Chapter 2

Cardiac disease in pregnancy

D. L. Adamson, M. K. Dhanjal, C. Nelson-Piercy, R. Collis

SYNOPSIS

Heart disease and its management in obstetrics

- Aortic dissection
- Ischemic heart disease
- Arrhythmias
- Obstetric anesthetic management of the mother with cardiac disease
- Anesthetic antenatal assessment and classification of mother with cardiac disease
- Anesthetic interventions
- Anesthetic techniques

Heart disease and its management in obstetrics

D. L. Adamson, M. K. Dhanjal, C. Nelson-Piercy

INTRODUCTION

Pregnancies complicated by significant heart disease are uncommon in the UK, Europe and the developed world. However cardiac disease is now the leading cause of maternal death in the UK. There were 44 indirect deaths attributed to cardiac disease in 2000-2002, giving a death rate of 2.2 per 100,000 maternities. The maternal mortality rate from cardiac disease has continued to rise in the UK since the early 1980s. The major causes of cardiac deaths over the last 10 years are cardiomyopathy (predominantly peripartum), myocardial infarction (most commonly due to coronary artery dissection), dissection of the thoracic aorta and pulmonary hypertension. In the UK, rheumatic heart disease is now rare in women of childbearing age and mostly confined to immigrants.
Women with congenital heart disease are surviving longer thanks to corrective or palliative surgery in childhood. They are therefore not uncommonly encountered in pregnancy. These women may have complicated pregnancies. Women with mechanical prosthetic valves face difficult decisions regarding anticoagulation in pregnancy.

Because of significant physiological changes in pregnancy, symptoms, such as palpitations, and signs such as an ejection systolic murmur are very common innocent findings. Not all women with significant heart disease are able to meet these increased physiological demands. The care of the pregnant and parturient woman with heart disease requires a multidisciplinary approach, involving obstetricians, cardiologists and anesthetists, preferably in a dedicated antenatal cardiac clinic. This allows formulation of an agreed and documented management plan encompassing management of both planned and emergency delivery.

PHYSIOLOGICAL CHANGES IN PREGNANCY

Blood volume starts to increase by the fifth week after conception secondary to estrogen- and prostaglandin-induced relaxation of smooth muscle that increases the capacitance of the venous bed. Plasma volume increases and red cell mass rises but to a lesser degree, thus explaining the physiological anemia of pregnancy. Relaxation of smooth muscle on the arterial side results in a profound fall in systemic vascular resistance and together with the increase in blood volume, determines the early increase in cardiac output. Blood pressure falls slightly but by term has usually returned to the pre-pregnancy value. The increased cardiac output is achieved by an increase in stroke volume and a lesser increase in resting heart rate of 10-20 beats/min. By the end of the second trimester the blood volume and stroke volume have risen by between 30 and 50%. This increase correlates with the size and weight of the products of conception and is therefore considerably greater in multiple pregnancies as is the risk of heart failure in heart disease.

Although there is no increase in pulmonary capillary wedge pressure (PCWP), plasma colloid oncotic pressure is reduced. The colloid oncotic pressure-pulmonary capillary wedge pressure gradient is reduced by 28%, making pregnant women particularly susceptible to pulmonary edema. Pulmonary edema may be precipitated if there is either an increase in cardiac pre-load (such as infusion of fluids), increase in left atrial and pulmonary venous pressure (such as in mitral stenosis) or increased pulmonary capillary permeability (such as in pre-eclampsia), or a combination of these factors.

In late pregnancy in the supine position, pressure of the gravid uterus on the inferior vena cava causes a reduction in venous return to the heart and a consequent fall in stroke volume and cardiac output. Turning from the lateral to the supine position may result in a 25% reduction in cardiac output. Pregnant women should therefore be nursed in the left or right lateral position wherever possible. If the mother has to be kept on her back, the pelvis should be rotated so that the uterus drops to the side and cardiac output as well as uteroplacental blood flow are optimized. Reduced cardiac output is associated with a reduction in uterine blood flow and therefore in placental perfusion; this can compromise the fetus.

Labor is associated with further increases in cardiac output (15% in the first stage and 50% in the second stage). Uterine contractions lead to autotransfusion of 300-500 mL of blood back into the circulation and the sympathetic response to pain and anxiety further elevate heart rate and blood pressure. Cardiac output is increased more during contractions but also between contractions. The rise in stroke volume with each contraction is attenuated by good pain relief and further reduced by epidural analgesia and the supine position. Epidural analgesia or anesthesia cause arterial vasodilation and a fall in blood pressure. General anesthesia is associated with a rise in blood pressure and heart rate during induction, but cardiovascular stability thereafter. Prostaglandins given to induce labor have little effect on hemodynamics but ergometrine causes vasoconstriction and syntocinon can cause vasodilation and fluid retention.

Following delivery of the baby up to 1 L of blood may be returned to the circulation due to the relief of inferior vena cava obstruction and contraction of the uterus. The intrathoracic and cardiac blood volumes rise, cardiac output increases by 60-80% followed by a rapid decline to pre-labor values within about 1 h of delivery. Transfer of fluid from the extravascular space increases venous return and stroke volume further. Those women with cardiovascular compromise are therefore most at risk of pulmonary edema during the second stage of labor and the immediate postpartum period. All the changes revert quite rapidly during the first week and more slowly over the following six weeks but even at a year significant changes still persist and are enhanced by a subsequent pregnancy.
NORMAL FINDINGS ON EXAMINATION OF THE CARDIOVASCULAR SYSTEM IN PREGNANCY

Normal findings on cardiovascular examination may include a loud first heart sound with exaggerated splitting of the second heart sound and a physiological third heart sound at the apex. A systolic ejection murmur at the left sternal edge is heard in nearly all women and may be remarkably loud and be audible all over the precordium. It varies with posture and if unaccompanied by any other abnormality it reflects the increased stroke output. Venous hums and mammary souffles may be heard. Because of the peripheral vasodilation the pulse may be bounding and in addition ectopic beats are very common in pregnancy.

CARDIAC INVESTIGATIONS IN PREGNANCY

The electrocardiographic (ECG) axis shifts superiorly in late pregnancy due to a more horizontal position of the heart. Small Q-waves and T-wave inversion in the right precordial leads are not uncommon. Atrial and ventricular ectopics are both common.

The amount of radiation received by the fetus during a maternal chest X-ray (CXR) is negligible and a CXR should never be withheld if clinically indicated in pregnancy. Transthoracic echocardiogram is the investigation of choice to exclude, confirm or monitor structural heart disease in pregnancy. Transesophageal echocardiograms (TEE) are also safe with the usual precautions to avoid aspiration. Magnetic resonance imaging (MRI) and CT pulmonary angiography are considered safe in pregnancy (see Ch. 21). Routine investigation with electrophysiological studies and angiography are normally postponed until after pregnancy but angiography should not be withheld if required, for example in acute coronary syndromes.

CONGENITAL HEART DISEASE

Congenital heart disease (CHD) in adults is increasing in frequency as a result of the success of congenital cardiology and cardiac surgery. It is the commonest birth defect with just under 1% of newborns having CHD. Half the CHD population are women, and are usually well enough to contemplate and undergo pregnancy. The majority of women with CHD who reach childbearing age do so because they either have a lesion which is associated with long-term survival or because they have had successful surgery. Whilst some may not be able to tolerate the hemodynamic changes of pregnancy, the majority will have enough cardiac reserve to carry a pregnancy to term. CHD is infrequently associated with maternal mortality, however, there may be a significant deterioration in the maternal condition. These complex patients should be jointly cared for by obstetricians, obstetric physicians and cardiologists with expertise in pregnancy and adult congenital heart disease and provide a challenge for such a team.

Pre-pregnancy counseling and assessment

Most women with congenital heart disease are known to a cardiologist prior to pregnancy. It is extremely important that such women are appropriately counseled about their individual health risks with pregnancy. If pregnancy carries an unacceptable risk, this should be explained and appropriate contraceptive advice given. Advising an individual may be difficult, particularly those with corrected complex disease, as data remain sparse for pregnancy outcomes following some forms of corrective and palliative surgery.

Maternal risk of pregnancy is assessed by obtaining an accurate history, which will define the functional status, examination and undertaking relevant investigations. This also provides a baseline of the cardiovascular status, allowing any change occurring in pregnancy to be objectively assessed. It may be necessary to treat the woman prior to pregnancy in order to minimize risk.

Poor maternal functional class (see Table 2.1), cyanosis, history of transient ischemic attack (TIA), heart failure, left arrhythmia, left heart outflow obstruction and impaired ventricular function have all prospectively been found to be associated with poor maternal outcome. Maternal cyanosis is also a risk factor for fetal and neonatal complications. There is only a 12% likelihood of a live birth if the resting arterial saturation is ≤ 85% whilst ≥ 85% is associated with a 63% likelihood of a live birth.

Most CHD is inherited in a multifactorial manner, however single gene disorders, chromosome abnormalities and maternal disease can be the cause. The risk of recurrent CHD is summarized in Table 2.2. The risk of cardiac malformation secondary to maternal diabetes mellitus is approximately 2-3%, while in single gene or chromosomal abnormalities such as Marfan, Noonan and Holt-Oram syndromes the risk of inherited heart disease is 50%.
Table 2.1 New York Heart Association (NYHA) functional classification

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>No breathlessness/uncompromised</td>
</tr>
<tr>
<td>II</td>
<td>Breathlessness on severe exertion/slightly compromised</td>
</tr>
<tr>
<td>III</td>
<td>Breathlessness on mild exertion/moderately compromised</td>
</tr>
<tr>
<td>IV</td>
<td>Breathlessness at rest/severely compromised</td>
</tr>
</tbody>
</table>

Table 2.2 Risk of congenital heart disease in a future pregnancy

<table>
<thead>
<tr>
<th>Risk factor</th>
<th>Risk of CHD in future pregnancy (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>One previous child with CHD</td>
<td>2</td>
</tr>
<tr>
<td>Two previous children with CHD</td>
<td>10</td>
</tr>
<tr>
<td>Mother with CHD</td>
<td>6</td>
</tr>
<tr>
<td>Father with CHD</td>
<td>2</td>
</tr>
</tbody>
</table>

The mode of delivery may also require some consideration prior to conception, although events in pregnancy may influence this. Cardiologists in the past have traditionally favored a Cesarean section (CS). In the majority of cases, however, vaginal delivery is safest for the mother. In comparison to CS, vaginal delivery is associated with a reduction in both blood loss and thromboembolic risk. There are also less abrupt hemodynamic changes, particularly when assisted by effective analgesia and a short second stage of labor. Vaginal delivery has to be balanced against the lack of predictable delivery time where a team can be prepared for all eventualities. Absolute indications for CS are Marfan syndrome with an aortic root dilated beyond 4.5 cm, aortic aneurysm and an acutely unwell mother.

A cyanotic heart disease

Septal defects

Atrial septal defects (ASD) are the most common congenital lesions found in adults, making up between 10-17%, while ventricular septal defects (VSD) are the commonest form in children (1.5-3.5 per 1000 live births). ASDs are often left uncorrected unless there is either a large hole or significant left to right shunt causing right ventricular overload and dilatation. If small or corrected, they cause no specific or significant problems during pregnancy and do not require antibiotic prophylaxis (Table 2.3). In an uncorrected defect, the effect of an increase in cardiac output on a volume-loaded right side is counterbalanced by the decrease in peripheral vascular resistance. The two main areas of concern are paradoxical embolus or arrhythmia. The former is rare but if the pregnant woman is immobilized or has other risk factors for thrombosis, thromboembolic prophylaxis is advised. The risk of atrial arrhythmias is well established with ASDs and remains even with correction. These should be treated dependent upon the rhythm (see arrhythmia section).

VSDs are less common and often corrected in childhood, unless they are small and hemodynamically insignificant. The only risk is that of endocarditis and prophylaxis is advised for delivery (see Table 2.3).

Valve disease

While all valves can be congenitally affected, the commonest lesion is that of a bicuspid aortic valve leading to aortic stenosis. This results in a fixed cardiac output. The majority of young women are asymptomatic and pregnancy is likely to be tolerated well. It is important to assess these women prior to pregnancy to determine whether it would be safer undergoing a pregnancy with the native valve or whether surgery should be contemplated prior to conception. Pregnancy is likely to be well tolerated if:

1. Resting ECG has no ST segment depression.
2. On exercise testing, there is an appropriate increase in BP and HR and no ST change.
3. The pressure across the aortic valve measures less than 80 mmHg at peak and mean pressure gradient is less than 50 mmHg.
4. There is good LV function

Women should be monitored throughout pregnancy to identify symptoms and signs suggestive of decompensation, such as an exaggerated tachycardia or dyspnea, angina, pulmonary edema or new ECG changes. A fall in pressure gradient across the aortic valve is suggestive of a reduction in LV function. Women with symptomatic aortic stenosis should be managed with bed rest to prevent the requirement of an increased cardiac output, and beta blockade which reduces cardiac output as well as lengthening diastole to aid coronary filling. In extreme cases of decompensation, emergency balloon aortic valvotomy or aortic valve replacement can be considered.

The risk of atrial arrhythmias is well established with ASDs and remains even with correction. These should be treated dependent upon the rhythm (see arrhythmia section).

VSDs are less common and often corrected in childhood, unless they are small and hemodynamically insignificant. The only risk is that of endocarditis and prophylaxis is advised for delivery (see Table 2.3).
but are associated with significant maternal and fetal morbidity and mortality. Where the woman has progressed well through pregnancy, a vaginal delivery can be considered but hypovolemia and vasodilators should be avoided. Some women with bicuspid aortic valves have post-stenotic dilatation of the aorta and are at risk of aortic dissection. A history of chest pain should thus be taken extremely seriously.

Aortic pathology

The commonest problems encountered are aortic dissection (see later) and coarctation of the aorta. Although women with aortic coarctation have usually undergone corrective surgery prior to pregnancy, recoarctation or aneurysm formation at the site of surgery can occur. The aorta should therefore be assessed prior to conception or as early in pregnancy as possible, preferably with MRI. If there is no significant defect, then a woman should be able to undergo a vaginal delivery with a short second stage. Patients with coarctation or aneurysm should have aggressive blood pressure management to minimize the risk of dissection (see aortic dissection). Pregnancy is usually well tolerated by all groups and mortality is low.

Congenitally corrected transposition of the great arteries

In this condition the ventricles are switched round, i.e. the right atrium drains into the left ventricle which is positioned where the right ventricle should be and then it drains back out into the pulmonary artery. Deoxygenated blood is still directed to the lungs, and oxygenated blood to the body. However, the morphological right ventricle now has to support a systemic circulation and pressure, which it is not designed to do. Additionally, the tricuspid valve between the left-sided atrium and ventricle is prone to regurgitation in this higher-pressure system. If patients are well and have no deterioration in right (systemic) heart function, they will probably tolerate pregnancy well. However, right heart function as well as systemic tricuspid ativoventricular valve regurgitation may deteriorate in pregnancy. Finally, the ativoventricular node is absent and thus patients often have heart block requiring pacing.

Hypertrophic cardiomyopathy

This condition, previously thought to be rare, may affect up to 2 in 1000 people. Up to 70% of cases are familial with autosomal dominant inheritance. The
diagnostic criteria for hypertrophic cardiomyopathy (HCM) are unexplained asymmetrical myocardial hypertrophy on echocardiography. Diagnosis is usually made as the result of echocardiography to investigate symptoms, a heart murmur or as part of familial screening.

Many women encountered in pregnancy are asymptomatic. Clinical features do not relate to the degree of left ventricular outflow tract obstruction and include chest pain, breathlessness, pre-syncope, syncope, atrial or ventricular arrhythmias, heart failure and sudden death. The overall risk of disease-related complications such as sudden death, endstage heart failure, and fatal stroke is roughly 1-2% per year. Risk factors for sudden death include a positive family history with 2 or more sudden cardiac deaths at < 40 years (as the risk of sudden cardiac death may be increased in certain genotypes), abnormal blood pressure response to exercise (failure of systolic blood pressure to rise by > 25 mmHg from baseline values) and ventricular tachycardia. Most sudden deaths occur in patients with left ventricular wall thickness less than 30 mm, so the presence of mild hypertrophy cannot be used to reassure patients that they are at low risk.

There are case reports of sudden death in pregnancy in patients with HCM however large series report few problems with pregnancy in patients with HCM. It is thought that deaths recorded may reflect the background expected rate and pregnancy seems not to be associated with an increased risk.

In pregnancy, beta-blockers should be commenced in women with symptoms. Caution is needed with regional anaesthesia/analgesia since vasodilation may be poorly tolerated. Any hypovolemia or blood loss should be aggressively corrected and the patient kept ‘well filled’ during labor and delivery. Asymptomatic HCM is not an indication for Cesarean section and most of these women may deliver in their local units.

**Cyanotic heart disease**

**Cyanotic heart disease with pulmonary hypertension**

Eisenmenger’s syndrome is a broad term applied to any cardiac anomaly in which the pathological process of increased pulmonary flow leads to obliterator pulmonary vascular disease. The result is pulmonary hypertension (PH), which is severe enough to reverse the left-to-right shunt to right-to-left. The PH is usually irreversible even after corrective surgery. Pulmonary hypertension of any cause carries a 40-50% risk of maternal death even if pulmonary pressures are only half of systemic.

Women with PH should be strongly advised against pregnancy and adequate contraception is imperative. The subdermal progestogen implant Implanon® is the most suitable contraceptive in view of its effectiveness and lack of cardiovascular risk. It is more effective than sterilization, which requires a general anesthetic. It can be inserted even with those on warfarin. If the patient is on the enzyme inducer Bosentan which renders Implanon® less effective, oral progestagen-only oral contraception should be added. IUDCs are usually best avoided due to the risk of a vagal response with cervical dilatation.

Women with PH who do become pregnant should be advised of the high risk of mortality and termination of pregnancy advised. Termination of pregnancy can be performed medically or surgically depending on facilities available. A medical termination can be performed at any gestation using mifepristone and then misoprostol or gemprost. The disadvantages are that it may take several days to complete during which there may be significant pain and bleeding. This can result in a tachycardia that may not be tolerated well. Adequate analgesia is essential. The procedure should be performed in a high dependency area with full monitoring. Expulsion of the products of conception or delivery of the fetus can be unpredictable. Products of conception may be retained requiring surgical evacuation.

Surgical termination can be performed by suction, or at gestations over 13 weeks, by dilatation and evacuation. Both procedures can be planned with appropriate medical staff available. The risks of a vagal response, uterine perforation and bleeding are reduced with skilled operators.

If women decline termination of pregnancy, they require careful management with a multidisciplinary team with expertise in PH. Bosentan has been shown to be teratogenic in the first trimester in animal studies, but it continued use may be necessary for maternal health. Prophylactic heparin throughout the pregnancy and elective admission for bed rest and oxygen therapy (which acts as a vasodilator) from 20-24 weeks’ gestation onwards are usually recommended. Most women with Eisenmenger’s syndrome who die as a result of pregnancy, do so soon after delivery. There is no evidence that Cesarean versus vaginal delivery, nor regional versus general analgesia/analgesia reduces this risk. The dangers relate to increasing the right-to-left shunt and escalating pulmonary hypertension often despite intensive and appropriate care. The principles of management include multidisciplinary discussion and planning of elective delivery. Postpartum management should be in an intensive care environment by intensivists,
anesthetists, cardiologists and obstetricians. Supplemental oxygen reduces pulmonary vascular resistance. It is important to avoid hypovolemia and acidosis, maintain pre-load, continue thromboprophylaxis and avoid pulmonary artery catheters, which carry a risk of potentially devastating in situ thrombosis. Systemic vasodilators (nitrates, epidural anesthesia and syntocinon) should be avoided but selective pulmonary vasodilators, e.g. inhaled nitric oxide, i.v. prostacyclin, sildenafil and the endothelin antagonist bosentan may be helpful although many women die despite optimal use of these measures.

**Cyanotic heart disease in the absence of pulmonary hypertension**

In complex cyanotic heart disease without PH maternal risk is dependent upon:

1. Ventricular function (see Table 2.1).
2. Hemorrhage due to impaired clotting factors and platelet function.
3. Paradoxical embolus as all cyanotic patients shunt from right to left.
4. Heart failure.
5. Increasing cyanosis secondary to the vasodilatation of pregnancy.

Cyanosis is associated with adverse effects on the fetus with an increased incidence of low birth weight, prematurity and fetal loss (see Table 2.4). These patients need careful monitoring in a unit that understands the complex physiology of the maternal circulation. Women often require bed rest and oxygen which may reduce cyanosis and, as such, improve fetal well-being.

While spontaneous vaginal delivery is best for the mother, fetal growth restriction or fetal distress often precipitates a Cesarean section. It is important in these situations to maintain the circulatory volume and avoid significant vasodilation. The use of aspirin in these women is controversial because of the increased bleeding risk associated with poor platelet function.

**Post surgical correction**

The majority of women who have had definitive correction of simple lesions such as ASD, VSD, patent ductus arteiosus (PDA) can be treated as normal provided there is no significant residual lesion. The increased risk of arrhythmias, particularly atrial arrhythmias remains. In normal pregnancy, right ventricular volumes increase and this may lead to problems in women with right ventricular dysfunction. The more complex procedures, particularly those with shunts are prone to specific complications and are discussed below.

**Repaired tetralogy of Fallot**

Despite the complexity of the original lesion, a successful correction is likely to lead to an uncomplicated successful pregnancy. One report of 40 pregnancies in 27 women demonstrated no serious complications in any of the pregnancies and an incidence of miscarriage comparable to that of the general population. A second report of 63 pregnancies in 29 women reported that 13 ended in termination and 6 pregnancies were complicated by arrhythmias and/or right sided heart failure.

These women require assessment to confirm that there is no significant right ventricular outflow obstruction, pulmonary regurgitation (PR) or reduction in right ventricular function. The PR may get worse as the pregnancy progresses causing increasing tiredness and breathlessness which is best treated with bed rest and diuretics.

**Repaired transposition of the great arteries**

The Mustard operation uses a baffle (conduit) to direct pulmonary venous return into the right ventricle and transposed aorta, and the systemic venous blood via the mitral valve and left ventricle. Essentially, this allows blood to go in the right direction but through the wrong ventricle. Therefore the right ventricle and tricuspid valve support the systemic circulation with the same problems as in congenitally corrected transposition (see above).

The Senning operation is physiologically the same as the Mustard. The difference is that with the Senning, the baffle is created from right atrial wall and atrial septal tissue whilst the Mustard creates the baffle using pericardium or synthetic material.

The Rastelli repair operation uses a valved conduit for certain complex congenital lesions including

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**Table 2.4 Livebirth rates with varying degrees of maternal cyanosis**

<table>
<thead>
<tr>
<th>Maternal oxygen saturation (%)</th>
<th>Livebirth rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;90</td>
<td>92</td>
</tr>
<tr>
<td>≥85–90</td>
<td>63</td>
</tr>
<tr>
<td>&lt;85</td>
<td>12</td>
</tr>
</tbody>
</table>
transposition with pulmonary stenosis. Women with either subaortic stenosis or residual conduit obstruction can develop greater obstruction in pregnancy.

The main complication from repaired transposition of the great arteries is narrowing of the pulmonary venous pathway because of the baffle structure. This fixed obstruction acts as a physiological equivalent of mitral stenosis. These women require complete assessment before or early in pregnancy. However, providing the right ventricular function is good, pregnancy is usually well tolerated.

Post-Fontan procedure

The Fontan circulation was developed for patients with a functional single ventricle. The single ventricle supports the systemic circulation while the systemic venous return is directed to the pulmonary artery directly through a baffle. This means there is no pump directing the blood through this shunt. While this repairs any shunts and thus abolishes cyanosis, the circulation is prothrombotic, has a limited ability to increase cardiac output and poorly tolerates atrial arrhythmias and hypovolemia. However, in those women with good ventricular function and few symptoms, pregnancy may be well tolerated.

ACQUIRED HEART DISEASE

Peripartum cardiomyopathy

Peripartum cardiomyopathy (PPCM) is a rare illness where there is an onset of heart failure with no identifiable cause in the last month of pregnancy or within the first 5 months postpartum, in the absence of heart disease before the last month of pregnancy. It is associated with increased maternal age, Afro-Caribbean race, multiparity, multiple pregnancy and hypertension. It is similar in its clinical presentation to dilated cardiomyopathy, but the latter is not related to pregnancy.

Clinical features

Presentation varies from an incidental finding during echocardiography through to severe heart failure and death. Cardiac decompensation in an otherwise stable patient may occur with iatrogenic fluid or syntocinon infusions, beta-agonists for tocolysis and steroids for fetal lung maturity, which may all cause fluid overload.

Clinically the patient may be tachycardic and tachypleic with congestive cardiac failure and may have an arrhythmia. A CXR will show an enlarged heart and pulmonary edema. The ECG confirms a tachycardia with possible atrial or ventricular arrhythmias, the signs of either left heart strain (ST depression and T-wave inversion in the chest leads) or left bundle branch block (LBBB). Complications include renal and hepatic dysfunction from low cardiac output, life threatening pulmonary edema, systemic or pulmonary emboli from mural thrombus, fatal arrhythmias and death.

The diagnosis is made with the temporal relationship with pregnancy and echocardiography which shows increased cardiac dimensions, left ventricular systolic dysfunction often with global/biventricular involvement (left ventricular ejection fraction < 45%; fractional shortening < 30% in M-mode; left ventricular end-diastolic dimension > 2.7 cm/m²; and dilatation, often of all four chambers).

Management

Treatment is supportive as the underlying condition can only be treated with delivery. Pulmonary edema should be treated by sitting the patient up, giving oxygen, diamorphine and loop diuretics. Vasodilators such as nitrates, isosorbide or hydralazine will help reduce afterload. These can be replaced with ACE inhibitors postpartum, which may also help with cardiac remodeling. Thromboprophylaxis is essential and low molecular weight heparin (LMWH) such as enoxaparin is valuable. A higher dose may be required particularly in those with associated arrhythmias. In such cases warfarin should be used postnatally and breast feeding can continue. Digoxin can be safely used for atrial fibrillation or flutter and beta-blockers may be used with caution for rate control in those with preserved cardiac output.

In many cases preterm delivery will be iatrogenic due to maternal cardiac deterioration. Caution should be used with the administration of steroids for fetal lung maturation in such cases as further fluid overload can occur. A pre-emptive increase in diuretics may prevent sudden deterioration.

In the patient who remains hypoxic and hypotensive, intubation, ventilation and inotropic support is necessary. Delivery by Cesarean section will assist in reducing the cardiac requirement. Occasionally an intra-aortic balloon pump or a left ventricular assist device may be needed in the interim until myocardial recovery occurs or until cardiac transplantation is performed.
Delivery

Vaginal delivery is appropriate in those who have relatively mild disease or in those who are adequately treated. Invasive monitoring in labor is recommended with arterial and central venous lines. Pulmonary wedge pressure readings are not usually necessary. Elevations in central venous pressure (CVP) should be treated with diuretics. Intravenous beta-blockers such as metoprolol may be required for tachycardia. However extreme caution must be used because of their negative inotropic effect. They should be avoided in women in frank pulmonary edema. Analgesia in the form of an epidural needs to be administered with due care and consultant anesthetic involvement is imperative.

Induction of labor is often considered in such patients to allow for insertion of lines, and to plan for all relevant senior staff to be available for labor and delivery. Often patients are on twice daily LMWH preparations, and induction of labor allows the heparin dose to be omitted prior to the onset of labor, thereby allowing the use of regional anesthesia which is very useful for pain control and hence prevention of maternal tachycardia.

In the event of an obstetric indication, or if there is significant maternal compromise or severe disease, a Cesarean section should be performed. Delivery should be planned and is often carried out in operating theatres with access to cardiothoracic facilities.

A dilute infusion of 5 units of syntocinon is used for the third stage. In the event of a postpartum hemorrhage, ergometrine can be used. Misoprostol is useful as an efficient uterine contractor. Thromboprophylaxis should be continued postpartum when there are no bleeding concerns.

Prognosis

There is a highly variable outcome that may not always be predicted by the initial severity of the left ventricular systolic dysfunction or dilatation. Patients may improve with treatment and return to normal (50%), improve slowly and be left with a degree of LV impairment which may over years improve, or may deteriorate despite full medical intervention and require heart transplant. Cardiomyopathy is the cause of almost a quarter of cardiac maternal deaths. In one study, the five-year survival was 94% but data from this study are likely to be over-reassuring as half of the patients with a diagnosis of PPCM had myocarditis on endomyocardial biopsy.

Subsequent pregnancy

Such women should have pre-pregnancy counseling where the risk of cardiac decompensation and maternal death should be discussed frankly.

In those with persistent LV dysfunction or dilatation 6 months after the initial diagnosis of PPCM, pregnancy should be actively discouraged as the risk of worsening heart failure is 50% and maternal death 25%. Adequate contraception should be used such as the intra-uterine progestogen-only system or the subdermal progestogen-only implant.

It is difficult to predict the outcome of a subsequent pregnancy in those whose LV function returns to normal. The contractile reserve may be diminished and this may only become apparent with the hemodynamic stress of a future pregnancy. A study using modified dobutamine stress echocardiography has shown that women who have ‘recovered’ with normal LVEF on standard echocardiography have impaired contractile reserve suggesting an increased risk of deterioration in a future pregnancy. There will be a few women who will tolerate another pregnancy, but it is currently not possible to identify this group. Approximately 20% will develop cardiac failure.

If the cardiac function has returned to normal and the woman wishes to embark on another pregnancy, she should have full hospital combined obstetric and cardiology care. A baseline echocardiogram should be performed. At booking it is essential to ensure that teratogenic medications are discontinued and replaced as necessary. If the pregnancy is unplanned consideration should be given to termination. Echocardiography should be arranged at regular intervals. If clinical or echocardiographic deterioration occurs, serious consideration should be given to discontinuation of the pregnancy either as a termination of pregnancy or as a preterm delivery.

Other dilated cardiomyopathies

These may be idiopathic or related to other conditions such as SLE (see Table 2.5). Women with known impaired LV function from any cause have high-risk pregnancies because they may be unable to meet the demands for an increased cardiac output and deterioration in LV function may occur in pregnancy. In patients with idiopathic dilated cardiomyopathy (DCM) outside pregnancy, their three-year survival falls from 92% in patients with an ejection fraction of greater than 40% to 71% in patients with an ejection fraction of less than 30%. These risks are higher than the mortality risk from peripartum cardiomyopathy. Pregnancy is contraindicated if the patient is New
Table 2.5 Causes of dilated cardiomyopathy

<table>
<thead>
<tr>
<th>Group</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infections</td>
<td>Viral, e.g. coxsackie B, HIV, Ebstein–Barr virus, varicella, echovirus, measles, mumps, pox</td>
</tr>
<tr>
<td>Neuromuscular</td>
<td>Muscular dystrophies</td>
</tr>
<tr>
<td>Nutritional</td>
<td>Niacin, thiamine, selenium deficiencies,</td>
</tr>
<tr>
<td>deficiencies</td>
<td>Kwashiorkor</td>
</tr>
<tr>
<td>Connective tissue</td>
<td>Rheumatoid arthritis, systemic lupus erythematosus, dermatomyositis</td>
</tr>
<tr>
<td>Vascular</td>
<td>Kawasaki disease</td>
</tr>
<tr>
<td>Hematological</td>
<td>Thalassemia, sickle cell disease, iron deficiency anemia</td>
</tr>
<tr>
<td>Drugs</td>
<td>Alcohol, chloroquine, iron overload, cyclophosphamide</td>
</tr>
<tr>
<td>Endocrine</td>
<td>Hypo- and hyperthyroidism, hypoparathyroidism, phaeochromocytoma</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Hemochromatosis, glycogen storage diseases</td>
</tr>
</tbody>
</table>

York Heart Association (NYHA) grade 3-4 (see Table 2.1) or if Left Ventricular Ejection Fraction (LVEF) is less than 30%.

Acquired valvular heart disease

Mitral valve prolapse

This is predominantly a benign condition that should not cause concern in pregnancy. An echocardiogram should be performed to exclude mitral regurgitation. If present, antibiotic prophylaxis against bacterial endocarditis is required (see Table 2.8).

Rheumatic heart disease

In young women most cases of acquired valvular heart disease are due to rheumatic heart disease, or previous endocarditis. Of these, mitral stenosis is the most common and potentially the most likely to cause maternal and fetal compromise.

Stenotic heart lesions

In mitral stenosis, the restriction in outflow from the left atrium results in higher left atrial pressures, left atrial enlargement and eventually right sided heart failure. The reduced blood to the left ventricle causes stroke volume to be reduced. A compensatory tachycardia occurs to maintain cardiac output. This however will reduce the time for filling of the left ventricle in diastole, and results in further increases in left atrial pressure. Thus a vicious circle is begun, ultimately causing pulmonary edema and PH. Left atrial dilatation predisposes to atrial arrhythmias.

Deterioration in pregnancy may occur due to a number of factors including the physiological increase in vascular volume that is maximal at around 20-24 weeks gestation, autotransfusion after delivery of the placenta, iatrogenic fluid infusions, or related to further tachycardia which may be due to pain at delivery, exercise, anxiety, intercurrent infection or arrhythmias.

Pre-pregnancy advice This is essential for women with valvular heart disease. It allows for a full cardiological clinical and echocardiographic assessment prior to embarking on pregnancy. The following points should be considered

1. Is surgical or interventional treatment of valve lesions required pre-pregnancy to optimize maternal and fetal well-being in pregnancy?
2. Is adaptation of potentially teratogenic medication required prior to pregnancy?
3. Is pregnancy contraindicated?

Pregnancy is absolutely contraindicated in women with associated PH, and those with two or more risk factors (see below), due to maternal death rates of 30-60%. Fetal and neonatal risks are also higher. Such women should be given adequate contraception such as a subcutaneous progesterone implant.

Assessment of the degree of cardiac compromise should be made in the remainder. Those women exhibiting any of the risk factors below should be advised of the higher rates of adverse maternal events including pulmonary edema, sustained brady- or tachyarrhythmias requiring treatment, stroke, cardiac arrest or death:

- Reduced left ventricular systolic function with ejection fractions of < 40%.
- Left heart obstruction – aortic or mitral stenosis with valve areas of < 1.5 cm² or < 2.0 cm², respectively.
- Previous cardiovascular events including heart failure, transient ischemic attacks or stroke.
- Reduced functional capacity: disease of NYHA class II or higher (see Table 2.1).

The absolute risks of these adverse events are 4% in women with no risk factors, 27% with one and 62% with two or more risk factors. The fetal and neonatal risks (preterm delivery, intrauterine growth restric-
tion, respiratory distress syndrome, intraventricular hemorrhage and death) are increased in those with left heart obstruction and NYHA class II or higher disease.

Surgical intervention with either balloon valvuloplasty or valve replacement pre-conception (especially if NYHA class III or IV, or mitral valve area < 1 cm²), will change the risk factor profile, allowing many of these women to undergo a relatively less complicated future pregnancy and labor than those treated medically.

Women should be advised to give up smoking and to start folic acid preconceptionally.

Booking visit Arrangements should be made for full hospital care with combined obstetric and cardiological input. A baseline echocardiogram, ECG and U+E should be performed. The ECG may show right axis deviation and P mitrale.

If the pregnancy is unplanned, detailed assessment of functional capacity, left ventricular function, degree of valvular obstruction and history of heart failure or embolic events should be made to see whether the pregnancy can continue or termination should be advised.

Antenatal care A combined assessment with a cardiologist or obstetric physician should be made each trimester at least, and more frequently if clinical deterioration occurs. Although routine echocardiography is unnecessary, it should be performed if there is any change in function.

With mitral stenosis, the heart rate should be controlled with beta-blockers which will allow more filling of the left ventricle in diastole and hence reduces the already elevated pressure in the left atrium. The benefits of this far outweigh the small risk of fetal growth restriction with beta-blockers.

The onset of palpitations warrants a 24-h ECG and echocardiography to establish the degree of heart chamber enlargement and to exclude mural thrombus. If an arrhythmia is detected it should be treated medically or by DC cardioversion (see later). Such a patient should be fully anticoagulated with treatment doses of LMWH.

Pulmonary edema should be treated with oxygen and loop diuretics. Diamorphine will assist in reducing anxiety. A CXR usually shows a small heart with an enlarged left atrium, and pulmonary congestion.

Further deterioration despite optimal drug treatment may require surgical intervention in pregnancy. Percutaneous balloon valvuloplasty of mitral and aortic valves has been performed in the second trimester with good outcome. Mitral valvotomy is best done using transesophageal echocardiography which eliminates the need for radiation. There is a 1% risk of major complications with this procedure which include dislodging of thrombi, cracking of a stenotic valve resulting in regurgitation which may be severe enough to require immediate valve replacement, and death. Comparatively, closed mitral valvotomy has a 3% maternal complication rate, which rises to 5% with open valvotomy. Fetal mortality is 5-15% with closed valvotomy increasing threefold if the procedure is open. The success of open valve replacements in pregnancy for severe mitral stenosis is similar to the non pregnant state, but the stillbirth rate is 10-30%.

Delivery Antibiotic prophylaxis should be administered to prevent bacterial endocarditis (see Table 2.8).

Vaginal delivery should be the aim unless there is an obstetric indication for Cesarean section. The ‘cardiac position’ is best adopted with the legs lower than the abdomen. Lithotomy and supine positions should be avoided. An epidural will provide adequate analgesia and will allow an instrumental delivery to be performed in the second stage. Those with moderate or severe mitral stenosis should ideally have invasive monitoring with an arterial line and CVP. Critical mitral stenosis may require pulmonary arterial (PA) catheterization though this has to be balanced with the risks of leaving a catheter in the PA in a pro-thrombotic patient. Pushing causes rises in the heart rate that may not be tolerated and hence assisted vaginal delivery is often performed.

A dilute infusion of syntocinon should be administered for the third stage (see peripartum cardiomyopathy). A degree of blood loss is tolerated well as it is the autotransfusion just after delivery which often precipitates pulmonary edema in those with critical mitral stenosis.

Regurgitant valve disease The systemic vasodilation and tachycardia that occurs in pregnancy reduces the regurgitant flow of blood allowing pregnancy to be tolerated far better than in those with stenotic valvular lesions. Arrhythmias can however result in pulmonary oedema, and severe regurgitation with ventricular decompensation may cause problems.

Artificial heart valves Bioprosthesis are superior to mechanical (metal) prostheses in all aspects except durability. They are less thrombogenic with reduced thromboembolism rates and hence do not require anticoagulation. The 10-year mortality in women with bioprosthetic valves
is lower than with mechanical valves despite the re-
operations required when bioprostheses require
replacement. It is the mandatory anticoagulation
requirement of mechanical valves that complicates
pregnancy. Pregnancy in a woman with a metal valve
is associated with a maternal mortality rate of 1-4%
with death usually as a result of thrombus formation
on the valve. The thrombotic risk varies and is out-
lined in Table 2.6.

There are four regimes for anticoagulation in preg-
nant women with mechanical heart valves:
1. Warfarin throughout pregnancy and unfraction-
ated heparin (UFH) or LMWH close to term.
2. Warfarin throughout pregnancy except weeks
6-12 and near term when UFH or LMWH is used.
3. UFH throughout pregnancy.
4. Dose adjusted LMWH throughout pregnancy
maintaining anti-Xa level at 0.5-1.2 U/mL

Table 2.6 Thrombotic risk with mechanical
heart valves

<table>
<thead>
<tr>
<th>Type of mechanical valve</th>
<th>Risk of thrombosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single tilting disc valves (e.g. Bjork Shiley)</td>
<td>Single tilting disc valves (e.g. Bjork Shiley)</td>
</tr>
<tr>
<td>Ball and cage valves (e.g. Starr-Edwards)</td>
<td>Ball and cage valves (e.g. Starr-Edwards)</td>
</tr>
<tr>
<td>Bileaflet valves (e.g. Carbomedics)</td>
<td>Bileaflet valves (e.g. Carbomedics)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Position of mechanical valve</th>
<th>Risk of thrombosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mitral &gt; aortic</td>
<td>Mitral &gt; aortic</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Number of mechanical valve</th>
<th>Risk of thrombosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 &gt; 1</td>
<td>2 &gt; 1</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Past history of embolic events</th>
<th>Risk of thrombosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes &gt; no</td>
<td>Yes &gt; no</td>
</tr>
</tbody>
</table>

Table 2.7 shows the fetal and maternal effects of
these regimes, which are all associated with signifi-
cant risks. In general, warfarin is safer for the moth-
but more harmful for the fetus, whereas heparin is the
converse. Warfarin and UFH have similar fetal
wastage of one-third of all pregnancies. Warfarin is
more protective against thrombosis and subsequent
maternal death than UFH, but is associated with sig-
nificant embryopathy in surviving babies. The risks
of congenital malformations and fetal loss are
dose dependent, being significantly higher if doses of
greater than 5 mg are required to maintain an inter-
national normalized ratio of greater than 2.0. Although
heparins do not cross the placenta, UFH is associated
with a 2% incidence of maternal osteoporosis and
thrombocytopenia.

LMWH is an attractive option as it has consider-
ably less maternal side effects of osteoporosis (0.04%)
and thrombocytopenia than UFH, less fetal wastage
and is easier to use. LMWHs have been used in preg-
nancy in women with mechanical heart valves, but
are not licensed for use and are specifically not rec-
ommended by the manufacturers as anticoagulants
in patients with prosthetic heart valves. This was fol-
lowing a series of maternal deaths resulting from
valve thromboses in women who had fixed dosing of
LMWH in pregnancy without monitoring anti-Xa
levels. More recent reports where dose adjusted
regimes have been used are much more encouraging.
Before using LMWHs, careful consideration should
be given to the individual’s thrombotic risk taking
the factors in Table 2.6 into consideration as well as
the presence of atrial fibrillation and impaired left
ventricular function. If used, an initial therapeutic
dose is advisable which is adjusted to maintain
the anti-Xa level between 0.5-1.2 U/mL.

Table 2.7 Fetal and maternal risk with different anticoagulation regimes in women with metal heart valves

<table>
<thead>
<tr>
<th>Anticoagulant regime</th>
<th>Spontaneous miscarriage (%)</th>
<th>Embryopathy (%)</th>
<th>Overall fetal loss (%)</th>
<th>Thrombosis (%)</th>
<th>Maternal death (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Warfarin*</td>
<td>24.7</td>
<td>6.4</td>
<td>33.6</td>
<td>3.9</td>
<td>1.8</td>
</tr>
<tr>
<td>UFH (dose-adjusted)</td>
<td>25</td>
<td>0</td>
<td>43.8</td>
<td>25</td>
<td>6.7</td>
</tr>
<tr>
<td>UFH (&lt;6-12 weeks) +</td>
<td>14.7</td>
<td>0</td>
<td>16.3</td>
<td>9.2</td>
<td>4.2</td>
</tr>
<tr>
<td>warfarin*</td>
<td>7.4</td>
<td>0</td>
<td>12.35</td>
<td>12.35</td>
<td>1.23</td>
</tr>
</tbody>
</table>

*Warfarin replaced by unfractionated heparin (UFH) at term.
*Spontaneous miscarriages, stillbirths and neonatal deaths.
*Only 10% of patients were dose-adjusted according to anti-Xa levels.
Every case should ideally be evaluated pre pregnancy and the woman fully counseled concerning the risks of each treatment regimen. If she has a small Bjork Shiley valve in the mitral position with a previous history of embolic events or arrhythmia and requiring <5 mg warfarin daily then counseling should be directive towards warfarin throughout pregnancy. For a woman with a caromedics valve in the aortic position and no history of previous embolic or arrhythmic events requiring >5 mg of warfarin then LMWH throughout pregnancy with careful monitoring would seem a reasonable option. Often the decision is not so straightforward and the woman may be unwilling to contemplate any risk of warfarin embryopathy especially if she has had a previously affected fetus.

**Delivery**

If warfarin is used, it should be stopped about 10 days pre-delivery to allow for clearance of the drug from the fetal circulation. Unfractionated heparin or LMWH treatment dose can be used until delivery. In labor or during induction, heparin should be stopped, but recommenced after delivery. Conversion back to warfarin should be delayed for at least 3 days postpartum to minimize the risk of obstetric hemorrhage.

The effects of unfractionated heparin can be reversed with protamine sulfate. This also partially reverses the effects of the longer acting LMWHs. Warfarin is reversed with fresh frozen plasma and vitamin K. Such agents may be required if bleeding occurs or if urgent delivery is necessary in the fully anticoagulated patient. It is best to avoid vitamin K if possible as anticoagulation with warfarin postpartum then becomes very difficult.

In the event of a valve thrombus occurring in pregnancy, thrombolytic treatment should be used. The risks of this treatment causing embolism, bleeding or placental abruption are lower than the risks associated with cardiothoracic surgery.

Antibiotic prophylaxis is mandatory to cover delivery in all women with artificial heart valves.

**ENDOCARDITIS PROPHYLAXIS**

Infected endocarditis is not common in pregnancy, but can have fatal consequences for both mother and fetus. The risk lies with any procedure causing bactemia and hence can occur antenatally as well as in labor. Indeed most maternal deaths from bacterial endocarditis in the Confidential Enquiries have not occurred in association with delivery. A propagation of bacteria occurs on a heart valve, mural endocardium or on implanted prosthetic material in the heart and can embolize to the pulmonary vasculature or systemically. Alternatively abscesses or fistulae can occur in the heart or valve prostheses can dehisce.

The indications for antibiotic prophylaxis are given in Table 2.3 and the recommended antibiotics in Table 2.8. Antibiotics should be given before a bacteremia is expected. If antibiotic prophylaxis is not given before this event, antibiotics may help a late clearance if given intravenously within 2-3 h.

**When to give endocarditis prophylaxis**

Prophylaxis against bacterial endocarditis should be given when any obstetric procedure is performed in the presence of infection. The vagina contains commensals some of which can cause systemic infection and endocarditis in the presence of ruptured membranes. Screening for vaginal infection is not routinely performed, hence prophylaxis should be given to women who have ruptured their membranes, at the onset of labor, or before Cesarean section. Insertion of cervical cerclage and urinary catheterization should also be covered. Procedures not requiring prophylaxis include choriovillous sampling, amniocentesis, vaginal examination, transvaginal ultrasound scanning and insertion of regional analgesia.

**AORTIC DISSECTION**

Rupture of the thoracic aorta and its branches has resulted in 19% of cardiac deaths in the UK 1991–2002.

Risk factors are Marfan syndrome with known aortic root dilatation (see above) and hypertension. More recently bicuspid aortic valves have been implicated.

**Table 2.8 Bacterial endocarditis prophylaxis regimens**

<table>
<thead>
<tr>
<th>Group</th>
<th>Penicillin allergy</th>
<th>90–1 h before procedure</th>
<th>6 h later</th>
</tr>
</thead>
<tbody>
<tr>
<td>High risk</td>
<td>No</td>
<td>Amoxyccillin 2 g i.v. +</td>
<td>Amoxyccillin 1 g p.o.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Gentamicin 1.5 mg/kg i.v.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Vancomycin 1 g i.v. over</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>1-2 hr + Gentamicin</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>1.5 mg/kg i.v.</td>
<td></td>
</tr>
<tr>
<td>Moderate risk</td>
<td>No</td>
<td>Amoxyccillin 2 g i.v.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Vancomycin 1 g i.v. over</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>1-2 hr</td>
<td></td>
</tr>
</tbody>
</table>
In pregnancy half of all reported aortic dissections are in women with Marfan syndrome. Most dissections will be Type A, which involve the ascending aorta with 88% of these occurring antenatally and 22% being fatal. Only 20% are Type B involving only the descending aorta which is not associated with previous aortic root enlargement or aneurysm.

The risk of Type A aortic dissection occurring in pregnant patients with Marfan syndrome increases with:

- Aortic root dilatation >4 cm.
- Progressive aortic root dilatation in pregnancy.
- Gestational age (most common in the third trimester).
- Maternal age.
- Family history of dissection.

However women with Marfan have a 1% risk of dissection during pregnancy, even in the presence of a normal sized aorta. Even women with no pre-existing cardiac disease can dissect, thought to be due to the increased cardiovascular stress in the aortic wall and the hormonal changes, which affect collagen tissue.

Pathology

Pregnancy causes alterations in the arterial wall with fragmentation in the reticulin fibers, and reduction in acid mucopolysaccharides and elastic fibers. When occurring in a woman with an inherited or acquired defect in the arterial wall, dissection may result. In Marfan syndrome there is a mutation within the fibrillin gene on chromosome 15q21 which often affects the cardiovascular system and predisposes to aortic root dilatation.

Preconception counseling

Women with Marfan syndrome or BAVD where the aortic root diameter is >4.5 cm should be offered prepregnancy aortic root replacement. The use of a composite graft (i.e. an aortic root replacement with an aortic valve replacement) will require anticoagulation with warfarin which can be changed to heparin in pregnancy (see artificial valves). Advice is less clear-cut for those with aortic roots of 4-4.5 cm.

Antenatal management

Where there is aortic root dilatation > 4 cm or a progression of aortic root enlargement, beta-blockers should be commenced. There is a small risk of intruterine growth restriction which is far outweighed by the benefits. Hypertension must be aggressively controlled as otherwise this can lead to intimal tears.

Echocardiography should be performed monthly, and delivery considered if the aortic root progressively dilates.

A patient having an aortic dissection will have sudden onset tearing chest pain radiating to the back associated with dyspnoea. Signs include new onset aortic regurgitation murmur and as the dissection advances, MI, CVA and hypotension. A dissection of the descending aorta may have few symptoms. Diagnosis is made with echocardiography, the transoesophageal route being far more sensitive, or CT.

Delivery and treatment of aortic dissection

In those with aortic root dilatation or aneurysm, it is imperative to prevent peaks of hypertension which may result in aortic dissection. The safest mode of delivery in such patients is by elective Cesarean section under regional blockade. Aortic repair should be performed postnatally as the risk of dissection remains.

If there is a Type A dissection, immediate surgery should ensue. In general after 28-30 weeks gestation a Cesarean section under GA should be followed by cardiac surgery which will usually involve replacement of the aortic root, aortic valve and reimplantation of the coronary arteries. Where the gestation is earlier than 28 weeks, aortic repair with the fetus in utero is recommended if there is no distal aortic involvement. Cardio pulmonary bypass will be required. A high-flow, high-pressure normothermic perfusion and a perfusion index of 3.0 is considered to be the safest for the fetus. Hypothermia can cause a fetal bradycardia resulting in hypoxic ischemic encephalopathy or even fetal death.

If there is associated distal involvement of the aorta in a Type A dissection, the fetus will need to be delivered as hypothermia is necessary for the open distal repair. It has been suggested that selective antegrade cerebral perfusion for maternal brain protection and moderate hypothermia (28 °C) can be used in this circumstance at gestations less than 28 weeks with the fetus left in utero. However, as this technique requires stopping cardiopulmonary bypass whilst the distal aorta is repaired, the fetus will not be perfused and hence this method is not recommended.

In those with a Type B dissection, conservative medical treatment should be used in the absence of rupture or hypotension. Delivery of the fetus should be considered as the fetal mortality is high. If there are any complications with a Type B dissection, immediate surgical intervention should proceed with delivery of the fetus and aortic repair.
ISCHEMIC HEART DISEASE

While acute coronary syndrome (ACS) is rare in pregnancy, as women delay childbirth until their late 30s and 40s, coronary artery problems and myocardial infarction (MI) are becoming more common pregnancy. Recent data from a retrospective study in the USA identified a threefold increase in the incidence of MI during pregnancy from 1 in 73400 pregnancies in 1990 to 1 in 24600 in 2000. Atherosclerosis is the predominant pathogenesis outside pregnancy, whereas in pregnancy coronary artery dissection and embolus in the absence of atheroma are more frequent. In the last Confidential Enquiry into maternal mortality, 18% of cardiac deaths were from MI with 63% of these secondary to spontaneous coronary dissection rather than plaque rupture. There is still on-going debate about the role of pregnancy-associated plasma protein A (PAPP-A) in ACS, which is a potential proatherosclerotic metalloproteinase. Diagnosis outside pregnancy relies on a combination of history, ECG changes and elevation of cardiac enzymes. ECG changes may require careful review as there is an incidence of up to 50% of abnormalities of unknown significance in women undergoing Cesarean section. Troponin I (TnI) and Troponin T (TnT) are thought not to be increased above the upper limit of normal both peri- and postpartum in healthy pregnant women and TnI is not affected by anesthesia or Cesarean section. It does however increase in pre-eclampsia, pulmonary embolism, atrial fibrillation and myocarditis.

While some drugs used in ACS are known to be safe in pregnancy (low dose aspirin, nitrates, heparin, beta-blockers and opiates) there are few data for the use of clopidogrel. Animal data appear promising, however experience is limited to isolated case reports. Statins and ACE inhibitors are contraindicated because of teratogenic side effects. There are no published data on either the safety or efficacy of GIIb/IIIa inhibitors, such as abciximab (antiplatelet agents used in the management of ACS).

Cases of MI during pregnancy have been described in a variety of patients however, all appear to be associated with a high maternal and fetal mortality. Coronary angiography should not be withheld in pregnant patients. Percutaneous coronary intervention may provide a better alternative to thrombolysis in these situations as it is associated with less bleeding risk and also allows management of spontaneous dissections with stent implantation. However, stent implantation may be associated with an increased risk of coronary dissection in a vulnerable vessel.

While evidence is scarce, it appears that women who have had ACS prior to conception should ideally delay pregnancy for a year after their event. These women should preferably be revascularized prior to conception or managed very aggressively if they present in pregnancy. Whether revascularized or medically managed, all women should ideally undergo stress testing prior to pregnancy to assess their residual ischemic burden so they may be best advised about the safety of pregnancy. Coronary artery bypass surgery (CABG) in pregnancy, has no increase in maternal risk compared to non-pregnant women, however is associated with a high fetal mortality.

Delivery is often influenced both by the maternal state as well as coexisting conditions such as diabetes and pre-eclampsia. If there is no subsequent angina, vaginal delivery is recommended. Agents that increase blood pressure such as beta-agonists, and ergot derivatives are best avoided or used in small doses.

ARRHYTHMIAS

The management of arrhythmias in pregnancy provides a complex dilemma for the physician. Many cardiologists do not have extensive experience in treating these women and the knowledge that therapy may have an adverse effect on the fetus is intimidating. Diagnosing arrhythmias with Holter monitoring can often be fruitless but treating arrhythmias blind means exposing the fetus to potentially unnecessary drugs. Once diagnosed, arrhythmia treatment requires a balance between maternal symptom control while avoiding or reducing any fetal complications from anti-arrhythmic medication. While there have been no documented maternal deaths from primary arrhythmias in the last UK Confidential Enquiry into maternal mortality, 9% of deaths were defined as sudden adult death syndrome, which raises the possibility of death from a primary arrhythmia. In women with heart disease arrhythmia is one of the five independent predictors of having a cardiac event during the pregnancy and should therefore be treated seriously.

Incidence of arrhythmia during pregnancy

The incidence of new onset and pre-existing arrhythmias are increased in pregnancy. As cardiac arrhythmias can be identified on Holter recordings in up to 60% of normal people under 40, it is not surprising that the antenatal clinic sees its fair share of palpitations. The increase in circulating hormones and
cardiac physiological changes, may explain why some women will present for the first time in pregnancy. The incidence of serious arrhythmias remains low in pregnancy despite the 25% increase in heart rate (HR).

There is no significant increase in arrhythmias in laboring women apart from isolated atrial premature beats (APBs) which can occur in up to 90% of women.

**Cause of arrhythmias**

The main causes of arrhythmias are similar in pregnant and non-pregnant women. Theories explaining the increase in arrhythmia frequency in pregnancy include: heightened awareness; increased plasma catecholamine concentrations and adrenergic receptor sensitivity; atrial stretch and increased end-diastolic volumes due to intravascular volume expansion; and hormonal and emotional changes. Patients with known underlying structural heart disease have a higher incidence of arrhythmias and many patients may already have a diagnosis prior to conception. Increased ectopy is benign and generally well tolerated but may trigger a more significant arrhythmia in a susceptible individual.

**Symptoms**

Palpitations, breathlessness, chest pain or pre-syncope may occur with arrhythmias. In the third trimester, patients may become more symptomatic with activity and thus even minor arrhythmias may present with these symptoms. An accurate history of the onset and offset of arrhythmias as well as the frequency, duration and character of the attacks aids the diagnosis of an arrhythmia and helps distinguish it from the physiological symptoms of advancing pregnancy.

**Diagnosis**

- ECG
- 24- or 48-h Holter monitor
- patient activated event recorder
- implantable loop recorders

In a symptomatic patient with palpitations, an arrhythmia must be distinguished from physiological awareness of the heart beat. An ECG is essential, although the paroxysmal, short-lived nature of most palpitations means they may have subsided by the time the patient reaches an ECG machine. If an ECG is obtained, the normal changes due to pregnancy should be remembered (see earlier). If not a 24-h or 48-h Holter ECG may be necessary, although capture may still be difficult. An accurate symptom diary is necessary which can be related to any abnormality on the Holter.

Asymptomatic arrhythmias should not be treated unless felt to be life threatening.

Less frequent episodes or those which have evaded detection are best documented using a patient activated event recorder. These come in a number of models: a continuous Holter recording; a solid state recorder placed on the chest when the patient is having an attack; or a wristwatch with a recording electrode. These devices record approximately 30 s of ECG which can either be stored or transmitted down a household telephone line to a central recording analyzer. They have the advantage of recording a sequential number of events as well as the ability to make remote diagnosis away from the hospital.

Finally, implantable loop recorders are increasingly being used to make diagnoses particularly in patients with unexplained syncope. There is no experience of these devices in pregnancy, however there is no theoretical reason why they would not be able to be used.

**Types of tachyarrhythmias**

Once an arrhythmia has been captured then it is important to correctly interpret the ECG.

**Broad complex tachycardias**

A broad complex tachycardia is likely to be either ventricular tachycardia (VT) or a supraventricular tachycardia (SVT) (Fig. 2.1) with aberrant conduction.

**VT in the structurally abnormal heart**

**Cardiomyopathy**

Those with cardiomyopathy, arrhythmias are the commonest cause of death outside pregnancy. Those with an ejection fraction under 35% post myocardial infarction are candidates for automatic implantable cardio-defibrillator (AICD) implantation. The data in the non-ischemic cardiomyopathies are less robust.

**Hypertrophic cardiomyopathy (HCM)** Non-sustained ventricular tachycardia (NSVT) is found in approximately 25% of adult patients with HCM although sustained monomorphic VT occurs in less than 1% of patients. Studies suggest that VT is the single best indicator of risk of sudden death in patients with hypertrophic obstructive cardiomyopathy.
Arrhythmogenic right ventricular cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy (ARVD) is a disease characterized by progressive fibrofatty replacement of right ventricular myocardium, initially with regional and later global involvement of the right ventricle. It presents in adolescents or young adults with a mean age of presentation in their mid-30s with a male predominance of up to 80%. The disease can be familial with an autosomal dominant inheritance and incomplete penetrance. Presentation is usually with arrhythmias, mainly ventricular and sudden death is common, particularly in the young. It can be diagnosed in its earlier stages using MRI and later by echocardiography. Those women diagnosed because of presentation with ventricular arrhythmias often have implantable defibrillators already inserted at the time of pregnancy.

Long QT syndromes In one study of 111 pregnancies, women with long QT had no increase in risk of cardiac events in pregnancy, but they did have a significant risk in the postpartum period. There is concern that the increase in heart rate after delivery coupled with the stress of caring for an infant may increase the risk of torsade. It is therefore imperative that women should continue their treatment (usually beta-blockers) throughout the pregnancy and puerperium.

Idiopathic VT

The incidence of VT in women of childbearing age is low unless they have significant underlying heart disease. VT in a young woman with a structurally normal heart, is more likely to be an idiopathic VT, which is associated with a good prognosis and low likelihood of degradation to ventricular fibrillation (VF). Right ventricular outflow tract (RVOT) tachycardia, as identified by left bundle branch block (LBBB), inferior axis and palpitations, is more common in women and states of hormone flux are the most common trigger. It is important to take a careful family history of collapse or sudden cardiac death and to review the 12 lead ECG when in sinus rhythm to confirm the patient does not have a condition associated with prolonged QT interval.

Supraventricular tachycardia

Premature atrial beats can be found in 50% of pregnant women but are generally well tolerated. In patients with documented SVT, sustained arrhythmias are infrequent (2 to 3 per 1000 pregnancies), but patients with paroxysmal SVT have exacerbation in 20% of pregnancies. New onset supraventricular tachycardias are also increased during pregnancy. Whilst SVTs are narrow complex because ventricular
activation is via the Hiss–Purkinje system, it is important to identify the differences between them and not treat them all as ‘SVTs’. They can be broadly divided into two types, those that use an additional connection between the atria and ventricles that set up a re-entrant circuit, and those that result from rapid, abnormal atrial activity (Fig. 2.1).

There are two re-entrant tachycardias: those that have an accessory pathway outside the node atrioventricular re-entrant tachycardia (AVRT), and those that have the additional pathway within the AV node, atrioventricular nodal re-entrant tachycardia (AVNRT). Both these arrhythmias are common with AVNRT being more common in women. The duration and frequency of attacks differs between patients and, in some, attacks are precipitated by exercise. The rate is usually between 130 and 250 beats per minute and is influenced by the sympathetic nervous system. As the circulating impulse re-enters the atria after ventricular activation, there will be no normal p waves, though each QRS complex may be followed by an inverted p wave (retrograde p). If the tachycardia is fast, this may not be detectable on the surface ECG.

The atrial arrhythmias are divided into atrial tachycardia, flutter and fibrillation. The mechanism is confined to the atria and the AV node merely transmits the impulses. Atrial fibrillation is the most easily diagnosed arrhythmia because of its irregular features, however this irregularity may be difficult to see with rapid rates. The typical ‘sawtooth’ flutter waves are not always seen in flutter especially at rates over 100 however a regular ECG at a rate of 150 should alert the clinician to think about atrial flutter with 2:1 conduction. The difference between atrial flutter and tachycardia is that in the latter, the atrial rate is slower therefore the ‘sawtooth’ pattern is not seen. It maybe therefore be difficult to distinguish atrial tachycardia from the re-entrant tachycardias.

The diagnosis of an SVT is often helped by adenosine, which can be safely given in pregnancy. One of the problems of using this drug is that, blocking the AV node, may encourage conduction down an accessory pathway and thus accelerate an arrhythmia. As such, experienced personnel should give it in a monitored area with equipment available for resuscitation.

**Electrophysiological studies in pregnancy**

While electrophysiological studies (EPS) are important in making accurate diagnoses of arrhythmias as well as providing the mapping for the curative radio-frequency ablation, they are often not required as an emergency. The vast majority of arrhythmias can be managed with anti-arrhythmic drugs alone and certainly for the duration of the pregnancy. They are often long procedures and involve a significant amount of radiation exposure. As such, both from the logistics of a pregnant women lying flat for many hours and the X-ray dose to the fetus even if screened, the experience of EPS studies in pregnancy is limited.

There are however case reports of women who were refractory to medical therapy and were experiencing recurrent hemodynamically significant arrhythmias, who underwent successful pathway ablation with limited fluoroscopy.

**Acute management of cardiac arrhythmias**

**Cardiac arrest**

Cardiac arrests are rare but if they occur it is important to be aware of the differences in management of pregnant women compared to other patients. In order to optimize maternal outcome, resuscitation should proceed following established guidelines from the resuscitation council. An obstetrician and pediatrician should be involved from an early stage. In addition to the standard causes of cardiac arrest, amniotic fluid embolism, pulmonary embolism, peripartum cardiomyopathy and acute coronary or aortic dissection are important causes in pregnant or recently delivered women. Cardiac resuscitation is more difficult in a pregnant woman because of:

- aortocaval compression;
- enlarged breasts;
- splinting of the diaphragm.

In the supine position the enlarged uterus can reduce venous return by aortocaval compression. It is imperative to relieve this using sand-bags or a ‘Cardiff wedge’ under the right side of the patient, manual displacement of the uterus to the left or raising of the woman’s right hip. Chest compression may be more difficult because of the enlarged breasts and splinting of the diaphragm. As gastric emptying is delayed in pregnancy, early intubation is recommended to prevent aspiration. After five minutes of resuscitation, the uterus should be emptied by Cesarean section. There are a number of reports where this has been associated with successful resuscitation of the mother.

**DC Cardioversion**

DC cardioversion is safe in all stages of pregnancy. The amniotic fluid buffers and protects the fetus, hence the amount of current reaching the fetus is
small, and is associated with only a small risk of inducing fetal arrhythmias. There have been reports of the need for emergency CS because of fetal arrhythmia particularly in women who are compromised; hence the fetus should be carefully monitored before, and throughout the procedure. One case report concluded that DC cardioversion had led to sustained uterine contraction and fetal distress necessitating urgent CS, but this is rare. In the latter stages of pregnancy, some anesthetists prefer to carry out the procedure using full general anesthetic and intubation in view of the more difficult airway and increased risk of gastric aspiration. Women should be nursed in the wedged position as for a cardiac arrest, otherwise the procedure is the same as for non-pregnant women.

**Sustained ventricular tachycardia**

Women with sustained VT who are compromised with hypotension and circulatory collapse should be treated immediately with cardioversion. An obstetrician should be involved and the fetus should be monitored. VT with hemodynamic stability can be safely treated with anti-arrhythmics. The most appropriate area to nurse the woman is often in the coronary care unit (CCU) or intensive care unit (ITU), which are familiar with the use of the drugs, and where appropriate monitoring and surveillance systems are in place. Lignocaine is the drug of choice because it is both effective and has been extensively used in pregnancy because of its local anesthetic properties. It is known to cross the placenta and result in fetal concentrations around half that of the mother but no fetal adverse effects have been reported. Oral beta-blockers have been successfully used in many pregnancies particularly in women with mitral stenosis or aortic dilatation. Disopyramide should be avoided as it has been reported to stimulate uterine contractions. As the efficacy of intravenous drugs in terminating arrhythmias is 50% or less, alternative methods have been explored. Reports are available of successful arrhythmia termination with anti-tachycardia pacing following the use of a flotation wire thus avoiding the radiation.

**Re-entrant supraventricular tachycardias**

Vagal maneuvers are easily and quickly administered and should be attempted first while drugs or anesthetists are being sought. Their success ranges widely from 20 to 90% and some studies report a greater success with carotid sinus massage, whilst others favour the Valsalva maneuver. The re-entrant tachycardias using the node (AVNRT) are more likely to be successfully terminated than those that involve an accessory pathway. An ECG should be used to monitor the effect of therapies as even when unsuccessful, valuable diagnostic information can be gained for the electrophysiologist.

If unsuccessful, termination with IV adenosine should be attempted. Adenosine is a naturally occurring purine nucleotide that transiently depresses sinus node activity and slows atrioventricular (AV) conduction. It has an efficacy of nearly 100% for terminating SVTs when given as a rapid bolus injection through a large bore cannula or central line. As the half-life of this drug is less than 10 seconds, it is rapidly metabolized by the maternal metabolism and has no appreciable effect on the fetus. Adenosine has been shown to be safe and effective in pregnancy. Most women respond to doses between 6 and 12 mg.

Verapamil is an effective second line treatment for the treatment of SVTs and can be used in doses up to 10 mg without effecting fetal heart rate. It should be given in 5 mg doses and repeated after 5 min if the first bolus is unsuccessful. Fetal distress has been associated with verapamil induced maternal hypotension.

Beta-blockers are the drugs of choice in women with Wolff–Parkinson-White syndrome (WPW), as AV nodal blocking drugs may accelerate conduction through the accessory pathway and cause a deterioration in maternal condition.

As with all arrhythmias, those associated with hemodynamic compromise should be treated with DC cardioversion.

**Atrial fibrillation and flutter**

These arrhythmias are uncommon in young women. If seen in pregnancy they are usually associated with congenital or valvular heart disease as well as metabolic disturbances such as thyrotoxicosis or electrolyte disturbance. Though they may be well tolerated in conditions other than severe mitral stenosis, it is advantageous to terminate the arrhythmia to avoid the need for anticoagulation, particularly as pregnancy is a pro-thrombotic state. Quinidine was thought to be safe in pregnancy, but now it is rarely used outside pregnancy due to the risk of torsade de pointes. Procainamide is a safe alternative. Beta-blockers, verapamil and digoxin can all be used to control ventricular rate. Mexiletine, sotalol and amiodarone (in the acute management) have also been used in small numbers of cases with success.
Chronic management

Drug treatment in pregnancy

The decision to treat a woman for chronic arrhythmias depends upon the frequency, duration and tolerability of the arrhythmia. It is a balance between the benefit of arrhythmia reduction or termination and the maternal and fetal side effects of any drug therapy. The smallest recommended dose should be used initially and be accompanied by regular monitoring of maternal and fetal clinical condition. Various drugs have been used to terminate fetal arrhythmias which provide useful data, although reports are predominantly case reports or small case series. Drugs used include digoxin, adenosine, amiodarone, flecaïnine, procainamide, propanolol, propafenone, quinidine, sotalol and verapamil (Table 2.9). The majority of drugs available only have class C evidence for use in pregnancy. For most women beta-blockers are first-line treatment for prophylaxis of SVT and VT in pregnancy unless they enter pregnancy already on alternative effective therapy such as flecaïnine. Amiodarone has the potential to cause thyroid dysfunction in the fetus and is usually therefore avoided.

Pregnancy and automatic implantable cardio-defibrillators

While the world experience is still low, with the increasing use of these devices, more reports of pregnancies in women with automatic implantable cardio-defibrillators (AICDs) are to be expected. Successful pregnancies are reported, but women with frequent

Table 2.9 Anti-arrhythmic drugs

<table>
<thead>
<tr>
<th>Drugs</th>
<th>Safety profile</th>
<th>Listed complications</th>
<th>Breastfeeding</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenosine</td>
<td>Safe to use in pregnancy with no detectable effect on fetal cardiac rhythm</td>
<td>Pregnant women may respond to lower doses due to a reduction in adenosine deaminase</td>
<td>Safe as short halflife</td>
</tr>
<tr>
<td>Atropine</td>
<td>Unknown but has been used for resuscitation</td>
<td>Insufficient data</td>
<td>Unknown</td>
</tr>
<tr>
<td>Amiodarone</td>
<td>Only for short-term use in emergencies</td>
<td>If prolonged use; fetal hypo- and hyperthyroidism, goitre, IUGR, prematurity</td>
<td>Avoid</td>
</tr>
<tr>
<td>Beta-blockers</td>
<td>Avoid atenolol in first trimester because of concern over IUGR</td>
<td>IUGR, bradycardia, apnoea, hypoglycemia</td>
<td>Safe</td>
</tr>
<tr>
<td>Dibicainide</td>
<td>Good safety profile</td>
<td>Miscarriage and fetal death in toxicity</td>
<td>Safe</td>
</tr>
<tr>
<td>Diltiazem</td>
<td>Too little experience to comment</td>
<td>Skeletal abnormalities, IUGR, fetal death</td>
<td>Unknown</td>
</tr>
<tr>
<td>Disopyramide</td>
<td>Too little data to recommend regular use</td>
<td>Premature uterine contractions</td>
<td>Unknown</td>
</tr>
<tr>
<td>Flecaïnine</td>
<td>Limited literature for treatment of maternal arrhythmias; however maternal ingestion is used to treat fetal SVT</td>
<td>Insufficient data but no reported significant complications. Concerns over its pro-arrhythmic potential in fetus have limited its use in past.</td>
<td>Unknown</td>
</tr>
<tr>
<td>Lignocaine</td>
<td>Good safety profile in pregnancy</td>
<td>Fetal distress may occur in fetal toxicity</td>
<td>Safe</td>
</tr>
<tr>
<td>Quinidine</td>
<td>Good</td>
<td>Rarely; mild uterine contractions, prem labor, neonatal TP, fetal VIIIln damage</td>
<td>Safe</td>
</tr>
<tr>
<td>Procainamide</td>
<td>Possibly as safe as quinidine short term in pregnancy</td>
<td>Chronic use may be associated with lupus-like syndrome, GI disturbance, hypotension, agranulocytosis</td>
<td>Safe</td>
</tr>
<tr>
<td>Propafenone</td>
<td>Unknown</td>
<td>Insufficient data</td>
<td>Unknown</td>
</tr>
<tr>
<td>Sotalol</td>
<td>Safe</td>
<td>Transient fetal bradycardia</td>
<td>Safe</td>
</tr>
<tr>
<td>Verapamil</td>
<td>Safe (first choice class IV drug)</td>
<td>Rapid injection may cause reduced maternal BP and fetal distress</td>
<td>Safe</td>
</tr>
</tbody>
</table>
arrhythmias require intensive management by a multidisciplinary team and often prolonged admission. One study reported a series of 44 women who underwent a pregnancy with an AICD implant in situ. They reported no increases in either device or therapy complications and no increases in the number of shocks the women received compared to pre-conception. In addition to standard AICD management, after each administered shock, monitoring of the baby with a cardiotocograph to ensure satisfactory fetal wellbeing may be considered.

Some groups have also reported the insertion of the AICD in women already pregnant using echocardiography to guide the positioning of the leads and avoiding radiation exposure to the fetus. AICD implantation may represent a safe alternative to medical therapy for ventricular tachycardia.

Bradyarrhythmias
Pathological bradycardia in pregnant women is rare. Some women who have physiological bradycardia may, in the second trimester feel dizzy and their blood pressure falls, however treatment is rarely required. Congenital heart block (CHB) is rare (prevalence 1: 20000) and the majority of these present before childbearing age. It does not usually pose a problem during the pregnancy. Temporary pacing is recommended during the delivery as the Valsalva associated with vaginal delivery increases the chance of worsening bradycardia and syncope as well as allowing for an adequate heart rate response for the increased cardiovascular stress.

Spinal anesthesia for CS can be associated with a high incidence of all grades of bradycardia (up to 13%). Rarely, symptomatic bradycardia has been attributed to aortocaval compression by the gravid uterus which responds to changing the maternal position.

In the rare case of a pregnant woman requiring a pacemaker transesophageal guided lead placement should be considered which avoids the standard X-ray screening techniques used.
Box 2.1 Group 1

Mothers with well-controlled and stable ischemic heart disease
  - Good biventricular function
  - No history of heart failure or pulmonary edema
  - No history of major dysrhythmias
  - Congenital heart disease which has been surgically completely corrected
  - Complete closure of patent ductus arteriosus
  - Complete closure of atrial or ventricular septal defect without residual pulmonary hypertension
  - Repaired coarctation of aorta
  - Small atrial or ventricular defects without a major shunt
  - Minor and asymptomatic pulmonary, aortic and mitral stenosis
  - Minor and asymptomatic mitral or aortic incompetence

Box 2.2 Group 2

Deteriorating NYHA: Any cardiac lesion
  - Continuing episodes of symptomatic ischemia
    - Moderately impaired left ventricular ischemia
      - EF < 40%
    - Treated heart failure or pulmonary edema
  - Episodes of symptomatic dysrhythmias
  - Palliative or partial correction of congenital heart disease
    - Partial correction of tetralogy of Fallot without cyanosis
    - Mustard’s or Senning’s procedure for transposition of the great vessels
  - Cardiomyopathy with mild to moderate ventricular impairment
    - Congenital; e.g. hypertrophic cardiomyopathy
    - Acquired; e.g. peripartum cardiomyopathy or secondary to viral infection or multisystem disease
  - Moderate pulmonary, aortic and mitral stenosis

during their pregnancy to confirm the patient’s history, unless otherwise well documented. Early and rapid availability of cardiology services is therefore required.

The obstetric anesthetist should have the opportunity to see the mother at least once during her pregnancy at the beginning of the third trimester and careful plans should be documented in the notes. The basis for seeing all mothers with cardiac disease antenatally however asymptomatic is so that analgesia and anesthesia can be given without delay once in labor, especially in an emergency. Ideally there should be 24-h anesthetic and obstetric cover on the delivery suite, but anesthetic intervention either anesthesia or analgesia can usually be considered routine with the normal provisos of pregnancy.

Group two

There is an intermediate group of mothers with cardiac disease (Box 2.2) that can present with deteriorating parameters during pregnancy or delivery, but usually tolerate the physiological demands of pregnancy without major problems. These mothers can usually tolerate labor without difficulty (with the possible exceptions of asymptomatic Marfan syndrome and aortic stenosis associated with a dilated aorta > 4 cm or any dilatation in pregnancy), but the anesthetic care may need to be modified.

These mothers should be seen from the early second trimester by a multidisciplinary team, including an obstetric anesthetist, and they are best looked after in a hospital that can offer 24-hour specialist care in case of sudden cardiac problems and obstetric emergencies. Detailed anesthetic plans should be made from the early part of the third trimester with desirable analgesia, levels of monitoring and the appropriate grade of the attending anesthetist recorded.

In many situations a routine low dose epidural for labour analgesia is a safe and appropriate choice, with pulse oximetry and non-invasive blood pressure for monitoring. If a mother needs an emergency caesar-ean section, the different physiological demands resulting from an extensive and dense regional block, a general anesthetic or blood loss more commonly associated with emergency Cesarean delivery, often require a greater level of experience and monitoring than can routinely be provided in many smaller hospitals. The major demands on this level of service provision are problems that arise outside office hours, even if an elective Cesarean section / delivery is planned.

Group three

These mothers have a high risk of deterioration during pregnancy (Box 2.3). The mother may present in the first or the early part of the second trimester with heart failure, where a termination of pregnancy may be necessary or may need delivery, usually by Cesarean...
ean section, early in the third trimester when the baby is viable.

A multidisciplinary team of an obstetrician, obstetric anesthetist and a physician with training in pregnancy, must assess these mothers early in pregnancy at around 20-24 weeks gestation. A pulmonary physician may need to be included if the mother has pulmonary hypertension. Flexible and ongoing assessment will be required throughout pregnancy as plans may rapidly change. The woman will need to be seen every two to four weeks in the mid-trimester of pregnancy and weekly from 30 weeks gestation. She will require monitoring (which may include invasive monitoring) in a high dependency area or intensive care setting at the time of delivery and all plans must be possible on any day and at any time.

ANESTHETIC INTERVENTIONS

Cardiac output increases during pregnancy and again during labor. Part of the increase in cardiac output in labor is caused by autotransfusion during uterine contractions and part by the physiological stress response to pain. Regional analgesia (epidural or combined spinal epidural) is the only form of pain-relief that can reliably obtund the pain response and is frequently offered to mothers with cardiac disease.

Mothers who fall into group one and many from group two will have tolerated pregnancy without difficulty and from the cardiovascular point of view will tolerate delivery well. These mothers can be offered regional analgesia safely, but those that feel they would prefer not to have an epidural need not have one.

Blood pressure and cardiac output control during anesthesia

To maintain blood pressure during anesthesia and prevent detrimental maternal problems (such as reduced coronary filling) and fetal distress, the anesthetist must control preload, afterload and heart rate.

Blood pressure (BP) is a function of cardiac output (CO) and systemic vascular resistance (SVR) and cardiac output is a function of heart rate (HR) and stroke volume (SV). This is summarized in the equation: $BP = (HR \times SV) \times SVR$.

Regional and general anesthesia can affect all of these parameters and it is the attention to the details of parameter control, rather than the absolute method of anesthesia, that will provide safe analgesia and anesthesia for the mother with cardiac disease.

Preload

Aortocaval compression

The majority of mothers will have a degree of aortocaval compression at term. For many mothers it will be asymptomatic, until they are given an anesthetic, where it can be a major contributing factor to significant hypotension in up to 80% of elective Cesarean sections under spinal anesthesia.

For the mother with cardiac disease with either poor ventricular function or a poorly compliant ventricle (especially in association with stenotic valvular lesions), the ability to maintain an adequate stroke volume will be heavily dependent on good venous return (preload) to the right side of the heart. Minor degrees of aortocaval compression causing even a small fall in venous return can cause serious cardiac decompensation either in relation to a fall in systemic vascular resistance associated with regional anesthesia or a further decline in ventricular contractility associated with the commonly used general anesthetic drugs. Mothers with cardiac disease should be nursed in the full lateral position to minimize aortocaval compression, especially in relation to establishing all regional anesthetic blocks. She should be placed on her back with visible left uterine displacement for the shortest possible time, should it be necessary, e.g. for delivery by Cesarean section, and hypotensive episodes treated with an increase in table tilt.

Hemorrhage

Hemorrhage will severely effect preload and may lead to early, rapid decompensation. This can be a problem
when there is no anesthesia and early aggressive fluid management needs to be instituted. If hemorrhage is associated with anesthesia, a fall in blood pressure and coronary filling can be even more alarming.

**Monitoring preload**
Early use of central venous pressure (CVP) monitoring can be very helpful to assess preload and should be considered in all mothers who have poor ventricular function and stenotic valvular lesions. An absolute filling pressure is very much less important than gradual changes or sudden falls in CVP readings. A sudden drop in CVP reading, which is usually rapidly followed by a drop in blood pressure, is often associated with aortocaval compression and re-positioning the mother is urgently required.

**Fluid therapy**
Gradual changes in CVP readings over the course of labor can indicate under filling due to dehydration or over filling associated with over enthusiastic intravenous fluid therapy. Both are detrimental, especially for the mother with poor ventricular function. An under filled mother will decompensate rapidly at delivery, should there be any hemorrhage. A mother who has been given too much fluid may suddenly develop pulmonary oedema at delivery, because of the additional burden of uterine autotransfusion.

In normovolemia, a CVP reading is unnecessary but intravenous fluids should be given to judiciously maintain a good urine output with a mother who is clinically warm and well perfused. In the face of hemorrhage, aggressive and early maintenance of hemoglobin concentration and intravascular volume must be instituted.

**Cardiac factors**

**Heart rate**
A normal HR is essential for maintaining cardiac output and blood pressure. In normal pregnancy, the mother’s increase in cardiac output is largely achieved by an increase in HR. For the mother with cardiac disease it is important to maintain the HR she tolerates best.

Tachycardia is especially poorly tolerated if the mother has ischemic heart disease, stenotic valvular disease or hypertrophic cardiomyopathy. Bradycardia is poorly tolerated in heart failure, a poorly compliant ventricle or regurgitant valvular disease.

**Vasopressors**
If anesthesia is associated with a drop in blood pressure, then having excluded aortocaval compression, the vasopressor of choice will be the drug that maintains or changes maternal heart rate to within the range tolerated best by that mother.

Ephedrine has traditionally been the vasopressor of choice in the obstetric population, with a mild inotropic and chronotropic effect. It is a useful drug in 3-6 mg boluses if the mother’s heart rate is below 80 beat/min or a mild tachycardia is desirable.

Phenylephrine has traditionally been avoided in obstetrics because it was feared that blood flow to the fetus may be affected by its pure alpha agonist, vasoconstrictor action. Recent studies have demonstrated that in the normal fetus, pH at birth may be improved by phenylephrine compared to ephedrine. This has led to a reassurance that phenylephrine can be used safely for the mother with cardiac disease. It can be given either as an infusion at 10-20 µg/min or boluses of 12-25 µg as required. It usually causes a reflex slowing of the maternal heart rate to 60-70 beat/min and blood pressure is well maintained.

**Cardiac contractility**
Regional anesthesia does not have an effect on cardiac contractility and cardiac output is well maintained as long as HR and preload are maintained. Regional anesthesia is particularly appropriate where cardiac function is poor and is the anesthetic of choice in cardiomyopathy and ischemic heart disease.

General anesthesia may be poorly tolerated in the mother with poor ventricular function as all general anesthetic drugs have a negatively inotropic action. If general anesthesia is required, then extreme care must be taken to maintain an appropriate heart rate using vasopressors and vagolytic drugs as required. Inotropic drugs may also be needed.

**Afterload**
Both regional and general anesthetic techniques are associated with a fall in afterload. This facilitates the forward flow of blood from the heart and blood pressure tends to fall.

As cardiac contractility is not affected by regional techniques, the fall in afterload can be beneficial in some conditions. For ischemic problems, cardiac work is reduced and the risk of cardiac failure improved. Regional techniques are also ideal for mothers with regurgitant valvular lesions, with improved forward flow and a reduction in ventricular dilatation and ventricular work.

A fall in afterload is particularly detrimental for the mother with aortic stenosis and a right to left cyanotic shunt. In aortic stenosis a fall in afterload can be associated with a dramatic fall in blood pressure...
leading to poor coronary filling, acute ischemia and dysrhythmias.

In cyanotic shunts a fall in afterload is associated with an increase in right to left shunt resulting in an increase in cyanosis. Phenylephrine is the agent of choice, as its pure alpha affect will minimize these changes.

The argument in these conditions is that general anesthesia is preferable to regional anesthesia, as it may be associated with a smaller fall in afterload. There are however many good descriptions in the literature of regional anesthesia being safely used in these conditions. It is the understanding of the pathophysiology of the condition and its management with tight, early control of cardiovascular parameters that is very much more important than the actual technique used.

ANESTHETIC TECHNIQUES

General anesthesia

For the mother with severe cardiac disease, a modified general anesthetic technique with a small intravenous induction dose of thiopentone or etomidate and intravenous opioids; fentanyl, alfentanil or remifentanil is appropriate. The mother is vulnerable to gastric regurgitation and aspiration and suxamethonium should still be given. The baby may have significant opioid induced respiratory depression due to this technique and the attending pediatrician must be made aware.

Regional techniques

Labor

For mothers with significant disease all regional techniques should be titrated slowly against analgesic requirements. A 0.1% bupivacaine solution with fentanyl or sufentanil given in 5 ml boluses is well tolerated. If analgesia is inadequate combined spinal epidural technique can be considered but intrathecal local anesthetics should be avoided and intrathecal fentanyl 25 µg or sufentanil 10 µg used as a sole agent. The epidural component can then be used to supplement the analgesia.

Cesarean section

Epidural anesthesia is tolerated if given in small incremental doses. To establish a block for Cesarean section, 3-5 ml boluses of 0.5% bupivacaine given every 10-15 min can be given. When establishing a regional block in this way it may take 45-60 min with a total of 25-30 mL of solution. Very careful assessment of the block is required to avoid problems of inadequate anesthesia during the surgery.

A combined spinal epidural technique can also be useful. A 5 mg intrathecal dose of bupivacaine can usefully provide sacral anesthesia with little in the way of hemodynamic instability and the epidural component can then be used in small incremental top-ups, until the desired block is achieved. Sacral anesthesia is more assured with this technique and a smaller dose of epidural bupivacaine will be required.

Uterotonics

Oxytocin

Oxytocin is the drug most commonly used to enhance uterine contractility after delivery and reduce the risk of postpartum hemorrhage. As stated above, the risks of hemorrhage are significant to the mother with cardiac disease. It is therefore important she is given oxytocin but the usual method of giving a bolus of 5-10 units at delivery should be avoided.

Women with right heart problems and pulmonary hypertension may tolerate the autotransfusion from natural uterine contraction poorly. Additional uterine contraction associated with oxytocic drug use may make the situation worse. Gentle manual rubbing of the uterus followed by a slow infusion (40 u over 4-6 h) to maintain uterine contractility is preferable.

Oxytocin also causes a profound fall in afterload and a reflex tachycardia with a significant increase in cardiac output. These physiological parameters are frequently detrimental to the mother with significant cardiac disease and bolus doses should be avoided if possible. A slow infusion will avoid these problems and should be given as above.

Ergometrine

Ergometrine, usually given with oxytocin, can counter the vasodilatation effect of oxytocin alone, but the effects are unpredictable and resulting hypertension can in itself be detrimental. In all but the most straightforward cases this combination should be avoided.

Carboprost

Carboprost is a prostaglandin used to treat post partum hemorrhage due to uterine atony in patients unresponsive to ergometrine and oxytocin. This drug can cause bronchospasm and hypertension, but when faced with severe hemorrhage its use should be con-
sidered, although myocardial ischemia is a known side-effect.

**Physical methods**

The mother with significant cardiac disease will not tolerate major and ongoing hemorrhage. If hemorrhage occurs after vaginal delivery then early transfer to theatre for examination under anesthesia is important. Early initiation of invasive monitoring (blood pressure and CVP) should be considered.

The early use of a uterine compression or brace suture such as a B-Lynch suture can lead to early control of hemorrhage due to placenta previa and uterine atony. Its use at laparotomy can avoid the consequences of major ongoing hemorrhage and avoids the continued use of the above drugs, in mothers who are intolerant of them.

**Further reading**


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