

Review Article

Heart Disease and Pregnancy

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Recent advances in paediatric cardiology and cardiac surgery have enabled more than 85% of patients with congenital heart disease to survive to adulthood. Half of these patients are women, the majority of them of childbearing age. Most interventions, however are not curative, about half of the women with congenital heart disease face the prospect of further surgery, arrhythmia, heart failure, and—if managed inappropriately—premature death. The additional burden of pregnancy represents a new challenge for this growing number of patients with heart disease.¹

The number of women of childbearing age with coronary artery disease is also expected to grow, because of both the advanced maternal age and fertility, and the adoption of the western style of life, which leads to a higher incidence of risk factors for coronary atherosclerosis.²

In the past few decades, rheumatic fever has decreased drastically in developed countries. Nevertheless, rheumatic fever remains problematic in the developing countries. Immigrants represent an additional population that may be at high risk, because of social reasons or because of lack of screening for congenital heart disease in their countries of origin.³

Physiological changes during pregnancy, delivery and the post-partumThe *main physiological changes during*

pregnancy are an increase in blood volume, heart rate and cardiac output and a decrease in peripheral vascular resistance (Figure 1).⁴

- The increase in blood volume (30-50%) represents an adaptation process induced by the increased metabolic demand of the foetus. This increase starts as early as the sixth week of pregnancy; the levels peak by the 20th to 24th weeks of pregnancy, and then are sustained until term or decrease. The underlying mechanism is presumably hormonal, but the exact sequence of events remains unclear.
- Cardiac output increases in parallel with the increase in blood volume (30-50%). The rise of cardiac output in pregnancy is disproportionately greater than the increase in heart rate and is therefore attributable to an augmentation of stroke volume. As pregnancy advances, heart rate increases, approximately by 10 to 20 beats per minute, and becomes the more predominant factor in increasing cardiac output.⁵
 - There is a remarkable fluctuation in resting cardiac output with changes in position; the compression of the inferior *vena cava* by the enlarged gravid uterus in the supine position results in decreased venous return and a concomitant significant decrease in cardiac output.⁶
 - Due to this hyperdynamic status, murmurs develop in nearly all women during pregnancy; they are usually soft mid-

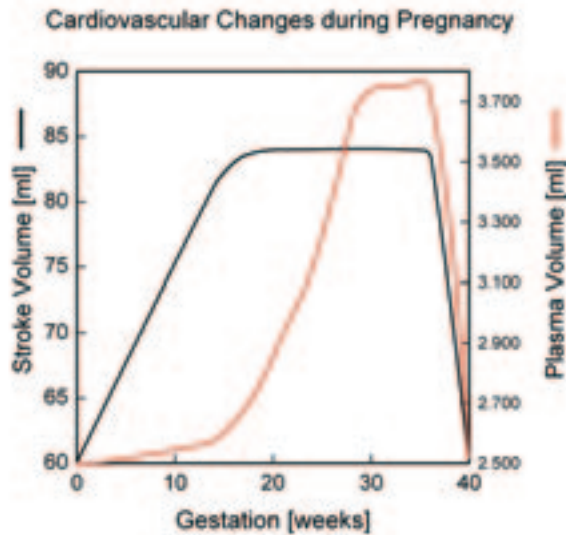


Figure 1. Cardiovascular changes during pregnancy. Changes of stroke volume and plasma volume.

systolic and their intensity may increase as cardiac output increases. A continuous murmur due to increased mammary blood flow may also be heard. Diastolic murmurs can be physiologic during pregnancy. However, echocardiography is also warranted when a diastolic murmur is present.

- Among healthy pregnant woman, serial echocardiography usually demonstrates minor increases in both ventricular diastolic dimensions, which remain within the normal range, with a slight decrease in left ventricular end-systolic diameter and a minimal increase in the size of the left atrium. Left ventricular contractility appears to be depressed in pregnancy, but the ejection fraction is maintained because of the altered loading conditions.⁷ Increased circulating volumes result in increased transvalvular flow velocities. Minor degrees of atrioventricular valve regurgitation are common and of no clinical significance.⁸
- Aortic root diameter increases during pregnancy, by about 2-3 mm.⁹
- The 30% fall in systemic vascular resistance is a fundamental physiological change during pregnancy.
 - This afterload reduction is due to the high flow, low resistance, maternal placental circulation.
 - Pulmonary pressures remain normal during pregnancy, suggesting a similar decrease in pulmonary vascular resistance to accommodate the increased cardiac output.

Haemodynamics are altered substantially during labour and delivery, secondary to anxiety, pain, and uterine contractions.

- Uterine contractions result in a 50% acute increase

in both heart rate and blood volume as uterine blood enters the circulating pool (300 to 400 ml of blood). The magnitude of the increase in cardiac output during contractions increases as labour progresses.

- Both systolic and diastolic blood pressures increase markedly during contractions because of compression of the abdominal aorta by the uterus, resulting in an increase in peripheral vascular resistance. The augmentation is greater during the second stage of delivery.
- In spite of the external haemorrhage associated with delivery, cardiac output is significantly higher one to two hours post-partum prior to initiation of placenta contractions. This 60% to 80% increase in cardiac output occurs immediately after delivery, because of the temporary increase in venous return due to relief of caval compression and the additional blood shifting from the contracting uterus into the systemic circulation.

The cardiovascular adaptations associated with pregnancy regress by approximately six weeks after delivery.¹⁰

Preconception counselling

Discussions about future pregnancies, family planning and contraception should begin in adolescence, both to prevent accidental and possibly dangerous pregnancies and to allow patients to plan their lives.¹¹

Counselling should address how pregnancy may affect not just the mother, but also the foetus. It should ideally be provided in a joint clinic by an obstetrician

with expertise in heart disease and a cardiologist with expertise in women with congenital heart disease. The information should include estimation of maternal mortality as well morbidity—for example, the likelihood of pregnancy resulting in heart failure requiring hospitalisation, exacerbation of arrhythmias, or long term deterioration in ventricular function. Parental life expectancy should also be discussed, as premature death, disability, or the need for major surgery will obviously affect a couple's ability to care for their child.¹²

If pregnancy is planned adequately it will allow the minimisation of maternal risk, if necessary, by catheter or surgical intervention before conception. Any surgical intervention should take into account the potential effect on pregnancy; special consideration should be given for women of reproductive potential to using tissue valves for valve replacement.

The timing of pregnancy is of importance. For example, for those with a systemic right ventricle or univentricular heart, pregnancy is likely to be tolerated better when the patient is in her twenties rather than her late thirties.

Careful planning also allows the estimation and minimisation of foetal risk (see “The risk for the foetus”).

Contraception

None of the contraceptive methods available provides 100% cover at zero risk for women with heart disease. “Natural methods” and “barrier methods” have unacceptably high failure rates and cannot be recommended for women in whom pregnancy carries a substantial risk. Combined oral contraceptives should be avoided in patients at risk of thromboembolism because of the thrombophilic properties of oestrogen.

Progesterone-only oral contraceptives (mini-pill) do not increase the risk for thromboembolism and have few serious side effects, but the failure rate is higher than that of the combined pill. Depot injections of progestogen are an alternative to the mini pill, especially for adolescents for whom compliance is a concern.

An intrauterine device impregnated with progestogen has been an important advance in contraception for patients with a high risk for pregnancy-related complications and thromboembolism. Such devices are highly effective and safe: they reduce menstrual bleeding and carry a very low risk of infection and ectopic pregnancy.

Sterilisation should be considered for women in whom pregnancy would carry a prohibitively high risk.

Termination of pregnancy

The risk of termination of pregnancy increases with increasing gestational age and should be performed as soon as the decision has been made, preferably in the first trimester. Suction curettage under local anaesthesia is the preferred method. Medical abortion with oral antiprogesterones and vaginally administered prostaglandins is probably contraindicated because the haemodynamic effects are unpredictable, although limited information exists regarding patients with heart disease.

The risk for the mother

Risk stratification is based on general knowledge of physiological changes during pregnancy, on existing knowledge of specific heart diseases, especially those which carry high risk for mortality, and on recent prospective clinical studies examining maternal and foetal outcomes during pregnancy.

Conditions with markedly reduced cardiac output, such as stenotic lesions on the left side, are not well tolerated. The decrease in systemic vascular resistance suggests that regurgitant lesions on the left side and left to right shunts will be generally well tolerated. For the same reason, right to left shunts increase risk, and conditions associated with cardiac cyanosis are not well tolerated.

Pulmonary hypertension continues to carry a prohibitive risk during pregnancy, with mortality rates between 30% and 50%, and this needs to be communicated early to the patient and the family (see below: “Pulmonary hypertension”).

In the largest prospective multi-centre study to date (CARPREG) of women with pregnancy and heart disease, the frequency and predictors of pregnancy-related complications among Canadian women with heart disease were examined.^{13,14} There were 562 women during 617 pregnancies. The study included all pregnant women with congenital or acquired cardiac lesions, excluding those who had mitral valve prolapse with less than moderate mitral regurgitation and those with arrhythmias. A primary cardiac event occurred in 80 completed pregnancies (13%); half of them occurred in the prepartum period. The most frequent cardiovascular complications were heart failure and arrhythmias. Three deaths were reported.

The risk factors for cardiovascular complications are shown in Table 1. The estimated risk of a cardiac

Table 1. Generic cardiac risk factors for adverse maternal outcomes during pregnancy.

- Poor functional class before pregnancy (NHYA class >II).
- Impaired left ventricular function (ejection fraction <40%).
- Left heart obstruction, mitral valve area <2 cm², aortic valve area <1.5 cm², left ventricular outflow tract peak blood pressure gradient measured by Doppler echocardiography >30 mmHg.
- Preconception history of adverse cardiac events such as symptomatic arrhythmia, stroke, transient ischaemic attack and pulmonary oedema.

The estimated risk of a cardiac event in pregnancies without risk factor was 5%, with one risk factor 27%, and 75% if two risk factors were present.

event in pregnancies without risk factor was 5%, with one risk factor 27%, and with two risk factors 75%.

Regarding patients with congenital heart disease in particular, a recent publication described 90 pregnancies in 53 women, with 19.4% of primary maternal cardiac events; pulmonary oedema occurred in 16.7% and sustained arrhythmias in 2.8%. There were no maternal deaths. In addition to the previous risk factors reported in the CARPREG study, right ventricular dysfunction in patients with ventriculo-arterial concordance and/or severe pulmonary regurgitation were independent risk factors for cardiac events, but not for death.¹⁵

The risk for the foetus

The potential for congenital heart disease in the offspring must be considered before conception. Overall, the chance is 4%¹⁶ compared with a background of 0.8% for the general population.¹⁷

For lesions with autosomal dominant inheritance – DiGeorge, Marfan syndrome, Noonan syndrome and hypertrophic cardiomyopathy – the risk of recurrence of heart disease can be as high as 50%. If the mutation is known, prenatal diagnosis at 12 weeks of gestation can be made by corionic biopsy.

A foetal nuchal translucency measurement at 12-13 weeks gestation is a useful first screening test (the incidence of congenital heart disease is only 1/1000 with normal nuchal thickness). A specialist foetal cardiac echo scan at 14-16 weeks gestation should be offered to detect moderate to severe congenital heart lesions, and this may need repeating at 20 weeks.

For women with cyanotic or stenotic lesions, routine ultrasound biometry of the foetus is warranted. Close assessment of foetal growth is also advisable in patients with systemic hypertension or taking beta-blockers. Maternal drugs may need changing before conception or once pregnancy is confirmed.

Overall, there is a higher incidence of foetal and neonatal adverse events, including intrauterine growth restriction, premature birth, intracranial haemorrhage, and foetal loss, in women with heart disease compared with the general population. By far the most common complication in the foetus is growth restriction, particularly when the increase in cardiac output is inadequate so that the flow to the placenta is restricted. This is amplified by any other obstetric risk factors.

In a prospective study that examined the incidence of adverse neonatal outcomes in women with heart disease compared to healthy women, the incidence was 18% and 7% respectively (Table 2). In patients with heart disease without obstetric complications and without known risk factors for neonatal complications the actual incidence of neonatal complications was similar to that in healthy women.¹⁸

Management of pregnancy, delivery and the post-partum

The level of antenatal care and monitoring required should be determined before pregnancy, or when this is not possible as soon as pregnancy is confirmed. See Table 3, which includes risk stratification of heart diseases during pregnancy. As most general obstetricians will see only few patients with moderate to severe heart disease, referral to a specialist centre for counselling is strongly advisable.

Low risk patients can continue with their antenatal care locally, taking into account the specialist recommendations.¹⁹ Moderate to high-risk patients should be cared for in a tertiary, multidisciplinary environment. Women with significant valvular heart disease should be evaluated periodically, including echocardiography (every trimester and whenever there is a change in symptoms).

Careful planning for antenatal care and delivery are clearly required. Some patients may benefit from hospi-

Table 2. Generic risk factors for adverse neonatal outcome.

- Poor functional class before pregnancy (NHYA class >II).
- Impaired left ventricular function (ejection fraction <40%).
- Left heart obstruction, mitral valve area <2 cm², aortic valve area <1.5 cm², left ventricular outflow tract peak blood pressure gradient measured by Doppler echocardiography >30 mmHg.
- Cyanosis.
- Anticoagulant therapy.
- Multiple gestation.
- Smoking.
- Maternal age less than 20 or more than 35.

Table 3. Risk stratification of heart diseases during pregnancy.

High risk	Moderate risk Estimated risk of mortality or major complications: 1- 5%	Low risk Estimated risk of mortality or major complications: >1‰ and <1%
Pulmonary hypertension (mortality 30-50%) <ul style="list-style-type: none"> • Eisenmenger syndrome • Primary pulmonary hypertension 	Cyanotic lesions without pulmonary hypertension	Left to right shunts
Univentricular physiology with or without Fontan with severe dysfunction	Single ventricle physiology (with or without Fontan) with good systemic ventricular function	Functionally normal bicuspid aortic valve
Severe aortic/mitral stenosis	Mechanical prosthesis	Biological prosthesis without residual cardiac dysfunction
Marfan syndrome with severe aortic root dilatation	Not severe aortic/mitral valve stenosis	Asymptomatic moderate to severe mitral or aortic regurgitation without left ventricular dysfunction
Acute myocardial infarction during pregnancy	Severe pulmonary stenosis	Moderate pulmonary stenosis
Severe systemic ventricular dysfunction: <ul style="list-style-type: none"> • Dilated cardiomyopathy • Previous peripartum cardiomyopathy • Right ventricular systemic ventricle with severe dysfunction (corrected congenital transposition of the great arteries, Mustard) 	Marfan syndrome without marked aortic root dilatation	Repair of Tetralogy of Fallot without residual lesions such as severe pulmonary regurgitation
	Unrepaired coarctation of the aorta	Other repaired lesions without residual cardiac dysfunction

talisation during the third trimester for bed rest, close monitoring and oxygen therapy of cyanotic patients.

Drugs used during pregnancy

Most commonly used cardiovascular drugs for patients with heart disease cross the placenta and expose the foetus to their pharmacological effects.²⁰ Drug effects are influenced by intrinsic pharmacokinetic properties and by the complex physiological changes occurring during pregnancy, requiring adjusting doses through pregnancy.

Some drugs also enter breast milk and may affect the neonate and infant that way. The benefits and risks for mother and foetus have to be weighed carefully. See Table 4, which shows the most common drugs used in cardiology.^{21,22}

Complications during pregnancy

Arrhythmias

Both ectopics and sustained arrhythmias become more frequent during pregnancy. In general they are treated in the same way as outside pregnancy but as conservatively as possible, although prevention of arrhythmia and avoidance of the need for anticoagulation are clearly desirable.

Antiarrhythmic drugs in pregnancy should be administered at the lowest effective dose and for the shortest duration possible. Monotherapy is also preferable.²³

Supraventricular tachycardias can be terminated by vagal stimulation, or failing that, with intravenous adenosine.²⁴ Beta blockers with beta-1 selectivity are the first choice for prophylaxis of supra- or ventricular tachycardia during pregnancy. Potentially life-threatening ventricular tachyarrhythmias are much less common and should be terminated by electrical cardioversion.

Electrical cardioversion is not contraindicated during pregnancy and should be used for any sustained tachycardia causing haemodynamic instability and thus threatening foetal and maternal well-being. Foetal heart rate must be monitored and maternal airways should be protected.

Amiodarone may be used with caution as a second line drug in cases resistant to other antiarrhythmic agents. It leads to neonatal hypothyroidism.

The presence of an implantable cardioverter-defibrillator does not itself contraindicate future pregnancies. In a series of 44 pregnancies of women with such a device the incidence of appropriate discharges was no greater during than outside pregnancy, with good outcomes for the mother and the foetus.²⁵

A pacemaker for the alleviation of symptomatic bradycardia can be implanted if needed. Echo guidance can minimise the effects of radiation.

Table 4. Drug therapy during pregnancy.

Drugs not safe	Drugs relatively safe
<p>Angiotensin converting enzyme inhibitors</p> <ul style="list-style-type: none"> ● produce foetotoxic effects principally affecting the developing foetal kidneys, especially during second and third trimester. ● should be avoided during pregnancy. <p>Angiotensin receptor blocking drugs</p> <ul style="list-style-type: none"> ● similar to Angiotensin converting enzyme inhibitors. ● should be avoided during pregnancy. <p>Amiodarone</p> <ul style="list-style-type: none"> ● neonatal hypothyroidism, prematurity and possibly neurodevelopmental problems. ● should be used as a second line drug in cases resistant to other antiarrhythmic agents whose safety has been more established. ● if used, foetal thyroid hormone levels should be monitored regularly. <p>Spironolactone</p> <ul style="list-style-type: none"> ● possible risk of anomalies of the external genitalia (animal studies only). ● if potassium-sparing diuretics are needed, amiloride is preferable. <p>Warfarin (see anticoagulation)</p> <ul style="list-style-type: none"> ● well known warfarin embryopathy syndrome. ● intracranial bleeding. 	<p>Aspirin</p> <ul style="list-style-type: none"> ● is relatively safe, and its use can prevent pre-eclampsia in high risk women. <p>Clopidogrel</p> <ul style="list-style-type: none"> ● no teratogenic effects in animal studies, ● serious complications have not been documented in case reports. <p>Diuretics</p> <ul style="list-style-type: none"> ● can be used during pregnancy, if used before pregnancy and whenever is necessary. ● not indicated in pre-eclampsia; there is some concern that their use might promote the occurrence of pre-eclampsia. <p>Beta-blockers</p> <ul style="list-style-type: none"> ● have been used extensively during pregnancy with good safety profile and no teratogenic effects. ● monitoring of foetal growth is recommended as foetal growth retardation has been described. ● can be used during breastfeeding, avoiding nursing infants at the time of peak beta-blocker plasma levels, usually occurring 3 to 4 hours after a dose. <p>Calcium channel blockers</p> <p>Digoxin</p> <ul style="list-style-type: none"> ● is considered safe through pregnancy when not exceeding therapeutic levels. ● has been considered the drug of choice in treating foetal arrhythmias. <p>Adenosine</p> <ul style="list-style-type: none"> ● treatment of choice for supraventricular tachycardia during pregnancy; short half-life. <p>Procainamide</p> <ul style="list-style-type: none"> ● can be used with relative safety to treat a variety of maternal and foetal arrhythmias. ● chronic therapy is not recommended during pregnancy because of lupus like effects. <p>Lidocaine</p> <ul style="list-style-type: none"> ● has been used as local anaesthetic during pregnancy and is relatively safe. <p>Flecainide</p> <ul style="list-style-type: none"> ● has become the treatment of choice for foetal supraventricular tachycardia. It is especially useful in treating cases refractory to digoxin and in those complicated by <i>hydrops foetalis</i>.

Cardiac arrest

There are specific issues that must be considered:²⁶

- Place women 15-30 degrees in the left lateral position.

- Chest compression should be done higher on the sternum.
- Do not use femoral lines for administration of drugs.
- Use cricoid compression before and during intubation to avoid aspiration.

- Emergency caesarean section should be started, if appropriate, as soon as cardiac arrest is confirmed.

The following should be considered: magnesium sulphate excess, eclampsia with multi-organ failure, acute myocardial infarction, aortic dissection, massive pulmonary embolism, amniotic fluid embolism, and trauma or drug overdose.

Thromboembolic disease

There is a five-fold increased risk of thromboembolic events during pregnancy in any woman and an eleven-fold increase during the puerperium, which is even greater following caesarean section. Since cyanotic heart disease is also an independent risk factor for thrombosis, pregnancy creates additional risk for these patients and prophylaxis with low molecular weight heparin is indicated in the second and third trimesters. Treatment of deep venous thrombosis with low molecular weight heparin is safe, but dose adjustment is required.

Patients with mechanical prostheses, who require anticoagulation, place themselves and the foetus at particular risk during pregnancy. Management decisions must weigh the risk and benefits of warfarin versus heparin (see below: “Anticoagulation in pregnancy”). Ideally, these issues should be considered much earlier and alternatives to a metallic prostheses should be sought.

Thrombosis of newer generation prosthetic valves, including valves in the aortic position, has been reported. Thrombolysis is recommended²⁷ but unfractionated heparin may be used initially for small non-obstructive thrombus.

Endocarditis

Infective endocarditis is uncommon in pregnancy, but when it occurs it presents difficulties in management. Antibiotics must be chosen to safeguard the life of the mother but also to try to avoid damage to the foetus. The need for surgical treatment must be weighed against the risk of foetal loss. It should not be delayed if endocarditis is life-threatening for the mother.

The American Heart Association and the European Society of Cardiology^{28,29} do not currently recommend the routine use of prophylactic antibiotics during labour for women with heart disease, but many high-risk centres practice it. Although proven benefits do not exist, the use of intravenous antibiotic prophylaxis seems reasonable in high-risk patients (such as those

with previous endocarditis or valvular prosthesis, complex cyanotic congenital heart disease, surgically created systemic to pulmonary artery shunt or conduit, bicuspid aortic valve), or those undergoing caesarean or operative vaginal delivery (forceps or ventouse extraction).

Heart failure

Normal pregnancy is accompanied by symptoms of fatigue, a decrease in exercise capacity, and dyspnoea. Many patients with heart disease who are asymptomatic may develop classic symptoms of heart failure while pregnant.

Deterioration in New York Heart Association (NYHA) functional class alone is not an indication for hospitalisation because of its subjectivity, taking into consideration the effect of general pregnancy discomfort. Augmentation of jugular venous pulsation related to increased blood volume and leg oedema (often observed in late pregnancy) could lead to an erroneous diagnosis of heart failure, or overestimation of its severity.

If heart failure is well documented, bed rest, low salt diet and the classic treatment of heart failure is recommended, including beta-blockers, digitalis and oral diuretics. Angiotensin converting enzyme inhibitors and angiotensin receptor antagonists are contraindicated during pregnancy (see “Drugs in pregnancy”). Hydralazine and nitrates can be used for this purpose.

In more severe cases, congestive heart failure will require hospitalisation with intravenous diuretic administration, plus vasodilators for further afterload reduction. In life threatening cases (see “Peripartum cardiomyopathy”) the temporary use of an intra-aortic balloon pump or left ventricular assist device may help to stabilise the patient’s condition.⁴

Systemic hypertension

Hypertensive disorders in pregnancy can be classified as:

- *Chronic hypertension.* Pre-existing hypertension, before the 20th week of pregnancy. Superimposed pre-eclampsia develops in 20-25% of women with chronic hypertension and carries risks to both mother and foetus.
- *Gestational hypertension.* Hypertension after the 20th week of pregnancy which is induced by pregnancy.
- *Pre-eclampsia.* Hypertension after the 20th week

of pregnancy which is induced by pregnancy and is accompanied by proteinuria.

Pre-eclampsia is the most common medical complication of pregnancy and is associated with substantial morbidity and mortality for both mother and baby. It can cause abruption of the placenta, pulmonary oedema, disseminated intravascular coagulation, hepatic failure and prematurity. Women should be screened regularly throughout pregnancy for pre-eclampsia (blood pressure and proteinuria) and those at high risk should be referred early for specialist care.³⁰

The primary goal of antihypertensive treatment is to prevent maternal cerebral complications. For ethical reasons there are no placebo controlled trials of pharmacological regimens.³¹ The recommended goal of therapy is the reduction of mean blood pressure to below 126 mmHg, but not less than 105 mmHg, and the diastolic pressure to between 90 and 105 mmHg.

Delivery is the only definitive treatment for pre-eclampsia and may be indicated depending on maternal symptoms. There is no evidence that any other therapy, including antihypertensive drugs, alters the underlying pathophysiology or improves outcome.

Methyldopa remains the first line agent for pre-eclampsia therapy, because of its safety and the absence of evidence of adverse effects in mothers or babies including long term paediatric follow-up. The dosage starts with 250 mg three times per day and increases progressively to a maximum of 3 to 4 g per day. Other drugs commonly used are labetalol, nifedipine, clonidine and hydralazine.

Patients with severe pre-eclampsia who are at 23 to 34 weeks of gestation should be treated with bed rest, intravenous magnesium sulphate for seizure prophylaxis, blood pressure control, foetal assessment, and corticosteroids for foetal lung maturity.

Hypertensive crisis management

Hydralazine may be given by slow intravenous injection of 5 mg, which can be repeated after 20 min; labetalol or nifedipine can also be used; in rare cases there may be a need for nitroglycerine or nitroprusside.

Magnesium sulphate has been demonstrated to be more effective than nimodipine for prophylaxis against seizures in women with severe pre-eclampsia.

Cardiac surgery during pregnancy

Cardiac surgery during pregnancy should be reserved for patients refractory to medical management in whom

further delay would prove detrimental to maternal health. Maternal mortality is similar to that for non-pregnant women. Foetal outcome is significantly influenced, with foetal loss 30%. The complexity of an operation and the duration of cardiopulmonary bypass (CPB) directly affect foetal mortality. Whenever possible, delivery should be performed through caesarean section after heparinisation and cannulation of the mother.

If cardiac surgery is required, monitoring of the foetus and uterine activity should be performed during CPB. Uterine contractions during CPB are considered to be the most important predictor of foetal death, and are more prone to occur during rewarming. Preparation and management should be oriented towards the avoidance of any sudden changes of maternal flow.

Whenever possible, normothermic CPB should be first choice for a pregnant patient.³²

Delivery and post-partum

Labour and delivery must be planned carefully and well in advance. Management of intrapartum care should be supervised by a team experienced in the care of women with cardiac disease (obstetrician, anaesthetist, midwife) with a cardiologist readily available.

Maternal monitoring during labour should be individualised, and usually includes continuous electrocardiographic monitoring and pulse oximetry, and occasionally invasive blood pressure recording.

The principle is to manage the stress of labour in such a way that it does not exceed the woman's capacity to cope with it.

Vaginal delivery carries about half the risk of complications of an elective caesarean section, for both the mother and the foetus. However, prolonged and difficult labour should be avoided, and detailed continuous monitoring of the foetus and the mother are mandatory.

The threshold for assisted delivery, by either vacuum or forceps, should be low, in order to limit or avoid active maternal "bearing down" and a prolonged second stage of labour. Labour should not be induced unless for obstetric reasons or because of cardiovascular deterioration. Spontaneous labour is usually quicker than induced and carries a higher chance of successful delivery.

Early epidural analgesia with a cardiostable drug at low dose is important. For patients on anticoagulation, even at prophylactic doses, care needs to be taken during regional anaesthesia, to minimise the risk of neuroaxial haematoma.

Oxytocic drugs such as oxytocin and ergometrine, which improve uterine contraction, also have major haemodynamic effects. Continuous infusion of oxytocin, at the lowest effective rate, in low volume fluid, has minimal cardiovascular effects and avoids the circulatory overload.

For most cardiac conditions a normal vaginal delivery, with good analgesia and a low threshold for forceps assistance, is the safest mode of delivery for the mother, since it is associated with less rapid haemodynamic changes, and a lower risk of haemorrhage and infection.³³ However, in some conditions caesarean section is indicated (Table 5).

Ongoing monitoring during the puerperium and post-partum period is necessary in high risk patients. Coronary unit monitoring should be used if necessary, especially in cyanotic patients and those with pulmonary hypertension, where the risk of maternal death remains high for up to 10 days. Most deaths in Eisenmenger patients have been reported to occur in the first or second week after delivery.¹⁹

Specific heart conditions

Congenital heart disease

Left to right shunts

The effect of the increased cardiac output on the already volume-overloaded right and left ventricle in patients with an atrial septal defect is counterbalanced by the decrease in peripheral vascular resistance. In the absence of pulmonary hypertension, pregnancy, labour and delivery are well tolerated and maternal and foetal complications are probably similar to those occurring in the normal population. Infrequently, among patients with an atrial septal defect, paradoxical embolisation may be encountered if systemic vasodilation and/or elevation of pulmonary resistance promote a transient right to left shunt.

Patients with restrictive ventricular septal defect and with small patent *ductus arteriosus* also tolerate pregnancy well. There are few data on the effects of

pregnancy in patients with atrioventricular septal defects. Although physiologically they behave similarly to other left to right shunts, a recent study examining the outcome of 62 pregnancies in 29 women with atrioventricular septal defects reported that pregnancy is not always well tolerated, predominantly because of a deterioration in NHYA class (23%) and worsening of pre-existing atrioventricular valvular regurgitation (17%). Offspring mortality was high (6.3%), primarily due to recurrence of complex congenital heart disease.³⁴

Stenotic lesions

Congenital aortic valve disease

Congenital bicuspid aortic valve is the most common cause of aortic valve stenosis that complicates pregnancy. Women with symptomatic aortic stenosis should delay pregnancy until after surgical repair. Women with moderate to severe aortic stenosis may develop heart failure, even if symptoms are not present prior to pregnancy. Early studies of critical aortic stenosis shared high mortality rates (17%).³⁵ Lower mortality and morbidity have been reported in recent studies, however, presumably reflecting better maternal care.³⁶

The non-pregnant Doppler gradient across the valve may double during pregnancy, merely as a result of the physiological changes. An apparent increase of the pressure gradient during pregnancy should not necessarily cause alarm. In contrast, absence of such an increase can be falsely reassuring, since it may herald left ventricular dysfunction or failure.

Cardiac surgery is indicated when there is clinical deterioration; successful palliation during pregnancy by balloon valvuloplasty has been reported.³⁷

Aortic stenosis is an adverse risk factor for foetal outcomes (see "Risk to the foetus").

Coarctation of the aorta

Maternal mortality with unrepaired coarctation of the aorta has been estimated at 3%. The main complications reported relate to severe hypertension, including aortic dissection. Changes in the aortic wall during gravid status increase the pre-existing risk of dissection in coarctation of the aorta.

There was a 30% incidence of hypertension during pregnancy amongst 50 women with coarctation of the aorta and one death in a patient with Turner syndrome (died of Type A dissection).³⁸

Coarctation of the aorta after repair is considered a

Table 5. Indications for caesarean section other than obstetric reasons.

- Marfan syndrome with aortic root dilatation >45 mm.
- Therapeutic treatment with warfarin at the onset of partum.
- If there is abrupt haemodynamic deterioration.
- Some authors recommend caesarean section in women with severe pulmonary hypertension or severe aortic stenosis.³⁶

low risk lesion. During pregnancy, following coarctation repair, the risk of dissection and rupture is likely to be reduced but not eliminated, especially in those with aneurysm at the site of repair. In a recent series of pregnancies after coarctation repair there were no serious cardiovascular complications, but the incidence of hypertension was still 22%.³⁹

Pulmonary stenosis

Mild to moderate pulmonary stenosis is well tolerated during pregnancy and foetal outcome is favourable. Pregnancy in women with severe pulmonary stenosis may precipitate right heart failure or atrial arrhythmia. Balloon valvuloplasty may be required and yield a good result for patients with valvar stenosis when symptoms of pulmonary stenosis develop or progress.⁴⁰

Unrepaired cyanotic conditions (without Eisenmenger physiology)

The maternal risks associated with cyanosis during pregnancy vary depending on the aetiology of the cyanosis, with a much higher rate of success if there is no pulmonary hypertension. Mortality during pregnancy in women with cyanosis without pulmonary hypertension is reported as 5%.

A large case series examining the outcome of 96 pregnancies in 44 women without Eisenmenger physiology reported a high rate of maternal cardiac events. One death occurred and cardiovascular complications were reported in 32% of the patients. Miscarriage, prematurity and low birth weight were all associated with cyanosis. The incidence of foetal complications was high, with a low live birth rate (43% of the pregnancies) and 37% premature births. The lowest live birth rate (12%) was observed in those mothers with an arterial oxygen saturation of <85% at the beginning of pregnancy. It has been suggested that bed rest and O₂ administration can improve foetal outcome.⁴¹

Repaired cyanotic conditions

Tetralogy of Fallot

Tetralogy of Fallot is the most common cyanotic condition, currently with a large number of patients who have reached reproductive age after successful repair during infancy. Classically, repaired tetralogy of Fallot has been considered as a low risk lesion in terms of both maternal and foetal outcomes, with no maternal deaths being reported.

Recently, concerns have been raised regarding the potential effect of pregnancy on right ventricular dilatation and function. In a recent retrospective study (43 pregnancies) that sought to determine outcomes in patients with tetralogy of Fallot, most of them repaired, 7% of the women had adverse cardiovascular events, including supraventricular tachycardia, heart failure, pulmonary embolism, and progression of right ventricular dilatation. Patients who had cardiovascular complications during pregnancy also had severe right ventricular dilation or right ventricular dysfunction. The same study also reported a higher rate of spontaneous foetal loss, compared with the expected average.⁴²

Children of mothers with tetralogy of Fallot are more likely to have congenital heart disease, with reported incidence approximately 3%. It is believed that 15% of patients with tetralogy of Fallot have DiGeorge syndrome, which is inherited with autosomal dominant pattern.⁴³

Pulmonary atresia

In an early retrospective study evaluating the outcome of pregnancy in patients with pulmonary atresia, comparing those with and without previous radical surgical repair, there were significant complications even in the repair group. Patients with residual systemic to pulmonary collaterals and those with elevated right ventricular pressures appeared to be at particularly high risk.⁴⁴

Systemic right ventricle

Both in the atrial switch operation (Mustard or Senning procedure) for repair of transposition of the great arteries, and in congenitally corrected transposition of the great arteries, the anatomic right ventricle supports the systemic circulation.

Transposition of great arteries with atrial switch

Cardiovascular complications are largely related with the presence of a systemic right ventricle. In women without systemic ventricular dysfunction and in NYHA classes I and II, pregnancy is well tolerated.⁴⁵ However, there are legitimate concerns of irreversible systemic ventricular dysfunction and dilatation, even in these women.⁴⁶

Transposition of great arteries with arterial switch

In patients with previous arterial switch for transposi-

tion of the great arteries little is known about their ability to cope with pregnancy, but it is fair to say that no major difficulties are anticipated.

Congenitally corrected transposition of the great arteries

In a study with 41 patients, there were 105 pregnancies with 73% live births and no maternal mortality. In most patients who developed complications the diagnosis was established during pregnancy.⁴⁷

Ebstein's anomaly

For the majority of patients with Ebstein's anomaly, pregnancy is well tolerated. However, increased risk of atrial arrhythmia can be anticipated and higher rates of foetal loss have been reported. Pregnancy is associated with more maternal complications among patients with Ebstein's anomaly when resting cyanosis is present prior to pregnancy.

Univentricular physiology with or without the Fontan operation

Overall, the risk for patients with single ventricle physiology (double-inlet ventricle, atrioventricular atresia and atrial isomerism) palliated with arterial shunt is related with the presence of generic risks for pregnancy. If pulmonary arterial hypertension is present the risk is prohibitive. Successful pregnancy under proper cardiologic and obstetric supervision has been reported.

The Fontan operation

This procedure eliminates cyanosis and partially reverses volume overload of the functioning single ventricle. Patients have a limited ability to increase cardiac output during pregnancy, however.

There is limited information about pregnancy and the Fontan operation. In the only series reported, in 21 women with 33 pregnancies, 15 resulted in live births, 5 in elective therapeutic abortions and 13 in spontaneous abortions during the first trimester. There was no maternal mortality in these 21 patients, although they probably represent a highly selected population.⁴⁸

If ventricular dysfunction or obstruction of the Fontan circulation exists, the risk of pregnancy is much higher. Atrial arrhythmias tend to develop or worsen, with a rate of 25% sustained supraventricular arrhythmias being reported in a recent review of the literature.⁴⁹

Right atrial thrombus formation may occur, with a risk of paradoxical embolism if the Fontan is fenestrated or following Fontan failure. Fontan patients with a large right atrium and venous congestion have to be monitored very carefully. It may be that such patients should be considered for conversion to total cavopulmonary connection before they embark on pregnancy.

The successful Fontan patient with a small right atrium or total cavopulmonary connection, in functional class I or II with good left ventricular function, can probably complete pregnancy with a normal live birth at a relatively low risk. The need for anticoagulant treatment has to be considered and plans should be made before pregnancy.

Marfan syndrome

Eighty percent of patients with Marfan syndrome have some cardiac involvement, including mitral valve prolapse and aortic root dilatation and dissection. Pregnancy is a high risk endeavour for affected females, as it is associated with a higher risk of dissection, occurring most often in the last trimester or the early post-partum period.⁵⁰

Full assessment should be performed before pregnancy. The risk is lower for pregnancy following elective aortic root replacement. A number of patients have had successful full term pregnancies without complications following elective aortic root replacement.

Women with minimal cardiac involvement (aortic root less than 40 mm, and without previous dissection, and no significant aortic or mitral regurgitation) should be informed of a 1% risk of aortic dissection during pregnancy.⁵¹ Patients with an aortic root diameter more than 40 mm should be told that the risk of dissection during pregnancy may be as high as 10%.

Pregnancy needs to be monitored by echocardiography at regular intervals throughout pregnancy and for six months post-partum. Beta-blocker therapy should be continued. If the aortic root diameter is 4.5 cm or greater caesarean delivery is advisable. If normal delivery is planned the second stage should be expedited. The woman may be allowed to labour on her left side or in a semi-erect position to minimise stress to the aorta.

Aortic dissection type A requires emergency cardiac surgery. Acute dissection type B should be managed medically with the same indications for surgery in non-pregnant women.

Hypertrophic cardiomyopathy

Asymptomatic patients with hypertrophic cardiomyopathy usually tolerate pregnancy well, as their left ventricle seems to adapt in a physiological way. Nevertheless, pulmonary congestion or even sudden pulmonary oedema, mostly commonly occurring in the peripartum have been described. In the largest series to date of 127 patients with 271 pregnancies, there was no death. The incidence of heart failure was 2%.⁵²

Beta-blockers should be continued during pregnancy. Normal delivery should be conducted with continuation of beta-blocker therapy and avoidance of systemic vasodilation.

Dilated cardiomyopathy

Occasionally dilated non-peripartum cardiomyopathy is documented before pregnancy. Provided that patients have completely or almost completely recovered their ventricular function, maternal risks should be low.

Peripartum cardiomyopathy

This is defined as unexplained left ventricular systolic dysfunction that develops in the last prenatal month or within six months of delivery. Most severe cases tend to develop and/or present with overt heart failure during the first few days post-partum. The pathogenesis is poorly understood; some form of myocarditis has been postulated.

Women who have developed peripartum cardiomyopathy usually present with heart failure and marked fluid retention. Treatment is the same as for any form of cardiomyopathy with impaired left ventricular function. Failure may be fulminant and may require inotropes, a ventricular assist device or even transplantation.

As in acute myocarditis outside pregnancy the most fulminant cases show the largest capacity for improvement and in them the use of a device as a bridge to recovery is particularly appropriate. Maternal mortality is as high as 20%.

If left ventricular dysfunction persists a year after delivery, the risk of death in a subsequent pregnancy is approximately 20%. There is also a substantial risk of recurrence of symptomatic heart failure and permanent impairment of left ventricular function in any subsequent pregnancy. Pregnancy should therefore be discouraged.⁵³

Acquired heart disease

Coronary artery disease

The incidence of acute coronary syndromes during pregnancy has been reported as 6.2 per 100,000 in the US. The principal risk factors were advanced maternal age, classic risk for atherosclerosis, anaemia, and transfusion during delivery.⁴

As the average maternal age increases and assisted reproductive technology allows for even more advanced maternal age, the incidence of myocardial infarction during pregnancy may also increase.⁵⁴ Acute myocardial infarction during pregnancy and puerperium is associated with significant maternal and foetal mortality, reported as between 5.7% and 37%.

Although atherosclerosis is often documented, coronary dissection with angiographically normal arteries is also common. A recent study of 859 pregnant women, in whom coronary angiography was performed in 51% of cases, showed atherosclerosis in 43%, thrombus in 21%; 29% had normal coronary arteries and 16% had coronary dissection.⁵⁵ Coronary dissection in otherwise healthy women occurs mostly near term or within three months post-partum. The dissection involves the left descending coronary artery in approximately 80% of cases. The overall mortality is of the order of 30 to 40%. Early diagnosis is often hindered by the normal changes of pregnancy and low level of suspicion.^{56,57}

Treatment of acute myocardial infarction

Diagnostic delay and no therapy administered are the main factors leading to poor outcomes of acute myocardial infarction during pregnancy.⁵⁸

Fibrinolytic drugs have been given during pregnancy for several indications, with no evidence of teratogenicity. Although most reported cases were associated with favourable maternal and foetal outcomes, therapy is associated with a risks of maternal haemorrhage, especially when thrombolytic agents are given at the time of delivery.

Due to the extension of myocardial infarction, which is most frequently anterior or anterolateral, and the relative contraindication for fibrinolytic agents, primary angioplasty can be the safest therapy for the woman and consequently for the foetus, especially after the second trimester. A radial approach has been suggested as safer in terms of radiation. The recent consensus document of the European Society of Cardi-

ology recommended primary angioplasty for the treatment of acute myocardial infarction in pregnancy.⁵⁹

Acquired valvular heart disease

Mitral stenosis

Rheumatic mitral stenosis is the most common clinically significant valvular abnormality in pregnant women. An increase in maternal heart rate decreases the diastolic filling period, further increasing left atrial pressures. Surgery or valvuloplasty has to be considered in all symptomatic patients before pregnancy, or those asymptomatic with severe mitral stenosis who contemplate pregnancy.⁶⁰ Close follow-up is necessary in every pregnant woman with significant mitral stenosis, even if she was totally asymptomatic before pregnancy, with echocardiographic assessment of mean transmitral gradient and pulmonary artery pressure.

Beta-blockers should be started in patients who have symptoms or estimated systolic pulmonary arterial pressure more than 50 mmHg, as well as low salt diet and reduction of physical activity. Diuretics need to be added if signs of pulmonary congestion develop or persist.

In patients who present with severe symptoms during pregnancy despite medical therapy, percutaneous balloon mitral valvuloplasty has to be considered. Performed during the second trimester, it has been associated with normal subsequent deliveries and excellent foetal outcomes. Published series comprise more than 250 valvuloplasties during pregnancy. One study compared percutaneous balloon mitral valvuloplasty with open mitral valve commissurotomy and showed that valvuloplasty is a better option, reducing foetal and neonatal mortality significantly.⁶¹ Appropriate anticoagulation should always be in place.

Aortic stenosis

See "Congenital aortic valve disease".

Regurgitant lesions

The fall in systemic vascular resistance reduces the regurgitant volume in mitral and aortic regurgitation. In aortic regurgitation the shortening of diastole due to tachycardia also contributes to the regurgitant volume reduction. Pregnancy is frequently well tolerated, even in patients with severe regurgitant lesions, if there is no left ventricular dysfunction and the patient is asymptomatic before pregnancy.^{62,63}

Mitral regurgitation

In young women this is most commonly due to mitral valve prolapse and is usually well tolerated during pregnancy.

Aortic regurgitation

Aortic regurgitation in young women may be congenital, and so due to a dilated aortic annulus (Marfan or bicuspid aortic valve), or acquired and mainly due to previous endocarditis or rheumatic fever.

Prosthetic valves

Mechanical prosthetic valves

Ideally, women of reproductive age with valvular heart disease who are undergoing surgery should have a tissue rather than a metallic prosthesis. The risk associated with pregnancy in women with mechanical prosthetic valves is related mainly to the increased incidence of thrombosis. Effective anticoagulation is critical in patients with mechanical prosthetic valves, but remains problematic because both oral anticoagulation and heparin have been associated with important maternal and foetal side effects.

Whatever the anticoagulation regimen, pregnancy in a patient with a mechanical prosthesis is associated with a maternal mortality between 1 and 4%. Foetal outcome is affected by the type of anticoagulation and the status of the mother, with a foetal loss rate of 20%, but a worse outcome is seen when warfarin is used. Heparin does not cross the placenta and is therefore safe for the foetus. However, long-term heparin therapy during pregnancy is difficult to manage and is reported to be less effective, significantly increasing the thromboembolic risk for the mother.

Anecdotally, a combination of high dosage of low molecular weight heparin with aspirin has been effective in some patients with a mechanical prosthesis (see below). The choice of anticoagulation needs to be made after detailed discussion with the patient of the risks inherent in the different anticoagulation modalities.^{64,65}

Warfarin

The use of warfarin, particularly between the 6th and 12th weeks of pregnancy, is associated with warfarin embryopathy. This is characterised by nasal hypopla-

sia and/or bone stippling. Its use during pregnancy may also be associated with intracranial bleeding and central nervous system abnormalities. In the main meta-analysis examining cases where warfarin was used throughout pregnancy the embryopathy risk was 6.4%, and not 30% as previously reported.⁶⁶ The risk has been thought to be dose related. It has been reported that warfarin risk embryopathy is low in women who needed less than 5 mg of warfarin to maintain an adequate international normalised ratio.⁶⁷

Warfarin should be replaced by unfractionated heparin from week 36 of the pregnancy. The international normalised ratio should be maintained at pre-pregnancy levels.

Unfractionated heparin (UFH)

If heparin is used during pregnancy the risk of embryopathy is 0% but maternal death is 6.7% with adjusted doses. In addition to the discomfort of its administration, heparin risks include thrombocytopenia and osteoporosis. Subcutaneous heparin during the first trimester is associated with a high incidence of thromboembolic complications, particularly prosthetic thrombosis, and maternal death.

The level of anticoagulation with heparin treatment should be closely monitored and an activated partial thromboplastin time of 2.5 times normal should be achieved. Resistance to moderate doses of UFH in high-risk women with old-generation prosthetic valves has been reported.

If subcutaneous UFH is used, it should be started in high doses (17,500 to 20,000 U) and adjusted to prolong a 6-hour post-injection activated partial thromboplastin time to within the therapeutic range.

Low molecular weight heparin (LMWH)

In a non-pregnant status LMWH is superior in some ways to UFH. LMWH does not cross the placenta barrier and it offers potential advantages, including fewer bleeding complications, a lower frequency of heparin-induced thrombocytopenia (although heparin-induced thrombocytopenia seems to be rare in pregnancy), a lower incidence of osteoporosis, superior subcutaneous absorption and bioavailability, a longer half life, and a more predictable dose response. A lower rate of spontaneous abortion in patients with prosthetic heart valves has been shown when LMWH was used compared with UFH.

Some concerns have been raised about the use of

LMWH during pregnancy. A recent report cited a high incidence of valve thrombosis related to the use of LMWH, with higher mortality rates. A careful review of the reported cases, however, indicates that most, if not all of them were associated with an inadequate dose, lack of monitoring, or sub-therapeutic anti-Xa levels.⁶⁸

Because of changes in both intravascular volume and body gain during pregnancy, administration of LMWH on the basis of weight alone is inadequate, and measurement of anti-Xa activity is necessary to ensure effective anticoagulation. The most recent recommendations of the American College of Chest Physicians call for the use of LMWH given twice daily to maintain four-hour post-injection anti-Xa levels of around 1 U/ml.⁶⁹

Recent data suggest that such peak levels were associated with sub-therapeutic trough levels of less than 0.5 U/ml in the great majority of cases, and mention the importance of routine measurements and maintenance of trough levels within a therapeutic range (0.6 to 0.7 U/ml). Peak levels should also be monitored to prevent excessive anticoagulation (i.e. >1.5 U/ml), in which case an 8-hourly rather than 12-hourly dosing should be used. To ensure patient compliance and adequate prophylaxis, anti-Xa activity or activated partial thromboplastin time should be measured at least every 2 weeks.

The European Consensus does not approve the use of LMWH during pregnancy.⁵⁹

To summarise:

- A completely safe treatment for pregnant women who have a mechanical prosthesis does not exist.
- The European Consensus and the American College of Cardiology guidelines recommend the use of warfarin through pregnancy given the high reported mortality with UFH and the impression that warfarin embryopathy has been overstated. However, there is an underreported incidence of intracranial bleeding.
- If women do not accept the risks of warfarin, heparin has been proposed during the first trimester, with a change to warfarin until the 35th week of pregnancy.
- Any anticoagulation therapy must be replaced by UFH from the 36th week of gestation.
- The use of LMWH during pregnancy is not officially approved; there is a lack of data. If it is used without dose control the risk of thrombosis is very high and the use of concomitant aspirin should be considered.

Delivery and anticoagulation

Because of the high incidence of premature labour in women with prosthetic valves, warfarin should be replaced by heparin at the 36th week to avoid the risk of foetal intracranial haemorrhage. If labour begins during treatment with warfarin, caesarean section should be performed.

Anticoagulation also has implications for analgesic or anaesthetic options for delivery, since epidural or spinal techniques may carry a risk of intraspinal bleeding, depending on the dose and the timing of anticoagulant treatment. Catheter placement for epidural anaesthesia is not advisable within 10 to 12 hours of the last dose of LMWH, which should be withdrawn 18 hours before elective delivery and replaced by intravenous UFH.

Heparin should be withdrawn 4 hours before caesarean section or at the onset of labour. In the absence of significant bleeding, heparin can be resumed 4 to 6 hours after either surgical or vaginal delivery, and warfarin begun orally.

Biological prosthetic valves

These valves eliminate the need for anticoagulant therapy associated with mechanical prostheses.

Degeneration of bioprosthetic valves during pregnancy has not been confirmed, although most available data may support accelerated structural valve degeneration. In the aortic position it seems that homografts offer longer durability than stented porcine xenografts.

Pregnancy after the Ross procedure seems to be safe without maternal complications.⁶⁴

Pulmonary hypertension

Pulmonary vascular disease is the worst risk factor for pregnancy and the post-partum. Inability to decrease pulmonary vascular resistance to compensate for the volume overload exposes pregnant women with pulmonary hypertension to a high risk.⁷⁰ Despite improvements in medical therapy, rates of mortality are 30% in idiopathic pulmonary arterial hypertension, 50% in those associated with collagen diseases, and 30 to 50% in Eisenmenger syndrome.

In Eisenmenger syndrome, while systemic vascular resistance falls to compensate for the increased cardiac output, pulmonary resistance does not, leading to an increase in right to left shunting, cyanosis and heart

failure. Added to this is the risk of thromboembolic disease, bleeding and sudden death.⁷¹ Most deaths occur at term or during the first post-partum week. There is also little doubt about the high risk to the foetus.⁷²

In cases of mitral stenosis with secondary pulmonary arterial hypertension, the lower degree of pulmonary hypertension and the availability of mitral valvuloplasty may contribute to a more favourable outcome.

Conclusion

Ideally, timely pre-pregnancy counselling should be offered to all women with heart disease so as to allow patients to come to fully understand their childbearing potential. Preventing life-threatening cardiovascular complications with optimal care during pregnancy, delivery and the post-partum often requires a multidisciplinary team approach involving cardiologists, obstetricians and anaesthetists.

Conditions such as acute myocardial infarction require prompt catheter intervention. Areas such as anticoagulation for patients with mechanical prosthetic valves remain problematic, and preference should be given to tissue valves.

Successful pregnancy is feasible for most women with heart disease, whether congenital or acquired, and when optimal care is provided it can be undergone with relatively low risk.

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