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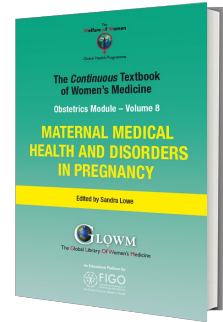
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## **The Continuous Textbook of Women's Medicine Series – Obstetrics Module Volume 8**

### **MATERNAL MEDICAL HEALTH AND DISORDERS IN PREGNANCY**

**Volume Editor:** *Clinical Associate Professor Sandra Lowe*



## *Chapter*

# **Pregnancy in the Woman with Cardiovascular Disease – Congenital and Acquired**

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## **INTRODUCTION**

Hemodynamic changes of pregnancy in women with pre-existing cardiovascular disease may complicate the course of pregnancy. Historically, underlying rheumatic heart disease was the commonest cause of cardiac complications in pregnancy. However, in the developed world, women with complex and/or repaired congenital heart disease or those with acquired myocardial disease comprise the highest risk group.<sup>1,2</sup> Many women with underlying cardiac disease are able to tolerate pregnancy successfully with careful management. Data from the Registry on Heart Disease and Pregnancy (ROPAC) demonstrates that planned cesarean section for women with heart disease confers little if any benefit for the mother and is associated with earlier delivery.<sup>3</sup> As such, guidance generally recommends vaginal delivery unless there are obstetric indications that favor cesarean section.

This chapter focuses on pregnancy in women with a history of heart disease, including valvular, congenital, aortic, and myocardial diseases. It should be noted that many women with heart disease will go on to have uncomplicated pregnancies. Nevertheless, the risks associated with pregnancy need to be identified and discussed, pre-pregnancy with the woman and her partner.

### **Cardiovascular changes during pregnancy and delivery**

The normal hemodynamic alterations that occur during pregnancy are summarized in Table 1. These changes begin as early as 6 weeks, peaking at approximately 20 weeks' gestation. The greatest reduction in blood pressure is seen in the first trimester and greatest increase in cardiac output in the second, whereas maximal heart rate increase happens in the third trimester.<sup>4</sup>

**Table 1** Summary of hemodynamic changes of pregnancy.

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

Stroke volume increases contributing to the increased cardiac output and preload early in pregnancy. Augmented preload may not be tolerated by obstructive cardiac lesions, such as mitral or aortic stenosis, or those with impaired ventricular function. Normal pulmonary compliance can accommodate this additional blood volume and preload burden. However, in the setting of pulmonary hypertension when there is reduced pulmonary vascular compliance, right-side heart failure may ensue and left-to-right shunting within the heart reverses, causing decreased oxygen saturation of the blood and cyanosis.

Systemic blood pressure decreases because systemic vascular resistance decreases beginning in early pregnancy. This effect is maximal in the second trimester. There is a decrease in the mean aortic pressure and a widening of the pulse pressure. This decrease in afterload from systemic vasodilation in a woman with aortic stenosis may further increase the gradient across the aortic valve, adding to the left ventricular work. Conversely, certain cardiac lesions may benefit from afterload reduction, such as mitral regurgitation and aortic insufficiency. These murmurs may soften and the echocardiographic severity of regurgitation decreases during pregnancy.

In the supine position, the gravid uterus compresses the inferior vena cava, acutely decreasing preload and causing the syndrome of supine syncope.

## During delivery and postpartum

The hemodynamic changes of labor and delivery pose potential problems for the woman with heart disease, and should be anticipated. With each uterine contraction during labor, there is a bolus of fluid into the intravascular space.<sup>5</sup> This change, although transient, is repetitive and may exacerbate certain cardiac problems by augmenting cardiac output by 15–20%, with a 10% increase in mean systemic arterial blood pressure.<sup>6</sup> A reflex bradycardia may occur. These changes have been shown to be attenuated with the patient lying in the left lateral decubitus position.<sup>6</sup> Pain and anxiety stimulate the sympathetic nervous system, causing an increase in blood pressure and heart rate. Prolonged Valsalva maneuver required during the second stage of labor may increase blood pressure and afterload, potentially complicating aortic disease.

Epidural anesthesia with appropriate pre-hydration provides pain control and may prevent marked fluctuations in blood pressure. The vasodilatation secondary to epidural analgesia or anesthesia may result in a precipitous decrease in blood pressure, and the woman with obstructive valvular disease or a cardiomyopathy may experience pulmonary edema with vigorous fluid resuscitation.

There is some information about the circulatory changes that occur during pregnancy, labor, and delivery; but relatively little is known about what occurs after the time of delivery. Postpartum hemodynamic changes may resolve by 6 weeks, but it may take up to 12 weeks for the cardiovascular changes of pregnancy to resolve.

Preconception evaluation

Women with cardiac disease should undergo a thorough preconception evaluation to identify all possible risks to the woman and the baby when embarking upon pregnancy, and to evaluate for the potential need for pre-emptive treatment to improve her cardiac status prior to pregnancy.<sup>1,2</sup> Specific diagnostic modalities are outlined to help focus diagnostic testing for specific clinical problems.

Full cardiac evaluation includes diagnostic tests appropriate to assess the feasibility and safety of pregnancy. This may include stress testing to quantify functional capacity. Exercise testing with impaired chronotropic response prior to pregnancy in women with congenital heart disease has been shown to relate to adverse obstetric events.<sup>7</sup> Cardiac catheterization may be indicated to assess hemodynamics, complex anatomy, coronary artery disease, and need for catheter-based interventions.<sup>8</sup> Evaluation may conclude that cardiac surgery is indicated to actually improve the outcome of a future pregnancy. Genetic counselling may be indicated for the woman with congenital heart disease. This would include conditions such as Marfan syndrome, many of the cardiomyopathies, and the inherited arrhythmias such as long QT or catecholaminergic polymorphic ventricular tachycardia.

Evaluation during pregnancy

Symptoms that arise during the course of a normal pregnancy are similar to those reported by patients with cardiac disease (Table 2). Distinguishing the symptoms of normal pregnancy from pathological symptoms is very difficult.

Table 2 Cardiovascular symptoms during pregnancy.

<p><b>Normal pregnancy:</b></p> <p>Fatigue</p> <p>Palpitations</p> <p>Lower extremity swelling</p> <p>Dyspnea</p> <p>Chest pain</p> <p>Syncope, presyncope</p>
<p><b>Potentially pathological symptoms that may indicate cardiac disease and require further investigation:</b></p> <p>Palpitations: symptomatic at rest, persistent, with associated symptoms</p> <p>Edema</p> <p>Dyspnea: progressive, nocturnal or at rest</p> <p>Orthopnea</p> <p>Chest pain: exertional or at rest</p> <p>Syncope, presyncope: exertional</p>

Physical findings detected during a normal pregnancy may be similar to those of patients who present with cardiac disease (Table 3). The physical findings of pregnancy are caused by expected hemodynamic alterations with increase in heart rate, fluid retention, expanded blood volume, and decreased afterload. Sinus tachycardia may be present at rest, especially during the third trimester. The cardiac examination in pregnancy may be notable for a hyperdynamic, diffuse precordial impulse that becomes displaced to the left as pregnancy progresses. The right ventricular impulse may be more prominent. The first and second heart sound may increase. Heart murmurs are heard in nearly 96% of pregnant women; classically, it is a mid-systolic murmur that is heard best along the left sternal border. Murmurs of mitral and aortic regurgitation may soften because of the decrease in afterload that occurs during the pregnancy. The murmurs of mitral stenosis and aortic stenosis are intensified because of increased preload and increased flow across the stenotic valve. Peripheral edema is caused by fluid retention and decreased plasma oncotic pressure during pregnancy.

Table 3 Cardiovascular examination during pregnancy.

<p><b>Normal pregnancy:</b></p> <p>Tachycardia</p> <p>Dilated neck veins</p>
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Bounding pulses, dynamic precordium  
 Third heart sound  
 Systolic murmur (intensity 1–2/6)  
 Peripheral edema (dependent)

**Potentially pathological symptoms that may indicate cardiac disease that requires further investigation:**

Bradycardia (pulse <50 beats/min)  
 Tachycardia  
 Elevated jugular venous pressure  
 Cardiomegaly  
 Right ventricular heave  
 Loud pulmonic component of second heart sound (S2)  
 Summation gallop, i.e. 3rd and 4th heart sounds  
 Loud systolic murmur (intensity 3–6/6)  
 Diastolic murmur  
 Cyanosis or clubbing  
 Persistent crackles  
 Persistent edema unresponsive to elevation

## Cardiovascular diagnostic testing during pregnancy

### Electrocardiographic

Table 4 outlines diagnostic tests used in pregnancy. The resting electrocardiogram is altered by pregnancy. The QRS axis may shift leftward as the heart becomes displaced laterally in the thorax, and nonspecific ST-T wave changes may be noted. Women have increased atrial and ventricular ectopy during pregnancy and may experience palpitations.<sup>9</sup> For detection of an intermittent arrhythmia not documented on a standard 12-lead electrocardiogram, a 24-hour or 48-hour ambulatory Holter monitor may be very helpful. If the arrhythmia is sporadic but is associated with symptoms, then a 30-day event monitor may be more useful.

**Table 4** Cardiac diagnostic testing during pregnancy.

Diagnostic modality	Indication
ECG, Holter monitor, event monitor	Detect arrhythmia
Doppler echocardiography	Assess valvular heart disease Estimate pulmonary artery pressure Assess cardiac function
Transesophageal echocardiography	Detect atrial thrombus Diagnose atrial septal defect or endocarditis Diagnose aortic dissection
Stress testing (heart rate limited) may be combined with echocardiogram	Provoke arrhythmia; assess valve disease severity Detect ischemia
Radionuclear imaging	Is rarely performed in pregnancy because of potential radiation effects
Cardiac catheterization/angiography	To diagnose and treat coronary artery disease
Magnetic resonance imaging	Assess whole aorta, congenital heart disease, some myocardial diseases

An electrocardiographic stress test may be performed during pregnancy to assess functional capacity, identify ischemia,

or provoke suspected arrhythmias. Stress testing can be performed safely during pregnancy, usually to the predetermined heart rate endpoint of 120–140 beats/min, depending on the patient's functional state and her exercise history.<sup>10</sup> Exercise stress testing in conjunction with radionuclide isotopes imaging is generally avoided because these agents concentrate in the bladder and constitute a radiation risk to the developing fetus. Stress testing combined with echocardiography may be useful to detect ischemia occurring with exercise.

### ***Echocardiography***

Echocardiography provides an excellent diagnostic modality for most cardiac conditions during pregnancy. Serial two-dimensional echocardiography can help to detect changes in cardiac chamber dimensions, wall thickness, or the presence of a pericardial effusion occurring or changing during pregnancy. In women with established heart disease, in particular those with congenital heart disease, repeat or serial imaging during pregnancy is generally dictated up their underlying lesion. In the presence of suspected heart failure in pregnancy, echocardiography can provide the clinician with immediate information regarding left ventricular systolic function. Chamber enlargement may occur in a normal pregnancy, with an increase in the fractional shortening. Left ventricular muscle mass as determined by echocardiography may increase during a normal pregnancy and may increase further in the setting of pre-eclampsia.<sup>11,12</sup> Doppler echocardiography can help to diagnose and follow the course of stenotic or regurgitant valvular lesions and estimate right-side cardiac pressures to detect pulmonary hypertension. Mitral and tricuspid valve regurgitation have been noted during pregnancy in the structurally normal heart. Physiologic mitral regurgitation has been thought to be secondary to annular dilatation with the expected ventricular enlargement. The transesophageal echocardiogram performed under sedation is particularly useful for the diagnosis of atrial thrombus, atrial septal defect, and aortic disease, such as thoracic aortic dissection.

### ***Cardiac catheterization***

Cardiac catheterization and angiography may be required to diagnose the severity of coronary artery disease in patients with anginal symptoms. Right-side heart catheterization may be needed to definitively diagnose the severity of pulmonary hypertension, quantify intracardiac shunts, and assess severe valvular disease.

### ***Magnetic resonance imaging***

Magnetic resonance imaging defines the cardiac anatomy and is particularly useful in the diagnosis of congenital heart disease and aortic disease. It may aid clinical decision making in pregnancy.<sup>13</sup> Gadolinium is not recommended during pregnancy.

### ***Risk prediction***

Several different risk prediction models have been diagnosed to help predict complications for women with heart disease and pregnancy.<sup>14,15,16</sup> The modified World Heart Organization (mWHO) is the most utilized model and is probably the most accurate with regards to risk assessment of the parturient with heart disease. The mWHO criteria,<sup>17</sup> as described in the most recent European Society of Cardiology (ESC) guidelines also assist by suggesting where delivery should be planned and how often women should be assessed during pregnancy.<sup>2</sup> Other risk prediction scores have been identified such as the CARPREG I and II and the ZAHARA scoring systems.<sup>14,15,16</sup> These can further assist in classifying maternal and fetal risks in women with underlying cardiac disease.

It is important to carefully assess the woman's functional capacity, her exercise history, and her ability to perform daily activities. The woman with normal function status (NYHA I) and normal ventricular function despite the presence of cardiovascular disease is more likely to sustain a normal pregnancy.

Women who fall into category IV of the mWHO model are advised against pregnancy because of the high risk of maternal mortality. This includes women with pulmonary arterial hypertension, previous peripartum cardiomyopathy with residual myocardial dysfunction, any severely impaired left ventricular dysfunction, severe mitral or aortic stenosis or significant aortic disease i.e. aneurysmal or coarctation.

### ***Cardiac medications***

The ROPAC registry showed that almost a third of women with heart disease took some form of cardiac medication

during pregnancy.<sup>18</sup> Women who were more likely to be prescribed cardiac medications were older, parous and more likely to have valvular heart disease. Women maybe prescribed a variety of cardiac medications and it is the job of the treating medical team to understand any risks of these medications during pregnancy and breastfeeding. Where there is limited pregnancy specific data, women should be counselled appropriately and where appropriate switched to be alternate agent that is considered safe. If a medication is considered of benefit during pregnancy but there is inadequate data on its safety profile, the treating medical team should discuss the risks and benefits of continuing the medication if this is felt to be clinically appropriate. The following section covers commonly used cardiac medications.

### ***Beta-blocker***

Beta-blocker are frequently used in women with cardiac disease and cohort studies suggest these are the most commonly prescribed type of cardiac medication.<sup>18</sup> Beta-blocker e.g. atenolol, metoprolol, bisoprolol, sotalol and carvedilol can be continued during pregnancy but they are potentially associated with impaired fetal growth.<sup>19,20</sup> transient neonatal hypoglycaemia and bradycardia. Beta-blockers with vasodilator properties e.g. labetalol (used for hypertension) do not cause reduced fetal weight.

### ***Calcium channel blockers***

Verapamil and diltiazem are used in pregnancy for management of arrhythmias. Diltiazem may be teratogenic in animals and there is inadequate data in humans. The ESC recommends against its use in the first trimester.<sup>2</sup> These are considered safe used in during pregnancy and in breastfeeding. Caution should be used in women with severe pre-eclampsia as the combined used of magnesium sulphate and calcium channel blockers can potentiate profound hypotension.

### ***Antiarrhythmic agents***

Adenosine, digoxin, lignocaine/lignocaine procainamide and quinidine are all considered safe for use in pregnancy and in breastfeeding. There is insufficient human data regarding the use of flecainide in the first trimester but animal data does suggest teratogenicity.<sup>21</sup> It is used later in pregnancy for both maternal and fetal arrhythmias. For women with arrhythmias that are refractory to these agents, amiodarone can be considered but it may cause neonatal hypothyroidism, hyperthyroidism, goitre, fetal bradycardia, prolonged QT, small for gestational age and prematurity.<sup>22</sup>

### ***Diuretics and aldosterone antagonists***

Furosemide, bumetanide and hydrochlorothiazide may be used in pregnancy and breastfeeding. These medications should be used for the management of heart failure and not pre-eclampsia. Spironolactone is anti-androgenic and in animal studies, using high doses, it was associated with feminisation of male rats and permanent dose-related changes in the reproductive tracts in offspring of both sexes when used later in pregnancy.<sup>23</sup>

### ***Anticoagulants***

Low molecular weight heparins (LMWH), e.g. enoxaparin, dalteparin, tinzaparin, are large molecules that have no placental transfer and can be used in pregnancy and breastfeeding, They can cause maternal bruising but rarely cause thrombocytopenia.

### ***Vitamin K antagonists***

These agents are considered teratogenic when used in the first trimester (see section on metallic heart valves). When used in the second and third trimesters the fetal International Normalised Ratio (INR) will be much greater than maternal INR and there is a risk of neonatal intraventricular hemorrhage. Vaginal delivery is contraindicated in women taking VKAs because the resulting anticoagulation effects on the fetus increase the risk of cranial hemorrhage during the birth.

### ***Other oral anticoagulants***

Novel and direct oral anticoagulants are not recommended for use in pregnancy or breastfeeding. There is limited data regarding their use but clinicians should ensure women of child bearing age using these medicines have access to

reliable contraception.<sup>24</sup>

### **Antiplatelet agents**

Low-dose aspirin is safe for use throughout pregnancy. Clopidogrel also appears to be safe in pregnancy but the data regarding its use is sparse. Reassuringly there is no data to show there is an increased risk of neonatal hemorrhage, or that there is an increased risk of placental abruption with the use of clopidogrel.<sup>25</sup> It is generally recommended to stop Clopidogrel 7–10 days prior to planned delivery because of increased risk of maternal bleeding at delivery.<sup>26</sup>

### **Statins**

At present there is a paucity of data to recommend statins use in pregnancy. However, there is no data to suggest there is an increased risk of teratogenicity if these drugs are used in the first trimester.<sup>27</sup> Furthermore, statins have been used in clinical trials relating to early onset pre-eclampsia.<sup>28</sup> Statins therefore may be used in selected cases in pregnancy if there is a compelling benefit for their use.

## **VALVULAR HEART DISEASE**

Historically, rheumatic heart disease was the major cause of valvular heart disease in women of childbearing age. Although rheumatic heart disease occurs less commonly in developed countries, the sequelae of rheumatic heart disease remain a significant problem in low and middle income countries (LMIC); and it remains a significant health issue for women and their caregivers in those areas as well as predominantly migrant women in the developed . More recently congenital valvular heart disease is increasingly recognized.<sup>29</sup> This includes valvular disease diagnosed at birth or during childhood, with its sequelae diagnosed during adulthood in the young woman. Potential complications are summarized in Table 5. Valvular heart disease in general is associated with not only increased risk for cardiovascular complications but also obstetric complications. There is increased incidence of preterm delivery, intrauterine growth retardation, and lower birth weight in women with valvular heart disease.<sup>29</sup>

**Table 5** Valvular heart disease in pregnancy.

Valve lesion	Potential risks in pregnancy
Tricuspid regurgitation	Associated with other cardiac lesions but usually well tolerated
Pulmonic stenosis/insufficiency	Usually well tolerated or asymptomatic prior to pregnancy. May be of greater concern after surgery for congenital heart disease
Mitral stenosis	Atrial fibrillation, cerebral or systemic thromboembolism, pulmonary edema
Mitral regurgitation	Well tolerated and may decrease with decreased afterload
Aortic stenosis	Arrhythmia, heart failure, or syncope
Aortic insufficiency	Well tolerated and may decrease with decreased afterload

### **Tricuspid valve disease**

Tricuspid valve involvement in rheumatic heart disease usually is mild and occurs with multi-valvular involvement. Rheumatic tricuspid stenosis usually occurs with mitral and/or aortic valve involvement. This occurs more commonly in women and is often well tolerated, even when intervention has been required for mitral or aortic disease.

The tricuspid valve is often involved in complex congenital cardiac disease or after surgical repair. In general, tricuspid regurgitation is well tolerated during pregnancy. Severe tricuspid valve regurgitation caused by previous endocarditis is rare but has been associated with illicit intravenous drug use.



## Pulmonic valve disease

Pulmonic valve stenosis is most often congenital in origin. In adults, it is well tolerated when of mild or moderate severity.<sup>30</sup> Severe pulmonic stenosis may require percutaneous balloon valvotomy, which is best performed before pregnancy. The woman with severe pulmonic stenosis may become symptomatic, with the increased preload of pregnancy, and may have secondary tricuspid regurgitation. Pulmonic stenosis is often associated with other congenital heart disease such as tetralogy of Fallot. It may also be present after tetralogy of Fallot repair. Pulmonic insufficiency usually is well tolerated during pregnancy as long as right ventricular function is normal and tricuspid regurgitation is minimal.

## Mitral valve disease

### *Mitral stenosis*

Mitral stenosis (MS) is the most common rheumatic valvular lesion diagnosed during pregnancy. Rheumatic carditis can result in MS, mitral regurgitation, or both. In uncomplicated MS, pulmonary venous hypertension develops as a result of the gradient required to maintain adequate transvalvar blood flow. Increased pulmonary pressure causes right ventricular dilation with dysfunction and tricuspid regurgitation. Patients may present with overt right heart failure. The increased volume load during pregnancy secondary to obstruction to outflow from the left atrium, may cause a woman to have symptoms for the first time in her life. The expanded stroke volume, augmented cardiac output, and increased heart rate of pregnancy hemodynamically challenge the woman with MS. Expanded blood volume and left atrial enlargement enhance the risk of atrial fibrillation. Atrial dilation and atrial fibrillation in the hypercoagulable pregnant patient can promote thrombus formation, necessitating therapeutic anticoagulation to prevent systemic embolization. The degree of MS can be assessed by two-dimensional and Doppler echocardiography evaluating left atrial size and ventricular function. Pulmonary arterial pressure can be estimated by the velocity of the tricuspid regurgitation jet and the trans-mitral gradient assessed with Doppler echocardiography. Thrombus in the left atrial appendage are best visualized with trans-esophageal echocardiogram. During pregnancy, echocardiography, at least once per trimester, is helpful to monitor left atrial size, left ventricular function, or worsening of the MS.

The major risks during pregnancy are atrial fibrillation, embolic events, and pulmonary edema.<sup>31</sup> The European Registry of Heart Disease and Pregnancy authors published contemporary data on 273 women with MS and pregnancy.<sup>32</sup> Maternal mortality during pregnancy was 1.9% and heart failure occurred in nearly a quarter of women with moderate or severe MS. Women with mild or symptomatic disease tolerated pregnancy well.

Atrial fibrillation in a pregnant woman with MS may result in pulmonary edema or systemic embolization, causing a cerebrovascular event. Selective beta-adrenergic blockade is recommended if needed to maintain sinus rhythm and slow the heart rate in atrial fibrillation. If atrial fibrillation leads to hemodynamic compromise, electrical cardioversion may be required after adequate systemic anticoagulation.<sup>33</sup>

Percutaneous trans-septal mitral balloon valvuloplasty has been performed in pregnant women with symptoms refractory to medical therapy.<sup>34</sup> This procedure may forestall the need for surgical valve replacement. Several series of patients have been reported in recent years.<sup>34,35,36</sup> Balloon mitral valvuloplasty should not be performed if significant mitral regurgitation or left atrial thrombus is present.

Cardiac surgery is indicated when a woman's symptoms do not resolve, despite medical therapy, and if balloon valvuloplasty is not an option.<sup>37</sup> Cardiopulmonary bypass may impact placental circulation and systemic hypothermia may precipitate uterine contractions. Surgery, if necessary, should only be performed in centers with appropriate expertise, preferably during the second trimester and using high-flow, high-pressure, normothermic perfusion and continuous fetal heart rate monitoring.<sup>38</sup>

### *Mitral regurgitation*

Mitral regurgitation (MR) may be the result of mitral valve prolapse (MVP), previous endocarditis, ruptured chordae tendoneae, rheumatic disease, or after congenital cardiac repair. MR and MVP may occur as part of the Marfan syndrome. In the presence of normal left ventricular function, pregnancy is well tolerated. The pregnancy related decrease in systemic vascular resistance reduces the severity of MR. Women with severe MR and limited exercise



capacity may benefit from mitral valve repair before conception. Severe symptomatic MR in a woman with NYHA functional class III–IV refractory to medical therapy may require surgical intervention.

## Aortic valve

### *Aortic stenosis*

Aortic stenosis (AS) is not common in women of childbearing age. It can be a result of rheumatic disease, or previous endocarditis. More often it is due to a congenital bicuspid aortic valve that may not have been detected before adulthood. The bicuspid aortic valve may be clinically silent or, if stenotic, symptoms may first develop during pregnancy. This is often associated with a degree of proximal aortopathy. As seen in MS, the severely stenotic aortic valve may not tolerate the increased cardiac output, stroke volume, and heart rate of pregnancy, resulting in congestive heart failure. The ROPAC investigators reported pregnancy outcomes for 96 women with at least moderate AS (34 with severe AS-peak gradient  $\geq 64$  mmHg).<sup>39</sup> There were no maternal deaths in their study, but over a fifth of women were hospitalized for cardiac reasons during the course of their pregnancy. Almost 7% of women developed heart failure. Infants born to mothers with severe AS were more likely to have a lower birth weight. Mild to moderate aortic stenosis is typically well tolerated.<sup>40</sup>

Women with critical AS, a valve area of less than  $1.0 \text{ cm}^2$ , should be discouraged from becoming pregnant, especially if forward cardiac output is compromised, causing symptoms and/or reduced functional capacity. Critical AS that is identified before conception may benefit from surgical repair. Percutaneous balloon aortic valvuloplasty has been reported during pregnancy when symptoms develop in the second or third trimester.<sup>41</sup> This may be a palliative measure to alleviate symptoms during pregnancy. Surgical intervention may be required if medical management fails and the patient is not a candidate for percutaneous valvuloplasty. Therefore, if moderate aortic stenosis is identified before pregnancy, it may be best to advise the patient to proceed with pregnancy before stenosis progresses and surgery is required.

Currently, women of childbearing age with critical aortic disease (stenosis and/or insufficiency) may have had a Ross procedure. A pulmonary autograft replaces the aortic valve and a homograft is placed in the aortic position. The coronary arteries are re-implanted. This surgery obviates the need for a mechanical prosthesis. Women undergoing pregnancy with a prior Ross procedure generally have a minimal pregnancy complications.<sup>42</sup>

### *Aortic insufficiency*

Aortic valve insufficiency (AI) may be the result of rheumatic fever, endocarditis, vasculitis, aortic dilation, Marfan syndrome, or commonly a congenitally bicuspid aortic valve. Mild to moderate AI with normal ventricular size and function is often well tolerated because of the vasodilatation and decrease in systemic vascular resistance. Therefore, the murmur of aortic insufficiency may decrease as pregnancy progresses along with the severity of regurgitation by echocardiography. Severe AI results in left ventricular dilation, which ultimately causes decreased left ventricular function. The woman with marked left ventricular dilation and left ventricular dysfunction secondary to AI would not tolerate the further volume load of pregnancy.

## Valve replacement and pregnancy

A woman of childbearing age requiring porcine valve replacement may face a reoperation, in the future. The benefit of a porcine valve or homograft is that therapeutic anticoagulation is not required. The use of bioprostheses (porcine) may eliminate the need for anticoagulation in the aortic position but not in the mitral position. Structural valve deterioration has been described in the teenage years and young adulthood, but pregnancy may not contribute to this phenomenon.<sup>43</sup> The changes in the bioprostheses appear to be independent of pregnancy. These risks must be considered in any woman of childbearing age who requires valve replacement.

A mechanical valve requires full therapeutic anticoagulation, which further complicates pregnancy by posing risks to mother and fetus.<sup>44,45</sup> Decisions around anticoagulation in the setting of a metallic prosthesis should be multidisciplinary and should involve the patient and her partner. These women should be managed by individuals with expertise in this area and care should involve if necessary an obstetric hematologist.

Low molecular weight heparin is commonly used but requires twice daily dosing to ensure a therapeutic range is

established.<sup>46</sup> Both trough and peak level monitoring have been proposed, with target levels of  $\geq 0.6$  IU/ml (trough) and  $\leq 1.5$  IU/ml (peak).<sup>47</sup> The ESC and ACC guidelines recommend dosing according to peak levels only.<sup>2,48</sup> In low-resource settings, unfractionated heparin, given as continuous infusion in the first trimester, aiming for an activated partial thromboplastin time of greater than twice the control value may be the only safe option. It is also recommended to add aspirin after the first trimester.<sup>47</sup>

Warfarin can also be considered for management of anticoagulation for women with a metallic prosthesis. When used in the first trimester, warfarin may cause a dose-dependent embryopathy that includes nasal hypoplasia, saddle nose, and stippled epiphysis in up to 10% of exposed cases.<sup>34</sup> The risk of embryopathy has been noted to be increased when the dose is more than 5 mg of coumadin.<sup>49</sup> It causes fetopathy in 0.7–2% of cases when used in the second and third trimester which may include neonatal intraventricular hemorrhage, ocular and central nervous system abnormalities.

Furthermore, warfarin is associated with a risk of miscarriage that is higher than that associated with heparin use.<sup>50</sup> However, warfarin is a superior agent when compared with low molecular weight heparin (LMWH) with regards to the risk of valve thrombosis, but valve thrombosis but even occur with warfarin use. Several different regimens for the use of warfarin and/or LMWH usage have been proposed for use in pregnancy.<sup>2</sup> Intravenous unfractionated heparin may also be used instead of warfarin but requires inpatient admission and intensive monitoring and regular dose adjustments.

Data from a systematic review showed that women taking vitamin K antagonists had the lowest risk of maternal complications but also the fewest live births.<sup>51</sup> Elkayam proposes a management strategy for metallic heart valves in pregnancy which includes switching warfarin to LMWH in the first trimester and then admission at 37 weeks' gestation to convert the woman to unfractionated heparin (UFH) by continuous infusion with prior to induction of labor.<sup>46</sup> All this requires meticulous planning and active patient engagement. The European Registry of Heart Disease and Pregnancy showed the risk of maternal mortality with a metallic valve in pregnancy was 1.5%;<sup>52</sup> a recent UK cohort study showed the risk to be much higher at 9%.<sup>53</sup>

## CONGENITAL HEART DISEASE

Due to the advances in the medical and surgical treatment of congenital heart disease more women are reaching childbearing age.<sup>54</sup> In developed countries complications as a result of congenital heart disease in pregnancy are more common than rheumatic heart disease. The risks and outcome of pregnancy in women with congenital heart disease will vary with the type of congenital heart disease, whether there has been surgical repair, the type of repair, and the sequelae of intervention.

Congenital heart diseases that may remain unrepaired seen in the adult patient include atrial septal defects, ventricular septal defect, patent ductus arteriosus, and Ebstein's anomaly (Table 6). Unrepaired complex cyanotic congenital heart disease rarely presents in adults. These patients will usually have undergone palliation or, more often, repair. Although they have had a physiologic (but not necessarily anatomic) correction or repair as a child, residual heart disease and/or surgical sequelae may persist into childbearing age. The type of surgery and subsequent cardiac status need to be thoroughly assessed and treated before conception.

**Table 6** Potential complications of unrepaired or incompletely repaired congenital heart disease in pregnancy.

Cardiac lesion	Potential complications during pregnancy
Atrial septal defect	Paradoxical embolus Eisenmenger's physiology and cyanosis
Ventricular septal defect	Endocarditis Eisenmenger's physiology and cyanosis
Patent ductus arteriosus	Eisenmenger's physiology and cyanosis

Eisenmenger's syndrome Cardiac lesion	Cyanosis Potential complications during pregnancy
	Fetal growth retardation Maternal mortality
Coarctation of the aorta	Decreased uterine perfusion Hypertension Aortic dissection Intracranial hemorrhage from associated cerebral aneurysms Bicuspid aortic valve pathology
Ebstein's anomaly	Increased tricuspid insufficiency Right ventricular failure Arrhythmias; heart block

Maternal cyanosis is associated with higher incidence of abortion, stillbirth, and small-for-gestational-age babies.<sup>55</sup> In addition, the offspring of women (or men) with congenital heart disease are at increased risk for congenital heart disease. Overall the risk of congenital heart disease in the offspring is between 3 and 5%. Fetal echocardiography, optimally performed between 18 and 22 weeks' gestation, can identify most major cardiac malformations to help anticipate and treat problems in the neonatal period. Fetal echocardiography has been shown to detect major congenital cardiac anomalies in the fetus.<sup>56</sup>

### Atrial septal defect

Atrial septal defect (ASD) is one of the most common congenital heart lesions found in adults. If undetected or unrepaired during childhood, a secundum-type defect is more common than a primum or sinus venous type of defect. The secundum atrial septal defect usually persists or enlarges in the region of the fossa ovalis. This defect is more common in females, and mitral valve prolapse is often seen. The ostium primum defect usually is detected and closed during childhood; however, after surgery a shunt may persist and often is associated with cleft mitral valve, which can be regurgitant in adults even after valvular repair.

The theoretical risk of an uncorrected ASD during pregnancy would be volume overload, which would further enlarge the right atrium and ventricle. ASDs complicated by severe pulmonary hypertension usually are detected before pregnancy and constitute a contraindication to pregnancy. Data from a multicenter study showed that when women with a repaired ASD were compared with an unrepaired ASD they had comparable outcomes with regards to pregnancy and cardiac complications.<sup>57</sup>

### Ventricular septal defect

The ventricular septal defect (VSD) is the most common congenital heart disease lesion identified at birth; however, the defect may close spontaneously within the first 5 years of life. If the VSD is closed early in childhood, and pulmonary pressures are normal, pregnancy is not of increased risk if pulmonary pressures are normal. Uncorrected VSDs can be associated with pulmonary hypertension, and Eisenmenger's syndrome and are managed as high risk. Women with unrepaired VSD and no evidence of a shunt appear to have similar pregnancy outcomes to those with a repaired VSD.<sup>58</sup>

### Patent ductus arteriosus

When diagnosed during childhood, a patent ductus arteriosus (PDA) is usually ligated. It rarely is detected during adulthood. Elevation of pulmonary artery pressure and potential reversal of the shunt with profound systemic hypotension pose the same risks as do septal defects. Previous repair without another congenital lesion, normal pulmonary pressures, and preservation of left ventricular function usually do not impact the course of pregnancy.

## Eisenmenger's syndrome

Eisenmenger's complex is the hemodynamic consequence of a shunt lesion. Eisenmenger's syndrome occurs when an intracardiac shunt results in severe pulmonary vascular disease, increasing right ventricular pressure causing right-to-left shunting of deoxygenated blood. The patient becomes cyanotic and experiences digital clubbing and polycythemia. Pregnancy is contraindicated in women with Eisenmenger's syndrome and associated pulmonary hypertension, which confers a maternal mortality between 5 and 50%.<sup>59,60</sup> A recent French multicenter study showed that the mortality rate was 5%, probably reflecting the fact that these women were managed at tertiary specialist centers.<sup>60</sup>

For women presenting in the first trimester of pregnancy, termination should be considered for the sake of maternal health. Later in pregnancy, termination can pose substantial risk and should be considered very carefully.

Data regarding pregnancy outcome in women with Eisenmenger's are sparse, and generally limited to case reports and small series. Rates of preterm delivery exceed 70%, typically due to iatrogenic preterm delivery.<sup>59,60</sup> Similarly, rates of small for gestational age are higher as this is related to the severity of maternal hypoxemia.

## Coarctation of the aorta

Coarctation of the aorta involves a focal narrowing of the distal aorta arch or descending aorta, typically beyond the left subclavian, and usually is diagnosed during childhood. In adults, it can be complicated by hypertension, aortic dissection, and arteritis. It is rarely an isolated lesion and may be associated with a range of cardiac defects including a bicuspid aortic valve, a ventricular septal defect, or a patent ductus arteriosus. Following repair, a woman who wishes to conceive should be evaluated for significant restenosis (gradient >20 mmHg). After surgical repair, patients often require antihypertensive medication, which should be assessed before conception. Hypertension may worsen during pregnancy and pre-eclampsia may occur. Obstetric outcomes for women with coarctation are favorable.<sup>61</sup>

## Ebstein's anomaly

Ebstein's anomaly is characterized by a malformed, elongated tricuspid valve with distal displacement of the septal leaflet of the valve into the right ventricle. In women without cyanosis or signs of heart failure, pregnancy is well tolerated. Ebstein anomaly is associated with an increased risk of preterm birth and low fetal birthweight, especially in the presence of cyanosis. Overall pregnancy outcomes are very good, especially in women who are NYHA Class 1.<sup>62</sup>

## Tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic congenital heart disease. It is a complex cardiac defect that includes a large ventricular septal defect, infundibular pulmonic stenosis, right ventricular hypertrophy and an overriding aorta. The majority of women with tetralogy of Fallot will have undergone surgical repair in infancy which may leave mild residual hemodynamic abnormalities, such as tricuspid regurgitation, pulmonic stenosis, or pulmonic regurgitation. These are all well tolerated in pregnancy. Typically the most common cardiovascular complication is that of supraventricular arrhythmia which complicates less than 10% of pregnancies.<sup>63</sup>

Women with marked tricuspid regurgitation and right ventricular volume overload, may experience right ventricular failure in pregnancy. It is important to evaluate these women with echocardiography to assess baseline pulmonary pressures and valvular abnormalities before conception. Significant pulmonary insufficiency and/or right ventricular outflow tract obstruction cause right ventricular dilatation and dysfunction. This hemodynamic abnormality is strongly associated with supraventricular and ventricular arrhythmias. There is a long-term risk for tachyarrhythmia and bradyarrhythmias postoperatively; therefore, Holter monitoring should be part of the preconception assessment and repeated during pregnancy if symptoms of palpitations, near-syncope, or syncope.

## Single ventricle

As surgical and perioperative care has improved, more women with a single ventricle are now embarking upon pregnancy. These women will have usually undergone several procedures in infancy to have a Fontan (single ventricle) circulation created. Pregnancy in women with a Fontan repair is considered to be high risk, with at least a 50% resulting

in miscarriage.<sup>64,65</sup> Women embarking upon pregnancy with low baseline oxygen saturations (<85%) seem to be at the greatest risk of miscarriage.<sup>64</sup> Furthermore, women are much more likely to have a preterm infant; a UK multicenter study showed the median gestation at delivery to be 32 weeks with many babies being born small for gestational age.<sup>64</sup> The woman with a single ventricle is at greater risk of thromboembolism; however, optimal anticoagulation in pregnancy is controversial.<sup>65</sup> Women should be individually risk assessed and considered for some form of anticoagulation or antiplatelet treatment in pregnancy. Cardiac complications are also more common in this group including arrhythmias and heart failure. A recent systematic review demonstrated that the risk of arrhythmia in pregnancy was about 8% and heart failure was 4%.<sup>65</sup> It remains unknown whether pregnancy has any adverse impact upon the Fontan repair long term.

### ***Transposition of the great arteries***

Transposition of the great arteries (TGA) accounts for 5% of all congenital heart malformations. In this condition, the aorta arises from the right ventricle and the pulmonary artery from the morphologic left ventricle. This was repaired historically by an atrial switch procedure to redirect incoming venous flow to the appropriate ventricle referred to as the Mustard or Senning procedure. More recently, women will have undergone the arterial switch procedure (Jatene) to reattach the great vessel to the appropriate ventricle and re-implantation of the coronary arteries.

After the Mustard or Senning repair, there are two clinical issues pertinent to pregnancy. First, is whether the morphologic right ventricle, acting as the systemic ventricle, can withstand the additional volume load of pregnancy and, once dilated, whether it will return to its original size and function after pregnancy. Second, arrhythmias commonly occur years after the atrial switch operation and may cause complications such as junctional rhythms with the loss of sinus mechanism, atrial fibrillation, and heart block. Normal functional capacity, sinus rhythm, and an intact repair contribute to a favorable outcome of pregnancy. A registry in the Netherlands reported 28 patients with 49 (71%) completed pregnancies.<sup>66</sup> There were 17 spontaneous miscarriages (25%). Arrhythmias occurred in 54% of the 11 patients with cardiac complications. A decline in NYHA functional class occurred in 77%, but only two patients developed heart failure. A single center UK cohort study has shown favorable maternal and neonatal outcomes in women following the arterial switch procedure undergoing pregnancy.<sup>67</sup>

Levo- or L-looped TGA (also known as congenitally corrected transposition, double discordance, or ventricular inversion) is characterized by the aorta arising from the morphologic right ventricle which receives blood from the left atrium and the pulmonary artery arising from a morphologic left ventricle. A woman may reach adulthood without known cardiac problems. Cardiac complications of L-TGA include heart block, systemic, or atrio-ventricular (A-V) valve regurgitation or systemic ventricular failure secondary to the volume load of pregnancy. A report of 22 patients having 60 pregnancies noted congestive heart failure during pregnancy related to severe systemic A-V valve regurgitation.<sup>68</sup>

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## **DISEASES INVOLVING THE AORTA**

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### **Bicuspid aortic valve**

A congenital bicuspid aortic valve (BAV) is present in approximately 1–2% of the general population. In the absence of any associated aortic dilatation pregnancy in these individuals is tolerated well. The overall risk of aortic dissection is increased in individuals with a bicuspid valve, but risk of aortic dissection in pregnancy remains very low. Pregnancy in women with a dilated aorta >50 mm and a bicuspid valve, however, is discouraged because of the increased risk of dissection.<sup>2</sup> A single center American study showed that in following women with a BAV pregnancy there appeared to be a significant rate of progressive aortic enlargement which requires long-term follow-up.<sup>69</sup>

### **Marfan syndrome**

Marfan syndrome (MFS), a disorder of fibrillin is an autosomal dominantly inherited disorder. Diagnosis is made by the modified Ghent criteria.<sup>70</sup> Cardiovascular manifestations of MFS include aortic dilatation as well as aortic and mitral regurgitation. Aortic disease is responsible for significant morbidity that is associated with the disorder, pregnancy has been shown to increase the risk of dissection in women with MFS.<sup>71</sup> Furthermore it appears that pregnancy may

accelerate aortic root growth.<sup>72</sup> Predicting which women with MFS are at greater risk of dissection in pregnancy remains challenging, a recent UK multicenter study failed to show any factors that were associated with increased risk of dissection in pregnancy.<sup>73</sup>

Most dissections appear to be type B (i.e. a tear in the descending aorta compared with type A dissections which affect the ascending aorta) and typically occur in the third trimester or postpartum.<sup>73</sup> Diagnosis of dissection can be made via echocardiography, cardiac CT scan or by MRI. The choice of imaging modality will depend upon the setting in which the patient presents. In the emergency setting, CT is commonly performed. CT has excellent specificity and sensitivity, particularly when compared to echocardiography.<sup>74</sup> MRI scanning is the most accurate technique for diagnosing dissection, and it is particularly good at assessing the descending aorta. However, MRI is not easily available in all hospital settings. A type B dissection is usually managed medically with aggressive antihypertensive therapy including beta-blockers to reduce both blood pressure and heart rate which will reduce further shear stress. Stenting can be undertaken in cases of aortic rupture. The timing of delivery in women with a known aortic dissection should be individualized, but delivery is probably best undertaken by planned cesarean in a cardiac theater section permitting full cardiothoracic support around 37 weeks' gestation.

Recent guidance from the ESC suggests that women should be monitored with echocardiography during pregnancy, at least once each trimester, to evaluate aortic root size, and that betablockers may be considered, but the data to support their use in pregnancy to reduce the risk of dissection are unproven.<sup>2</sup> ESC guidance also suggests that women with an aortic root of greater than 45 mm should be advised against pregnancy because of risks regarding dissection.<sup>2</sup> Vaginal delivery is encouraged for the majority of women with MFS, except in cases of aortic root dilatation where cesarean section may need to be considered.<sup>2</sup> Women with previous aortic root replacement may have an uncomplicated pregnancy, although there are some reports suggesting that these women have a greater risk of dissection in pregnancy.<sup>75</sup>

## Ischemic heart disease

Coronary artery disease (CAD) is rare in women of childbearing age. However, coronary disease may be caused by long-standing diabetes mellitus, hypercholesterolemia, or smoking. A recent systematic review demonstrated that women with underlying CAD are at greater risk of cardiac and obstetric complications in pregnancy.<sup>76</sup> Cardiac events occurred in approximately 9% of women, with only a fifth of women having an uncomplicated pregnancy. A UK cohort study was reassuring in that it showed no maternal deaths in pregnancy in a cohort of 79 women with CAD, but approximately 15% of women developed pre-eclampsia despite the use of aspirin in pregnancy.<sup>77</sup>

Women presenting with new suspected coronary artery disease in pregnancy should be managed in the same way as the non-pregnant individual. A marginal rise in cardiac troponins may be associated with pre-eclampsia but not at the threshold to diagnose acute coronary syndrome.<sup>78</sup> Women with ST segment elevation on 12-lead ECG and elevated troponins require urgent percutaneous coronary intervention. This is best performed through a radial approach with appropriate abdominal shielding.<sup>26</sup> When coronary stents are required both bare metal stents and drug eluting stents may be required. The treating interventionalist is best placed to decide which is most appropriate in each case. In rare cases coronary artery bypass surgery may be indicated. Medical management of symptomatic CAD during pregnancy should be similar to that in the non-pregnant patient. Beta-adrenergic blockers, specifically the selective agents, are indicated for ischemic disease, hypertension, and arrhythmia and should be continued throughout pregnancy. Nitrates should be utilized for symptomatic control of angina because calcium-channel blockers, such as nifedipine, verapamil, diltiazem, and amlodipine, may also be used, especially if there is vasospastic component to the angina. Low-dose aspirin is well tolerated in pregnancy. Women can be reassured regarding the use of clopidogrel, but this should be stopped at least a week prior to planned delivery, as it is associated with an increased risk of maternal hemorrhage.<sup>26</sup> Women with known CAD who are taking statins prior to pregnancy are advised to stop these medications because of a paucity of safety data in pregnancy. However, statins have been approved for use in clinical trials evaluating their impact upon early onset pre-eclampsia. Women taking angiotensin converting enzyme inhibitors (ACEI) should be advised to stop taking these during pregnancy because of possible teratogenesis and fetal renal impairment later in pregnancy.

Spontaneous coronary artery dissection (SCAD) describes a tear that occurs in the wall of the coronary artery typically



causing an accompanying hematoma. It is one of the commonest causes of pregnancy-associated myocardial infarction, and yet the etiology remains poorly understood.<sup>79</sup> Women with pregnancy-associated SCAD typically present with ST elevation changes, and are more likely to present with symptoms of heart failure. SCAD is diagnosed on coronary angiography and can be treated conservatively, medically or with coronary stenting.<sup>79</sup> Recurrence rates in pregnancy are reported to approach 20%.<sup>77</sup>

## Myocardial disease

### *Peripartum cardiomyopathy*

Peripartum cardiomyopathy (PPCM) is defined as an idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular systolic dysfunction towards the end of pregnancy or in the months following delivery where no other cause can be identified. The ejection fraction is nearly always reduced below 45%. The reported incidence varies between 1 in 300 and in 15000 live births.<sup>80,81</sup> Risk factors include multiple pregnancy, pre-eclampsia, maternal age, pre-existing hypertension and black race. Management is supportive with some experts recommending cessation of breast feeding and the use of bromocriptine.<sup>2,82</sup> Approximately 50% of women will recover left ventricular function. Data from a prospective North American study showed the maternal mortality to be approximately 5%.<sup>83</sup> Women who wish to consider pregnancy after previous PPCM require careful assessment and counselling. Data regarding recurrence risk of PPCM of women with previous PPCM show that the relapse rates vary between 20 and 50% with women entering pregnancy with impaired left ventricular function having worse outcomes.<sup>84</sup>

### *Other cardiomyopathies*

Dilated cardiomyopathy (DCM) encompasses a number of disorders resulting in left ventricular dilatation and dysfunction. Approximately 50% are idiopathic, of which more than 20% are hereditary. About 40% of genetic causes of DCM have been identified. A Canadian study of 36 pregnancies in 32 women with DCM showed that cardiac events were common in pregnancy and were related to NYHA functional class.<sup>85</sup> In the short-term pregnancy does not appear to adversely impact the natural course of DCM. Women with DCM should have regular (at least 4 weekly) clinical follow-up in pregnancy. Women with DCM should be considered as high risk and seen in a combined obstetric cardiac clinic with regular follow-up and echocardiographic assessment during pregnancy. Beta-blockers and diuretics may be required for ongoing management but ACEI and angiotensin receptor blockers should be stopped.

In hypertrophic cardiomyopathy, the heart muscle is thickened and has limited compliance. It may manifest as asymmetric hypertrophy of the septum or as a global, left ventricular hypertrophy. Data from the European Registry on Heart Disease and Pregnancy showed good maternal outcomes, with no maternal deaths, although cardiovascular events occurred in 23% of women.<sup>86</sup> Preterm birth occurred in almost 25% of pregnancies and 16% of infants were born small for gestational age. A further meta-analysis of 408 pregnancies in 237 women showed maternal mortality to be 0.9%.<sup>87</sup> Caution in these women should be noted with regional analgesia and obstetric hemorrhage because these women do not respond well to profound hypotension. Left ventricular non-compaction (LVNC) is a form of familial cardiomyopathy due to intrauterine arrest of compaction of the loose interwoven meshwork. There are limited reports of pregnancy and LVNC but certainly these women merit specialist combined care during pregnancy with regular follow up in pregnancy.

## Cardiac transplantation

There is now more information regarding pregnancy post solid organ transplant owing to data derived from the National Transplant Registry Pregnancy.<sup>88</sup> They collate data on all solid organ transplants and subsequent pregnancies. Women with heart transplants in pregnancy appear to have a higher rate of rejection when compared to kidney transplants, but lower rates of preterm birth. Late maternal deaths have been reported following pregnancy in women with heart transplants.<sup>89</sup>

These women require multidisciplinary management during pregnancy and meticulous monitoring of their immunosuppressive treatment. Pregnancy does not appear to increase the chance of rejection per se. Rates of gestational hypertension and pre-eclampsia are much greater in parturients with a heart transplant compared to normal women. Women should be reassured that they can breastfeed on most immunosuppressive drugs.



## CONCLUSION

In conclusion, preconception counselling and multidisciplinary care facilitates successful pregnancy outcomes in women with pre-existing cardiovascular disease. Although some women with severe, debilitating forms of cardiovascular disease may be further compromised by the hemodynamic changes of pregnancy, most can sustain pregnancy. Successful outcomes often require a multidisciplinary strategy formulated by the obstetrician, cardiologist, obstetric physician, neonatologist, geneticist and anesthesiologist, who can anticipate, treat, and even prevent cardiovascular, obstetric and neonatal complications.

## PRACTICE RECOMMENDATIONS

- All women with known cardiac disease should have access to preconception counselling to discuss the risks of pregnancy to her and her baby.
- Women with pre-existing cardiac disease should undergo cardiac evaluation prior to pregnancy, the extent of which will be determined by the nature of the underlying diagnosis
- Where possible women with cardiac disease should be cared for in a combined obstetric/cardiac clinic. The frequency of monitoring and follow-up during pregnancy will largely be dictated by their underlying condition.
- Underlying cardiac disease per se is rarely a reason to recommend delivery by cesarean section.
- Women with the most severe forms of cardiac disease may need to be advised against pregnancy because of significant maternal risks.

### *Relevant Guidelines for Heart Disease and Pregnancy*

- European Society of Cardiology Management of Cardiovascular Disease in Pregnancy 2018
- Management of Pregnancy in Patients With Complex Congenital Heart Disease: A Scientific Statement for Healthcare Professionals From the American Heart Association

## CONFLICTS OF INTEREST

*The authors of this chapter declare that they have no interests that conflict with the contents of the chapter.*

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