancy poses a reasonable risk if the problem is corrected first, and still other types should rule out pregnancy altogether.

This article reviews the impact of pregnancy on a number of congenital and acquired heart diseases (and vice versa) and offers recommendations for their management. It also provides a method to assess risk to help in deciding whether to refer a patient for specialty care.

## CARDIOVASCULAR CHANGES DURING PREGNANCY

During pregnancy, intravascular volume and cardiac output increase by 50%, peaking during the second trimester and remaining high through the rest of pregnancy. This "physiologic" high-output state is accompanied by decreases in peripheral vascular resistance and blood pressure. The hemodynamic changes of pregnancy may not fully resolve until 6 months after delivery.

Owing to these normal changes, many healthy pregnant women have symptoms mimicking those of cardiac disease, including fatigue, dyspnea, and light-headedness, and a number of "abnormal" findings on physical examination, electrocardiography, and echocardiography (TABLE 1).

Acknowledgment: The authors' work is supported in part by an operating grant from the Canadian Institutes for Health Research.

TABLE 1

**Common cardiac findings in normal pregnancy****Symptoms**

Fatigue  
Dyspnea  
Light-headedness

**Physical findings**

Displaced apical impulse  
Prominent jugular venous pulsations  
Widely split first and second heart sounds  
Soft ejection systolic murmur

**Electrocardiographic findings**

Sinus tachycardia  
Premature atrial or ventricular ectopic beats  
Right or left axis deviation  
ST-segment depression  
T-wave changes

**Echocardiographic findings**

Mild increase in left ventricular diastolic dimension with preservation of ejection fraction  
Functional tricuspid and mitral regurgitation  
Small pericardial effusion

## ■ OUTCOMES VARY WITH DIFFERENT CARDIAC LESIONS

Few women with heart disease actually die during pregnancy; the high-risk exceptions are women with Eisenmenger syndrome, pulmonary vascular obstructive disease, or Marfan syndrome with aortopathy. However, pregnant women with heart disease are at risk for other complications such as heart failure, arrhythmias, and stroke.<sup>1-6</sup> Their babies are also at risk of complications such as birth weight that is low for the gestational age, premature birth, and death.<sup>1,2,7</sup>

Congenital heart disease is now the most common heart problem in pregnant women seen at referral centers in North America.<sup>5</sup> Peripartum cardiomyopathy is infrequent. Isolated mitral valve prolapse is probably the most common cardiac lesion in pregnant women, but it has an excellent prognosis in pregnancy, and patients with it may not need to be referred to a cardiovascular specialist.<sup>8</sup>

## ■ CONGENITAL HEART LESIONS

**Left-to-right cardiac shunts**

In patients with an atrial septal defect, ventricular septal defect, or patent ductus arterio-

sus, blood can shunt from the high-pressure left side of the heart to the lower-pressure right side. During pregnancy, as cardiac output increases, one would expect this left-to-right shunting to increase. However, this effect may be attenuated by the decrease in peripheral vascular resistance.

If there is no pulmonary hypertension, then pregnancy, labor, and delivery are well tolerated.<sup>2,4,5,9</sup> However, during labor and delivery, a risk of paradoxical embolism exists (ie, a venous thromboembolism passing from the right side of the heart to the left), particularly with an atrial shunt such as a patent foramen ovale.

**Aortic stenosis, left ventricular outflow tract obstruction**

The most common cause of aortic stenosis in pregnant women is a congenital bicuspid aortic valve, but fixed subvalvular and supra-valvular aortic stenoses have similar hemodynamic implications.

If the stenosis is severe, the heart must strain to increase its output during pregnancy, and heart failure or ischemia may develop. The left ventricle can become hypertrophied and noncompliant, and in this condition, any condition that decreases preload—such

**In Marfan syndrome, aortic root replacement does not eliminate the risk of dissection**



as compression of the inferior vena cava in late pregnancy, anesthetic agents with vasodilatory effects, peripartum blood loss, and bearing-down maneuvers—can lead to an exaggerated drop in cardiac output and to hypotension.

In a 1993 overview of 106 pregnancies in women with congenital aortic stenosis, the maternal mortality rate was 11% and the perinatal mortality rate was 4%.<sup>10</sup> However, in a more recent series of 49 pregnancies (59% in women with severe stenosis), no women died.<sup>11</sup> Adverse maternal cardiac events occurred in 3 women (6%), all of whom had severe stenosis, defined as an aortic valve area of 1 cm<sup>2</sup> or smaller or a transvalvular pressure gradient of 64 mm Hg or greater.

Aortic dissection has been reported in pregnant women with a bicuspid aortic valve and ascending aortopathy,<sup>12</sup> although this risk is probably lower than for women with Marfan syndrome with aortopathy.

**Recommendations.** Women with symptomatic aortic stenosis should delay getting pregnant until the stenosis is surgically corrected.<sup>13</sup> However, absence of symptoms does not guarantee that pregnancy will be well tolerated. Balloon valvuloplasty during labor and delivery may be palliative in certain cases.

### Coarctation of the aorta

Coarctation of the aorta is commonly associated with a bicuspid aortic valve; other associations include aneurysms of the circle of Willis, ventricular septal defects, and Turner syndrome.

The coarctation often is corrected before a woman becomes pregnant; if it is not, aortic rupture is a risk in the third trimester and during labor. In early series of uncorrected cases, the maternal mortality rate was 3% to 4%, or higher if there were associated cardiac defects, aortopathy, or long-standing hypertension.

Even if the coarctation is corrected before pregnancy, pregnancy-induced hypertension can occur,<sup>4,5,14</sup> probably due to residual abnormalities in aortic compliance.

Maternal death is rare. Recent studies in patients with both corrected and uncorrected aortas have been encouraging, with only one maternal death reported in 182 pregnancies.<sup>14</sup>

### Pulmonary valve stenosis

Pulmonary valve stenosis can be classified by echocardiographic estimates of the peak pressure gradient across the valve:

- Mild (< 50 mm Hg)
- Moderate (50–79 mm Hg)
- Severe ( $\geq$  80 mm Hg).

The gradient increases with cardiac output during pregnancy, so the severity may be overestimated if no antenatal study is available.

Pulmonary valve stenosis that is mild or that has been treated by valvuloplasty or surgery is well tolerated during pregnancy.<sup>4,5</sup> Fetal outcome is also favorable.<sup>4,5</sup>

Severe stenosis, even if asymptomatic before pregnancy, may lead to right-sided heart failure or atrial arrhythmias owing to the increased hemodynamic load of pregnancy.

**Recommendations.** Patients with severe pulmonary valve stenosis should be considered for correction before pregnancy. Balloon valvuloplasty may be feasible during pregnancy if symptoms progress.

### Cyanotic heart disease: Unrepaired and repaired

The most common form of cyanotic congenital heart disease is the tetralogy of Fallot, the essential features of which are right ventricular outflow tract obstruction and a large, non-restrictive ventricular septal defect.

If the problem is not corrected or palliated, the pregnancy-associated fall in systemic vascular resistance and rise in cardiac output exacerbate right-to-left shunting, leading to increased maternal hypoxemia and cyanosis. The fetal loss rate may be as high as 30%, and the maternal mortality rate is 4% to 15%.<sup>15</sup>

In a series of 96 pregnancies in 44 women with a variety of cyanotic congenital heart defects, there were high rates of maternal cardiac events (32%, including 1 death) and prematurity (37%), and a low live birth rate (43%).<sup>16</sup> The lowest live birth rate (12%) was in mothers with arterial oxygen saturation of 85% or lower.

Risk is low in women in whom the tetralogy has been successfully corrected.<sup>2,4,5</sup>

### Marfan syndrome

Marfan syndrome is a connective tissue disorder inherited in an autosomal-dominant pat-

**Women with Eisenmenger syndrome should be offered sterilization or pregnancy termination**

tern. Life-threatening aortic complications are due to medial aortopathy, resulting in dilatation, dissection, and valvular regurgitation.

The aortopathy is a generalized process. Therefore, in patients with aortic root dilatation, prophylactically replacing the root before pregnancy may not fully eliminate the risk of dissection of the residual native aorta.

Risk is increased in pregnancy, owing to hemodynamic stress and perhaps hormonal effects. Although the mortality rate was very high (around 30%) in older case reports, more recent data suggest an overall maternal mortality rate of 1% and a fetal mortality rate of 22%.<sup>17</sup>

In a prospective study of 45 pregnancies in 21 patients, most patients had no obstetric complications or significant change in aortic root size. However, in 8 patients with a dilated aortic root (> 40 mm) or prior aortic root surgery, 3 of 9 pregnancies were complicated by either aortic dissection or rapid aortic dilatation.<sup>18</sup>

**Recommendations.** Patients with aortic root involvement should receive preconception counseling emphasizing their risk. If the patient is seen in early pregnancy, she should be offered the option to terminate the pregnancy.

In contrast, women with little cardiovascular involvement and an aortic root diameter smaller than 40 mm by echocardiography tolerate pregnancy well. Serial echocardiography should be done to monitor for progressive aortic root dilatation, and beta-blockers should be given prophylactically.<sup>19</sup> The possibility of dissection even with a normal aortic root should be acknowledged to the patient.

### **Eisenmenger syndrome and pulmonary vascular obstructive disease**

Eisenmenger syndrome involves pulmonary vascular obstructive disease resulting from a pre-existing left-to-right shunt. Over time, pulmonary pressures rise to systemic levels, changing the shunt flow to right-to-left.

Most complications during pregnancy occur at term and during the first postpartum week. Spontaneous abortion, intrauterine growth restriction, and preterm labor are frequent. Perinatal mortality is due mainly to prematurity.

Maternal and neonatal mortality rates are high in patients with pulmonary hypertension. A 1998 review of 125 pregnancies found the maternal mortality rate to be 30% in those with primary pulmonary hypertension, 36% in those with Eisenmenger syndrome, and 56% in those with secondary vascular pulmonary hypertension. The overall neonatal mortality rate was 12%.<sup>20</sup>

**Recommendations.** Preconception counseling should stress the extreme risks from pregnancy. Patients with Eisenmenger syndrome should always be offered sterilization or pregnancy termination.

## **■ RHEUMATIC HEART DISEASE**

### **Mitral stenosis**

Mitral stenosis is the most common rheumatic valvular lesion of pregnancy. The hypervolemia and tachycardia associated with pregnancy exacerbate the transmitral gradient.

Atrial fibrillation may result from the elevated left atrial pressure. This can precipitate heart failure, primarily due to an uncontrolled ventricular rate; equivalent tachycardia from any cause may be equally detrimental. Even patients with only mild to moderate mitral stenosis (who have no symptoms before pregnancy) may develop atrial fibrillation and heart failure during the antepartum and peripartum periods.

Recent studies found no mortality but substantial morbidity from heart failure and arrhythmia.<sup>4,5,21,22</sup> The risk of complications is higher in women with a history of cardiac events (arrhythmias, stroke, or pulmonary edema) and with moderate or severe mitral stenosis.

The risk of adverse fetal or neonatal outcomes also increases with increasing severity of mitral stenosis.<sup>22</sup>

Percutaneous mitral valvuloplasty during pregnancy should be considered in patients who, despite optimal medical therapy, are in New York Heart Association (NYHA) functional class III or IV (markedly limited ability or inability to engage in any physical activity without symptoms).<sup>23</sup>

### **Other rheumatic lesions**

**Rheumatic aortic stenosis** poses a risk during pregnancy similar to that of congenital

**Maternal cardiac status is an important predictor of pregnancy outcome**



aortic stenosis.

**Aortic or mitral regurgitation**, even if severe, is generally well tolerated during pregnancy, although function may deteriorate as measured by the NYHA classification.

## ■ PERIPARTUM CARDIOMYOPATHY

Peripartum cardiomyopathy, an idiopathic dilated cardiomyopathy, involves ventricular systolic dysfunction that develops during the last month of pregnancy or in the first 5 months after delivery in patients with no known underlying disease.<sup>24</sup>

Heart failure is the most common manifestation, although arrhythmias and embolic events also occur. Many women improve in NYHA functional status and ventricular function postpartum, but others have persisting problems or even worsen.

The relapse rate during subsequent pregnancies is substantial in women with evidence of persisting cardiac enlargement or left ventricular dysfunction. However, pregnancy may not be risk-free even in those who recover systolic function, as subclinical abnormalities may persist.<sup>25</sup>

A multicenter survey examined the outcomes of 60 pregnancies in women with peripartum cardiomyopathy diagnosed during a prior pregnancy. Of those with a poor left ventricular ejection fraction ( $< 0.50$ ), 44% developed symptoms of congestive heart failure and 19% died. Of those with better heart function (ejection fraction  $\geq 0.50$ ), 21% developed symptoms of congestive heart failure and none died.<sup>26</sup>

## ■ MANAGEMENT

The following five areas should be considered in the clinical approach to the woman with heart disease who is pregnant or considering pregnancy: 1) risk stratification, 2) antepartum management, 3) peripartum management, 4) recurrence of congenital lesion in the neonate, and 5) site of antepartum and peripartum care.

### **Assess risk before pregnancy**

Risk should ideally be assessed before a patient becomes pregnant. The data needed for risk

assessment can be acquired from:

- A thorough cardiovascular history and examination
- A 12-lead electrocardiogram
- A transthoracic echocardiogram
- An arterial oxygen saturation measurement by percutaneous oximetry (in patients with cyanosis).

The underlying cardiac lesion should be defined, and ventricular function, pulmonary pressure, severity of obstructive lesions, persistence of shunts, and presence of hypoxemia should be assessed.

When possible, surgery to correct cyanosis should be done before conception to improve maternal and fetal outcomes,<sup>2</sup> and symptomatic obstructive lesions should be corrected.<sup>13</sup> During pregnancy, cardiovascular surgery is more dangerous, involving a 6% risk of maternal mortality and a 30% risk of fetal mortality.<sup>27</sup>

Risk can be stratified according to the nature of the cardiac lesion, maternal factors (functional class or cyanosis), and the use of a risk index.

Low-risk patients include those with small left-to-right shunts, repaired lesions without residual cardiac dysfunction, isolated mitral valve prolapse without significant regurgitation, bicuspid aortic valve without stenosis, mild-moderate pulmonic stenosis, or valvular regurgitation with normal ventricular systolic function. Those at high risk include patients with significant pulmonary hypertension, Marfan syndrome with aortic root or major valvular involvement, and peripartum cardiomyopathy with residual left ventricular systolic dysfunction. The remaining cardiac lesions are considered to be in the intermediate-risk group.

Maternal functional class is an important predictor of outcome. A 1982 study of 482 pregnancies in women with congenital heart disease found that those in NYHA functional class I (ie, without limitation of physical activity) have lower cardiovascular morbidity and higher live birth rates.<sup>1</sup> Similarly, NYHA classes III (markedly limited activity) and IV (unable to be active without symptoms) predict adverse maternal cardiac events.<sup>5,6</sup>

In a prospective, multicenter study of 599 completed pregnancies,<sup>5</sup> four risk factors were

**If possible,  
avoid  
antiarrhythmic  
drugs during  
the first  
trimester**

TABLE 2

### Determining cardiovascular risk in pregnancy

#### Low-risk features

Small left-to-right shunt  
 Repaired lesion without residual cardiac dysfunction  
 Isolated mitral valve prolapse without significant regurgitation  
 Bicuspid aortic valve without stenosis  
 Mild-to-moderate pulmonary stenosis  
 Valvular regurgitation with normal ventricular systolic function

#### Intermediate-risk features

Unrepaired or palliated cyanotic congenital heart disease  
 Large left-to-right shunt  
 Uncorrected coarctation of the aorta  
 Mitral stenosis  
 Moderate aortic stenosis  
 Prosthetic valve  
 Severe pulmonary stenosis  
 Moderate-to-severe systemic ventricular dysfunction

#### High-risk features

New York Heart Association class III or IV symptoms (markedly limited physical activity or unable to perform any physical activity without symptoms)  
 Significant pulmonary hypertension  
 Marfan syndrome with aortic root or major valvular involvement  
 Eisenmenger syndrome  
 Severe aortic stenosis

identified that predicted a cardiac event (cardiac death, stroke, pulmonary edema, or arrhythmia) in pregnancy:

- Poor functional status (NYHA class III or IV) or cyanosis
- Left ventricular systolic dysfunction (ejection fraction < 0.40)
- Left heart obstruction (mitral valve area < 2.0 cm<sup>2</sup>, aortic valve area < 1.5 cm<sup>2</sup>, or peak left ventricular outflow tract gradient > 30 mm Hg)
- A cardiac event (arrhythmia, stroke, transient ischemic attack, or pulmonary edema) before pregnancy but since a prior cardiac surgical procedure.

The authors developed a risk index incorporating these factors. In a woman with heart disease and no other risk factors, the likelihood of a cardiac event during pregnancy is about 5%, increasing to 25% with one risk factor and 75% with more than one risk factor.<sup>5</sup>

This index should be used in conjunction

with lesion-specific risk estimates, if available, to predict a low (risk index of zero without high-risk lesion), intermediate (risk index of 1 without high-risk lesion), or high risk (risk index > 1 or high-risk lesion). Women at highest risk (eg, those with Eisenmenger syndrome or Marfan syndrome with a dilated aortic root) are less likely to undergo pregnancy and so were under-represented in contemporary studies (see TABLE 2.)

**Neonatal risk.** Maternal heart disease also increases the risk of neonatal complications,<sup>1-5,7</sup> especially if the mother also has noncardiac risk factors for neonatal complications (TABLE 3).<sup>7</sup>

### Antepartum management

**Limiting activity** is helpful in severely affected women with ventricular dysfunction, left heart obstruction, or class III or IV symptoms. Hospital admission by mid-second trimester may be advisable for some.

**Problems should be identified early** and treated aggressively, especially pregnancy-induced hypertension, hyperthyroidism, infection, and anemia.

**Beta-blockers** rather than digoxin should be used to control the heart rate for patients with functionally significant mitral stenosis. Empiric therapy with beta-blockers is offered to patients with coarctation, Marfan syndrome, and ascending aortopathy for other reasons (eg, a bicuspid aortic valve).

**Arrhythmias should be treated if warranted.** Premature atrial or ventricular beats are common in normal pregnancy, and in patients with preexisting arrhythmias, pregnancy may exacerbate their frequency and hemodynamic severity. These usually are not treated.

Pharmacologic treatment is usually reserved for patients with severe symptoms or when sustained episodes are poorly tolerated in the presence of structural cardiac abnormalities. Sustained tachyarrhythmias, such as atrial flutter or atrial fibrillation, should be treated promptly.

If possible, all antiarrhythmic drugs should be avoided during the first trimester, and those known to be teratogenic should be avoided throughout pregnancy.

Because of their safety profiles, preferred



drugs include digoxin, beta-blockers (possibly excluding atenolol), and adenosine.<sup>28</sup> One can also consider quinidine, sotalol, lidocaine, flecainide, and propafenone, but published data on their use in pregnancy are more limited.<sup>29</sup>

Amiodarone is generally regarded as contraindicated in pregnancy, although case reports describe its successful use. It is not teratogenic, but may impair neonatal thyroid function.<sup>30,31</sup>

Electrical cardioversion is safe. A report of 44 pregnancies in women with implantable cardioverter-defibrillators reported favorable maternal and fetal outcomes.<sup>32</sup>

**Anticoagulation therapy.** No current strategy is equally safe for both mother and fetus.

Oral therapy with warfarin is effective and logistically easy. However, it can affect embryonic organ development, although some evidence shows that a dosage of 5 mg per day may not be teratogenic.<sup>33</sup> Fetal intracranial bleeding is a risk throughout pregnancy, particularly during vaginal delivery, unless warfarin is stopped before labor.

Heparin in adjusted subcutaneous doses does not cross the placenta and so has no teratogenic effects. However, it may cause maternal thrombocytopenia and osteoporosis and is less effective in preventing thrombosis in patients with prosthetic valves.

In an overview of anticoagulation and pregnancy outcomes in women with prosthetic heart valves, the overall maternal mortality rate was 3%. Oral anticoagulation throughout pregnancy was associated with the lowest rate of valve thrombosis or systemic embolism (4%), while unfractionated heparin between 6 weeks and 12 weeks gestational age was associated with an increased risk of valve thrombosis (9%).<sup>34</sup>

Previous practice guidelines from 1998 recommended using either warfarin plus low-dose aspirin or heparin during the first 35 weeks of pregnancy and then heparin from the 36th gestational week onwards.<sup>13</sup>

The warfarin/aspirin strategy may be most appropriate if therapeutic anticoagulation can be achieved with a warfarin dosage of 5 mg per day.<sup>33</sup>

More recent guidelines recommend either (1) adjusted-dose heparin during the entire

**TABLE 3**

## **Risk factors for neonatal complications**

### **Maternal risk factors**

#### **Cardiac**

Poor functional class or cyanosis

Left heart obstruction

#### **Other**

Age < 20 or > 35

Multiple gestation

Smoking

Anticoagulant therapy

History of premature delivery

Membrane rupture

Incompetent cervix

Cesarean section

Intrauterine growth retardation

Antepartum bleeding after 12 weeks' gestation

Febrile illness

Uterine or placental abnormalities

### **Neonatal complications**

Premature birth

Low birth weight for gestational age

Respiratory distress syndrome

Intraventricular hemorrhage

Fetal or neonatal death

pregnancy or (2) adjusted-dose heparin until the 13th week of gestation, warfarin from the 14th week to the middle of the third trimester, and then restart adjusted-dose heparin.<sup>35</sup>

Low-molecular-weight heparin in adjusted doses is easier to administer and has been suggested as an alternative to adjusted-dose unfractionated heparin.<sup>35</sup> We currently utilize adjusted-dose low-molecular-weight heparin when heparin will be part of the antithrombotic regimen during pregnancy.

In women with prosthetic valves at high risk of thromboembolic complications, adding low-dose aspirin should also be considered.<sup>35</sup> Although high-dose aspirin may promote premature duct closure, low-dose aspirin is safe for the fetus, even at term.<sup>36</sup>

### **Peripartum management**

**Cesarean section** is indicated only for the following conditions:

- Aortic dissection

- Marfan syndrome with dilated aortic root
- Taking warfarin within 2 weeks of labor.

**Preterm induction is uncommon.** However, once fetal lung maturity is assured, a planned induction and delivery may be warranted for high-risk patients to ensure that appropriate staff and equipment are available.

**Hemodynamic monitoring.** No consensus exists on using invasive hemodynamic monitoring during labor and delivery. We commonly use intra-arterial monitoring and may also use central venous pressure monitoring if interpreting a sudden drop in systemic blood pressure is of concern. A pulmonary artery catheter is rarely indicated.

**Heparin anticoagulation should be discontinued** at least 12 hours before induction, or reversed with protamine if spontaneous labor develops. It can usually be resumed 6 to 12 hours postpartum.

**Antibiotic prophylaxis for endocarditis is not routine.** American Heart Association guidelines do not recommend routine endocarditis prophylaxis for cesarean section delivery or for uncomplicated vaginal delivery without infection.<sup>37</sup>

However, some centers do administer endocarditis prophylaxis for vaginal delivery in women with structural heart disease, as an uncomplicated delivery cannot always be anticipated.

**Pain control** should be offered with epidural anesthesia and adequate volume preloading. Epidural fentanyl does not lower peripheral vascular resistance, making it particularly advantageous for cyanotic patients with shunt lesions or significant aortic stenosis. Air-and-particulate filters should be placed in all intravenous lines for patients with a shunt.

**Positioning the patient on her left side** lessens the hemodynamic fluctuations associated with contractions when the patient is supine.

**Forceps or vacuum extraction** should be considered at the end of the second stage of labor to shorten and ease delivery.

**Postpartum monitoring.** Because hemodynamics do not return to baseline for many days after delivery, patients at intermediate or

high risk may require monitoring for at least 72 hours postpartum.

Patients with Eisenmenger syndrome are at risk of death for up to 7 days postpartum, and so require close observation longer.

## ■ RISK OF CONGENITAL HEART DISEASE IN OFFSPRING

The risk of congenital heart disease is 0.4% to 0.6% in the general population; this risk increases about 10-fold if a first-degree relative is affected.<sup>38</sup>

Left-sided heart obstructive lesions have a higher rate of transmission to offspring. Certain conditions, such as Marfan syndrome and the 22q11 deletion syndromes, are autosomal-dominant, conferring a 50% risk of transmission.<sup>38</sup>

Patients of reproductive age with congenital heart disease should be offered genetic assessment and counseling so that they are fully informed of the transmission risk and of the options for prenatal diagnosis. Strategies to decrease the incidence of congenital defects, including taking multivitamins preconception, should also be discussed.<sup>39</sup>

## ■ HIGH-RISK PREGNANCY UNITS

Women at intermediate or high risk should be managed in a high-risk pregnancy unit by a multidisciplinary team staffed by obstetricians, cardiologists, anesthesiologists, and pediatricians. Candidates for this care include:

- Those with at least one risk factor according to the risk index (TABLE 2)
- Those with lesion-specific risks in the high-risk category
- Those at risk for neonatal complications (TABLE 3).<sup>4,5</sup>

The multidisciplinary team should meet with patients early in pregnancy and write a management plan for most contingencies.

Women with heart disease deemed to be at low risk can be managed in a community hospital. If the mother's status or her risk profile is in doubt, a consultation at a regional referral center should be arranged. ■

Lying on the left side decreases hemodynamic fluctuations during contractions



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