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Cardiac disease in pregnancy

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ABSTRACT – Cardiac disease is the leading cause of maternal mortality in the UK. The major causes of cardiac deaths in pregnancy include cardiomyopathy, myocardial infarction, ischaemic heart disease and dissection of the thoracic aorta. With increasing numbers of migrant women in the UK, rheumatic heart disease in pregnancy has also re-emerged. Women with uncorrected congenital heart disease and those who have undergone corrective or palliative surgery may have complicated pregnancies. Women with metal prosthetic valves face difficult decisions regarding anticoagulation in pregnancy and have an increased risk of haemorrhage. Not all women with significant heart disease are able to meet the increased physiological demands of pregnancy. The care of pregnant women with heart disease thus requires a multidisciplinary approach, involving obstetricians, cardiologists and anaesthetists. This allows appropriate surveillance of maternal and fetal well-being, as well as planning and documentation of the management of elective and emergency delivery. This review discusses common cardiac conditions encountered in pregnancy and their antenatal and intrapartum management.

KEY WORDS: cardiac disease, multidisciplinary team, obstetrics, pregnancy

Introduction

Cardiac disease remains the leading cause of maternal death in the UK. The major cardiac causes in the latest triennium of the *Centre for Maternal and Child Enquiries* (CMACE)'s confidential enquiry into maternal death included myocardial infarction (MI; mostly related to ischaemic heart disease), dissection of the thoracic aorta and cardiomyopathy (most commonly peripartum).[1] Heart disease complicates 0.2–4% of all pregnancies in the western world.[2] The UK Obstetric Surveillance System (UKOSS) study of acute MI in pregnancy estimated an incidence of 0.7 cases per 100,000 maternities (95% confidence interval (CI) 0.4 to 1.0).[3] The number of pregnant women who have had corrective or palliative surgery for congenital cardiac diseases and to fit pacemakers and prosthetic heart valves is increasing. In addition, the incidence of acquired heart disease is increasing due to older age at first pregnancy and a higher prevalence of cardiovascular risk factors such as hypertension, diabetes and obesity. Rheumatic valvular disease comprises 56–89% of all cardiovascular disease in pregnancy in developing countries but is relatively rare in the UK.[4]

Women with risk factors for adverse cardiac events should be managed and counselled by a multidisciplinary team, including cardiologists with expertise in pregnancy, obstetricians with expertise in cardiac disease, fetal medicine specialists, obstetric anaesthetists and paediatricians. This counselling ideally should occur before pregnancy. Once a woman is pregnant, a multidisciplinary plan should be documented for both elective and emergency delivery.

This review briefly describes the common cardiac conditions encountered in pregnancy and issues around their management. The haemodynamic changes and common examination findings in pregnancy and useful cardiac investigations are outlined briefly in Tables 1 and 2, respectively.

General considerations in pregnant women with heart disease

Potential adverse outcomes in the mother include stroke, arrhythmia, pulmonary oedema and death. For the fetus, growth restriction and fetal loss are more common. Table 3 summarises predictors of adverse outcomes. Table 4 summarises common cardiac conditions and risk estimates in pregnancy based on the World Health Organization (WHO)'s criteria. Potential adverse effects of drugs acting on the uterus in women with cardiac disease are summarised in Table 5.

Congenital heart disease

Asymptomatic women with simple defects or previously repaired defects tolerate pregnancy well. Women with congenital heart disease are at increased risk of having babies with congenital heart defects and so should be offered specialist fetal cardiac scans between 18 and 20 weeks. The most common congenital heart diseases in pregnant women, which account for nearly 60% of cases, are patent ductus arteriosus, atrial septal defect and ventricular septal defect.

Acyanotic congenital heart disease

Atrial septal defect

Pregnancy is well tolerated by most women with an unrepaired atrial septal defect (ASD). Paradoxical embolism and pulmonary

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Table 1. Haemodynamic changes in pregnancy.[5–7]

Haemodynamic parameter	Pregnancy	Clinical implication	Normal findings
Blood flow	↑	• Nose bleeds common • Baseline serum creatinine lower in pregnancy	• Bounding/collapsing pulse • Prominent non-displaced apical pulse • Ejection systolic murmur • Loud first heart sound • Third heart sound • Venous hum • Mammary souffle • Relative sinus tachycardia (10–20 bpm) • Ectopic beats • Peripheral oedema • Warm/erythematous extremities • Elevated JVP in late pregnancy
Blood volume (plasma and RBC)	↑	• Physiological anaemia in pregnancy • Higher risk of cardiac failure in multiple pregnancy	
Systemic vascular resistance	↓	• Risk of maternal fetal compromise in women with fixed cardiac outputs (stenotic lesions) • Sinus tachycardia towards end of pregnancy	
Stroke volume	↑		
Cardiac output	↑		
Heart rate	↑		
Blood pressure	↓		
Pulmonary capillary wedge pressure	↔	• Increased susceptibility to pulmonary oedema	
Colloid oncotic pressure	↓		
Central venous pressure	↔		
Maternal oxygen consumption	↑	• Tendency to ischaemia in pregnant women with cardiac disease	

bpm = beats per minute; RBC = red blood cell; JVP = jugular venous pressure.

hypertension are rare and arrhythmias uncommon in women younger than 40 years.[11] Mitral regurgitation caused by mitral leaflet prolapse develops in up to 15% of cases of uncorrected ASD. No problems are anticipated during labour, but acute blood loss is poorly tolerated and can cause a large increase in left-to-right shunting; precipitous falls in left ventricular output, blood pressure and coronary blood flow; and even cardiac arrest.[5]

Ventricular septal defect and patent ductus arteriosus

Ventricular septal defect (VSD) and patent ductus arteriosus (PDA) are well tolerated in pregnancy unless they are large or complicated by pulmonary vascular disease. Pre-pregnancy evaluation of the presence of a (residual) defect, cardiac dimensions and estimation of pulmonary pressures are recommended.[12]

Pulmonary stenosis

Pulmonary stenosis (PS) is well tolerated, although severe cases may precipitate right-sided heart failure, tricuspid regurgitation or atrial arrhythmia. Women with a pre-pregnancy peak-to-peak catheter gradient >50 mmHg or symptoms should be considered for balloon valvuloplasty or surgery before conception.[6]

Aortic stenosis and bicuspid aortic valves

In women of childbearing age, the main cause of aortic stenosis (AS) is congenital bicuspid aortic valves (BAoVs). Patients can be asymptomatic, even with

Table 2. Cardiac investigations in pregnancy.[5–7]

Investigations	Use in pregnancy
Troponin	• Not affected by pregnancy • Valid test for myocardial ischaemia
CK	• Normal or lower in pregnancy • Increases in labour
ECG	– Sinus tachycardia – 15° left-axis deviation – T-wave inversion (leads III and aVF) – Non-specific ST changes, eg depression – Supraventricular and ventricular ectopic beats – Small Q waves
Holter monitoring	• Usually indicated in syncopal/presyncopal episodes or palpitations
Echocardiography	• Preferred screening method to assess cardiac function
Transoesophageal echocardiography	• Rarely required but relatively safe in pregnancy
Chest X-ray	• No contraindication
MRI and CT	• Safe in pregnancy
Cardiac MRI	• Avoided in first trimester but can performed in second and third trimesters
Electrophysiological studies	• Usually postponed until after pregnancy
Cardiac catheterisation	• Should not be withheld if clinically indicated – eg, acute coronary syndromes • Shielding the gravid uterus is recommended
Exercise test	• Usually performed prior to pregnancy • Semi-recumbent cycle ergometry or even treadmill test may be performed in asymptomatic pregnant women with suspected cardiac disease to reach 80% of expected heart rate
Ventilation/perfusion scan	• Not contraindicated in pregnancy • Ventilation portion can be omitted in pregnant women if chest X-ray is normal

CK = creatine kinase; CT = computed tomography; ECG = electrocardiography; MRI = magnetic resonance imaging.

severe AS. Significant obstruction results if the aortic valve area is smaller than 1 cm^2 or if the non-pregnant mean gradient across the valve is >50 mmHg. Women with BAoV are at increased risk of aortopathy (and are therefore at higher risk of dissection) and arrhythmias. The aortic root and ascending aortic diameter should therefore be assessed before and during pregnancy. In such women, surgery before pregnancy should be considered if the aortic root is >50 mm in diameter. In pregnant women with severe AS, heart failure occurs in about 10% and arrhythmias in 3–25%.[13] All women with symptomatic AS (chest pain, syncope or pre-syncope) or asymptomatic AS but impaired left ventricular function on a pathological exercise test (without an appropriate increase in blood pressure or the development of ST- or T-wave changes) should be counselled against pregnancy, and valvuloplasty or surgery should be performed before pregnancy.

Pregnancy is usually associated with a progressive increase in the gradient across the aortic valve on Doppler ultrasound as the left ventricular stroke volume increases. A falling or static gradient therefore may be falsely reassuring. Medical treatment (with β blockers and diuretics) and restricted activities are indicated for patients who develop signs or symptoms of heart failure during pregnancy. If medical treatment fails, either balloon aortic valvotomy or, rarely, valve replacement after early delivery by caesarean section are options.[14]

Table 3. Predictors of adverse maternal and neonatal outcome in pregnancy with cardiac disease.[4,13]

Predictors for a maternal cardiac event	Predictors for adverse neonatal events
• Prior cardiac event (heart failure, transient ischaemic attack or stroke) or arrhythmia • Baseline NYHA class >II or cyanosis* • Left heart obstruction (mitral valve area <2 cm^2, aortic valve area <1.5 cm^2 or peak ventricular outflow tract gradient >30 mmHg by echocardiography) • Reduced systemic left ventricular function (ejection fraction <40%)	• NYHA class >II or cyanosis during the baseline prenatal visit* • Maternal left ventricular obstruction • Maternal smoking • Multiple gestation

NYHA = New York Heart Association.

*Women with cyanosis (oxygen saturation <80%) have an increased risk of fetal growth restriction, fetal loss and thromboembolism secondary to the reactive polycythaemia. Their chance of a live birth has been quoted to be less than 20%.[13]

Table 4. Modified World Health Organization (WHO) classification of maternal cardiovascular risk.[11,22]

Risk category	Description	Conditions
I	• No detectable increased risk in maternal mortality and no/mild increase in morbidity	• Uncomplicated, small or mild PS, PDA or mitral valve prolapse • Successfully repaired simple lesions (ASD, VSD, anomalous pulmonary venous drainage) • Atrial or ventricular ectopic beats, isolated
II	• Small increased risk of maternal mortality and moderate increase in morbidity	• Unoperated ASD or VSD • Repaired TOF • Most arrhythmias
II/III		• Mild left ventricular impairment • HCM • Native or tissue valvular heart disease not considered WHO class I or IV • Marfan's syndrome without aortic dilatation • Aorta <45 mm in aortic disease associated with BAoV • Repaired CoA
III	• Significantly increased risk of maternal mortality or severe morbidity • Expert counselling required • If pregnancy is decided upon, intensive specialist, cardiac and obstetric monitoring needed throughout pregnancy, childbirth and puerperium	• Mechanical valve • Systemic right ventricle • Fontan circulation • Cyanotic heart disease (unrepaired) • Other complex congenital heart disease • Aortic dilatation 40–45 mm in Marfan's syndrome • Aortic dilatation 45–50 mm in aortic disease associated with BAoV
IV	• Extremely high risk of maternal mortality or severe morbidity; pregnancy contraindicated • If pregnancy occurs, termination should be discussed • If pregnancy continues, care as for class III	• Pulmonary arterial hypertension of any cause • Severe systemic ventricular dysfunction (LVEF <30%, NYHA class III–IV) • Previous PPCM with any residual impairment of left ventricular function • Severe MS, severe symptomatic AS • Marfan's syndrome with aorta dilation >45 mm • Aortic dilatation >50 mm in aortic disease associated with BAoV • Native severe CoA

AS = aortic valve stenosis; ASD = atrial septal defect; BAoV = bicuspid aortic valve; CoA = coarctation of the aorta; HCM = hypertrophic cardiomyopathy; LVEF = left ventricular ejection fraction; MS = mitral stenosis; NYHA = New York Heart Association; PPCM = peripartum cardiomyopathy; PS = pulmonary stenosis; PDA = patent ductus arteriosus; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

Coarctation of the aorta

Most cases of coarctation of the aorta (CoA) encountered in pregnancy will already have been surgically corrected, although residual narrowing is not uncommon; CoA may also be first diagnosed during investigation for hypertension in pregnancy. Women with unrepaired native CoA and those with repaired CoA but residual hypertension or aortic aneurysms have an increased risk of aortic rupture and rupture of an associated cerebral aneurysm during pregnancy and delivery. Any narrowing or pre- or post-stenotic dilatation or aneurysm formation should be assessed with magnetic resonance imaging prior to pregnancy.[7]

Optimal treatment of hypertension (ideally with β blockers) is necessary, although aggressive treatment should be avoided. Strenuous exercise should be avoided, as adequate blood pressure control may not be maintained during exercise, increasing the risk of cerebral haemorrhage or aortic dissection. Women with CoA are at increased risk of hypertensive disorders of pregnancy.[15]

Percutaneous intervention for re-CoA is possible but associated with a higher risk of aortic dissection in pregnancy. The use of covered stents may reduce this risk. Normal delivery is usually possible, although severe CoA would warrant a shortened second stage of delivery.

Marfan's syndrome

About 80% of patients with Marfan's syndrome have some cardiac involvement, commonly mitral valve prolapse and regurgitation. Patients with Marfan's syndrome and a normal aortic root diameter have a 1% risk of aortic dissection or other serious cardiac complications during pregnancy.[7] As such, pregnancy, even in the absence of pre-existing disease, increases the susceptibility to aortic dissection due to haemodynamic and hormonal changes. Dissection occurs most often in the last trimester of pregnancy (50%) or the early postpartum period (33%).[16] Women with progressive aortic root dilatation and aortic root dimension >4 cm and those with a family history of dissection or sudden death, even in the absence of a dilated aortic root, are at increased risk of aortic rupture or dissection. Women with aortic roots >4.6 cm should be advised to delay pregnancy until after repair or root replacement. Outcome of pregnancy is usually good in women with minimal cardiac involvement and an aortic root <4 cm in pregnancy.[7]

Management includes monthly echocardiography to assess the aortic root in those with cardiac involvement and β blockers for hypertension or aortic root dilatation. Vaginal delivery for those with stable aortic root is possible, but elective caesarean section with regional anaesthesia is recommended if the aortic root is enlarged or dilating.

Cyanotic heart disease

Any uncorrected or inadequately corrected congenital heart disease that is associated with cyanosis is associated with an increased risk of miscarriage, poor fetal growth, prematurity and a small-for gestation fetus,[8] especially in women with resting arterial saturation <85% or haemoglobin >18 g/dl and haematocrit >55%.

Tetrology of Fallot

The association of severe right ventricular outflow tract obstruction with a large subaortic VSD and overriding aorta causes right ventricular hypertrophy and right-to-left shunting with cyanosis. Pregnancy is usually well tolerated in uncorrected cases, but subcutaneous low molecular weight heparin (LMWH) should be given to prevent venous thrombosis and paradoxical embolism. However, most women will have had previous surgical correction and will do well in pregnancy, although pulmonary regurgitation from previous correction of right ventricular outflow tract obstruction may lead to right ventricular failure.

Pulmonary hypertension

Pulmonary vascular disease, whether secondary to Eisenmenger's syndrome or lung or connective tissue disease (for example, scleroderma) or due to idiopathic arterial pulmonary hypertension, is extremely dangerous in pregnancy, with maternal mortality of 25–40%.[14] In cases of unplanned pregnancy, elective termination carries a 7% risk of death.

Inability to increase pulmonary blood flow in pregnancy leads to refractory hypoxaemia. If the systolic pulmonary pressure – estimated by measuring regurgitant jet velocity across the tricuspid valve on Doppler ultrasound – is thought to indicate pulmonary hypertension, a specialist cardiac opinion is recommended. Women with pulmonary hypertension who have left-to-right shunts are at lesser risk and may do well during pregnancy as the increased right-sided pressures are related to volume rather than increased pulmonary vascular resistance in these cases, although there is still a potential risk of developing pulmonary vascular disease. Management includes drugs such as sildenafil and bosentan, elective admission for bed rest, oxygen, thromboprophylaxis with LMWH and serial monitoring of fetal growth. Most fatalities occur during delivery or during the first week after birth. Nebulised or intravenous prostacyclin can be used to prevent pulmonary vasoconstriction, although resuscitation is rarely successful when sudden deterioration occurs. All cases should be discussed with a centre specialising in pulmonary hypertension.

Postoperative congenital heart disease

Survivors of neonatal palliative surgery for complex congenital heart disease need individual assessment. Following the Fontan operation for tricuspid atresia or transposition with pulmonary stenosis, the left ventricle provides the pump for both the systemic and pulmonary circulations. Increases in venous pressure can lead to hepatic congestion and gross oedema, but pregnancy can be successful. Anticoagulation with LMWH and optimal hydration peripartum are recommended to enable adequate left ventricular preload.[14]

Table 5. Potential adverse effects of drugs acting on the uterus when used in women with cardiac disease.[11,23]

Drugs	Primary use	Mechanism of action	Potential side effects	Comments for use in women with cardiac disease
Uterotonics				
Oxytocin	• Management of PPH • Augmentation of labour	• Increase in frequency and amplitude of contractions	• Water retention • Hypotension • Tachycardia • ECG changes	• Avoid bolus • Dilute in ≥20 ml normal saline
Ergometrine	• Management of PPH	• Increase in frequency and amplitude of contractions at lower doses • Basal tone of uterus increases at higher doses	• Severe hypertension • Coronary vasospasm • Elevated PA pressures • Pulmonary oedema	• Avoid in: – Pre-eclampsia – coronary artery disease – aortopathies – aneurysms
Carboprost	• Management of PPH	• PGF2α agonist	• Vasoconstriction • Bronchospasm	• Avoid in: – asthma – shunt lesions – elevated PA pressures – single ventricle – pulmonary hypertension
Misoprostol	• Management of PPH • Cervical priming for medical termination of pregnancy	• PGE1 agonist	• Fever • Diarrhoea • Nausea • Cramps • Headache	• Well tolerated but less successful than oxytocin or ergometrine
Prostin	• Induction of labour	• PGE2 agonist		• Usually well tolerated
Tocolytics				
Ritodrine	• To stop preterm labour, eg threatened preterm labour, uterine hyperstimulation	• β-agonist	• Pulmonary oedema • Profound ketoacidosis in women with type 1 diabetes and those with vomiting	• Avoid in: – cardiomyopathy – pre-eclampsia – impaired LV function • Rarely used now
Salbutamol Terbutaline			• Headache • Nausea • Dizziness • Flushing • Palpitations • Tremor • Arrhythmias • Myocardial ischaemia • Paradoxical bronchospams	• Use with caution in: – cardiac failure – arrhythmias – known IHD – HCM
Nifedipine		• Calcium channel blocker	• Headache • Hypotension • Palpitations • Dizziness • Vasodilation • Syncope	• Avoid in: – stenotic lesions such as AS, MS and CoA – cardiac shunt lesions such as VSD and ASD – HCM • Contraindicated in unstable angina and recent MI
Atosiban		• Oxytocin antagonist	• Nausea • Vomiting • Tachycardia • Hypotension • Headache • Dizziness • Hot flushes • Hyperglycaemia	• Use with caution in: – preeclampsia

AS = aortic valve stenosis; ASD = atrial septal defect; CoA = coarctation of the aorta; ECG = electrocardiogram; HCM = hypertrophic cardiomyopathy; IHD = ischaemic heart disease; LV = left ventricular; MI = myocardial infarction; MS = mitral stenosis; PA = pulmonary arterial; PGE = prostaglandin; PGE = prostaglandin; PPH = postpartum haemorrhage; VSD = ventricular septal defect.

Acquired heart disease

Mitral valve prolapse

Pregnancy is generally very well tolerated in isolated cases of mitral valve prolapse.[6]

Rheumatic heart disease

Mitral stenosis

Mitral stenosis (MS) remains the most common important pre-existing heart condition in pregnancy worldwide. Asymptomatic women with MS may deteriorate in pregnancy, and a previously uneventful pregnancy course does not preclude deterioration in a subsequent pregnancy, because degeneration of the valve may lead to increased stenosis over time. Mitral stenosis may be missed during routine antenatal examination because the murmur is low pitched, usually quiet, diastolic and submammary. Women may deteriorate secondary to tachycardia, arrhythmias or the increased cardiac output of pregnancy. Pulmonary oedema may also be precipitated by increased volume (for example, during the third stage of labour or following injudicious use of intravenous fluid therapy). The risks are increased in women with severe MS (mitral valve area <1.5 cm^2), with moderate or severe symptoms prior to pregnancy, and with a diagnosis late in pregnancy.[7,14]

Women with severe MS are advised to delay pregnancy until after balloon dilation, valvotomy or replacement. Beta blockers should be given to maintain heart rate <90 bpm, and diuretics can be given as indicated. Pulmonary oedema should be managed as with non-pregnant women. Digoxin should be given only in women with concurrent atrial fibrillation. If medical therapy fails, balloon mitral valvotomy may be used in pregnancy, although open surgery on the mitral valve should be avoided, if possible, until after delivery.

Regurgitant heart disease

Patients with mitral or aortic regurgitation tolerate pregnancy much better than patients with valvular stenosis.[6]

Mechanical heart valves

Women with mechanical heart valves require lifelong anticoagulation, including during pregnancy, because of the increased risk of thrombosis. Warfarin is associated with a risk of embryopathy between 6 and 12 weeks of gestation and a dose-dependent risk of fetal intracerebral haemorrhage, miscarriage and stillbirth despite maternal international normalised ratio (INR) being within the therapeutic range. Low molecular weight heparins have a better safety profile in pregnancy, as long as anti-Xa levels are monitored closely (keeping peak levels at 0.8–1.2 IU/ml), with appropriate dose adjustments and good compliance with twice-daily injections, together with low-dose aspirin. However, a risk of valve thrombosis and bleeding in the mother remains even with optimal management.[17] The choice of anticoagulant regimen depends on the type, size, position and number of mechanical valves; the dose of warfarin needed to maintain therapeutic INR; any previous history of embolic events or arrhythmias; and maternal preference after informed counselling.[14]

Myocardial infarction

Pregnancy increases the risk of MI,[1] and it is increasingly encountered in pregnant and postpartum women.[2] Pregnant women may present with a preceding history of typical angina or atypical epigastric pain; nausea; dizziness; or pain in the chest, neck or left arm. Spontaneous coronary artery dissection and thrombosis are more common in pregnancy, usually during late pregnancy or around the time of delivery. Coronary ischaemia may also be associated with cocaine abuse, an embolic source or infective endocarditis. The risk is increased in older multigravid women, smokers and obese women and in those with diabetes, hypertension, hypercholesterolaemia and a family history of coronary artery disease. Troponin I is not affected by pregnancy and should be requested along with serial electrocardiograms in women in whom acute coronary syndrome (ACS) is suspected. The management of ACS is the same as for non-pregnant women. Coronary angiography is not contraindicated in pregnancy, and intravenous and intracoronary thrombolysis, percutaneous transluminal coronary angioplasty and stenting have all been performed successfully in pregnancy. Both aspirin and β blockers are safe in pregnancy. If clopidogrel is indicated, it may be used in pregnancy; however, it must be stopped before delivery, so careful discussion regarding the type of stent insertion must occur to allow for the fact that treatment with clopidogrel will need to be interrupted. Glycoprotein IIb/IIIa inhibitors are normally avoided, and statins should be discontinued for the duration of pregnancy.[14]

Cardiomyopathy

Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) may be first diagnosed in pregnancy when a systolic murmur leads to electrocardiographic and echocardiographic studies. Most patients are asymptomatic and do well. Shortness of breath, chest pain, dizziness and syncope can be treated with β blockers. Non-sustained ventricular tachycardia on 24-hour tape is one of the risk factors for sudden death. In women with HCM, hypotension (such as may occur after epidural blockade) or hypovolaemia (for example, resulting from postpartum haemorrhage) may cause left ventricular outflow obstruction and should be avoided.[7]

Peripartum cardiomyopathy

Peripartum cardiomyopathy (PPCM) is a pregnancy-specific condition[5,18] defined as idiopathic cardiomyopathy presenting with heart failure secondary to left ventricular systolic dysfunction

toward the end of pregnancy or in the months following delivery, for which no other cause of heart failure is found and the following echocardiographic criteria are demonstrated:

- left ventricular ejection fraction <45%
- fractional shortening <30%
- left ventricular end-diastolic dimension >2.7 cm/m^2.

Peripartum cardiomyopathy does not differ clinically from dilated cardiomyopathy. Diagnosis should be suspected in peripartum patients with breathlessness, tachycardia or signs of heart failure when no other cause for heart failure is evident. Pulmonary oedema is often a major feature. Chest X-ray shows an enlarged heart, with pulmonary congestion or oedema and bilateral pleural effusions. Systemic embolism from mural thrombus may precede the onset of ventricular arrhythmias or development of clinical heart failure, and pulmonary embolism may further complicate the clinical picture.

Management includes elective delivery (if antenatal), thromboprophylaxis and conventional treatment of heart failure including diuretics, vasodilators (hydralazine and/or nitrates), cardioselective beta-blockers (bisoprolol) or beta-blockers with arteriolar vasodilating action (carvedilol), digoxin and inotropes and, after delivery, angiotensin-converting enzyme inhibitors.[5,18] Critically ill women may need intubation, ventilation and monitoring, with use of inotropes and an intra-aortic balloon pump or ventricular assist device. Heart transplantation may be the only chance of survival in severe cases. About 50% of women make a full recovery, with left ventricular function returning to normal in 23–41%. Women in whom left ventricular function and size do not return to normal within six months are at significant risk of worsening heart failure and death or recurrent PPCM in the next pregnancy. The mortality from PPCM varies worldwide but is about 30%.[18]

Arrhythmias

Atrial and ventricular premature complexes are common in pregnancy. The arrhythmia encountered most commonly in pregnancy is supraventricular tachycardia (SVT).[19] First-onset of SVT (accessory pathway mediated or atrioventricular nodal re-entrant) is rare in pregnancy, but exacerbation of symptoms is common in pregnancy. Propranolol, verapamil and adenosine (preferred) can be used for acute termination of SVT or for those who do not respond to vagal manoeuvres (50%). For prevention, β blockers or verapamil may be used. Flecainide is safe and is used in the treatment of fetal tachycardias. Propafenone and amiodarone should be avoided. Temporary and permanent pacing, cardioversion and automatic implantable defibrillators (AICD) are also safe in pregnancy. The latter device is usually inactivated during caesarean section, as the AICD may misinterpret diathermy as ventricular fibrillation.

Endocarditis prophylaxis

Infective endocarditis (IE) is rare in pregnancy but threatens the life of both mother and child. Treatment is essentially the same as outside pregnancy, with emergency valve replacement if indicated. The baby should be delivered, if viable, before the maternal operation. The current recommendations from the National Institute for Health and Clinical Excellence (NICE) in the UK are that antibiotic prophylaxis against IE is not required for childbirth.[20] The British Society for Antimicrobial Chemotherapy has recommended antibiotic cover only for patients deemed to be at high risk of developing IE (such as those with previous IE) and for those who have the poorest outcome if they develop IE (such as those with cyanotic congenital heart disease).[21] When antibiotic prophylaxis is used, it should be 2 g amoxicillin intravenously plus 120 mg gentamicin intravenously at the onset of labour or ruptured membranes or prior to caesarean section, followed six hours later by 500 mg amoxicillin orally (or intramuscularly or intravenously depending on the patient's condition); intravenous 1 g vancomycin or 400 mg teicoplanin should be used in women with penicillin allergy.

Cardiac arrest

Cardiac arrest should be managed according to the same algorithm for non-pregnant patients.[1] Pregnant women (especially those in advanced pregnancy) should be 'wedged' (left lateral) to relieve any obstruction to venous return from pressure of the gravid uterus on the inferior vena cava. If cardiopulmonary resuscitation is required, the pelvis can be tilted while keeping the torso flat to allow external chest compressions. Emergency caesarean section may be required to aid maternal resuscitation.[22]

Genetic and pre-pregnancy counselling and contraception

When available, genetic counselling should be offered in women with congenital heart disease, as the risk to the unborn child varies between 3% and 50% depending on the type of congenital heart disease (compared to 1% in women with no cardiac defect). Children of parents with autosomal dominant conditions such as Marfan's syndrome, hypertrophic cardiomyopathy or long QT syndrome have an inheritance risk of 50%.[7]

The Centre for Maternal and Child Enquiries recommends that every woman with known cardiac disease should be offered pre-conception counselling to prevent accidental and potentially dangerous pregnancies in adolescence and to ensure that those at risk of cardiac or obstetric complications enter pregnancy well informed and with a clear management plan.[1] The discussion should address the woman's ability to tolerate pregnancy and delivery and should provide information about maternal and fetal risks, advice on changes in drug therapy and safe options for contraception.[23] Contraception should be tailored to the underlying cardiac condition. Combined oral contraceptive pills are contraindicated in women with hypertension and those at risk of thromboembolism. Insertion of intrauterine devices (copper devices and, to a lesser extent, the levonorgestrel (Mirena) coil) is associated with a risk of bacteraemia and vasovagal syncope; the latter might pose a problem in haemodynamically unstable

patients, such as those with Fontan circulation or Eisenmenger's syndrome. Progesterone preparations are generally safe, and nexplanon is the method of choice for many women with heart conditions.[7,10]

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