

Management of heart disease in pregnancy



Dr Eileen Bass
Obstetric Physician
National Women's Health
Auckland Hospital

Cardiac disease affects only one to two per cent of all pregnancies but is a leading cause of maternal morbidity and mortality. Congenital heart disease is now commonly encountered in pregnancy, as advances in medical and surgical treatment have led to the improved survival and wellbeing of young women with congenital heart disease.

However, rheumatic heart disease remains prevalent in many parts of the world, particularly amongst the Pacific and indigenous communities of New Zealand and Australia. In addition, the older age of pregnant women, continued prevalence of smoking and the emerging obesity epidemic is further contributing to the incidence of acquired heart disease in pregnancy. Pregnant women with cardiac disease require careful

assessment and optimisation of cardiac status to ensure successful pregnancy outcome for mother and baby. Ideally, all women with cardiac disease would receive pre-pregnancy counselling, along with adequate contraceptive advice. This article covers important aspects of acquired and congenital heart disease in pregnancy.

Cardiovascular changes

Knowledge of the significant physiological changes of pregnancy is required to predict the effect of pregnancy on women with underlying cardiac disease. Cardiovascular changes occur from four to six weeks gestation, peaking in the second trimester. A 40 per cent increase in cardiac output is due to increased blood volume, along with decreased systemic and pulmonary vascular resistance and increased heart rate.

Labour and delivery result in a further increase in cardiac output. Maternal position is important – there can be a 25 to 30 per cent fall in cardiac output when supine, from compression of the vena cava by the gravid uterus. Maternal valsalva, sympathetic stimulation from pain causing hypertension and tachycardia, and 'auto transfusion' of myometrial blood with uterine contraction, also cause fluctuations in cardiac output. Caesarean section is also associated with significant cardiovascular changes.

The first 24 to 72 hours post delivery are associated with rapid intravascular volume shifts and many women with cardiac disease may present for the first time postpartum.

Predicting risk

Certain cardiac conditions poorly tolerate the cardiovascular changes of pregnancy. These include pulmonary hypertension, poor ventricular function, stenotic valvular lesions, aortic pathology and ischaemic heart disease. In one study, stroke, arrhythmia, pulmonary oedema and death complicated 13 per cent of high-risk pregnancies. The best predictors of poor outcome in pregnancy are:

- poor functional class (poor exercise tolerance);
- previous cardiac event (arrhythmia, pulmonary oedema, stroke);
- left heart obstruction.

Neonatal outcome correlates best with maternal outcome, particularly if cardiac compromise results in poor fetal growth or necessitates premature delivery.

Management in pregnancy

Ideally, women with cardiac disease are managed by a multidisciplinary team. This includes:

- a cardiologist with expertise in pregnancy or obstetric physician;
- an obstetrician or fetal medicine specialist;
- midwives with experience in medical problems;
- an obstetric anaesthetist; and
- a neonatal paediatrician.

In a rural setting, it may be necessary to refer to the regional tertiary centre for ongoing management, or if appropriate, review and advice regarding shared care.

Careful history, examination and investigations should document functional status and assess whether pregnancy-related cardiovascular changes are likely to be tolerated. The role of ECG, 24-hour holter monitor, echocardiography, cardiac MRI and exercise testing will depend on the nature of the underlying cardiac problem. Maternal medications need to be reviewed. Many cardiac medications are compatible with pregnancy although the use of warfarin, ACE inhibitors and amiodarone are of particular concern.

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Regular antenatal visits to monitor maternal and fetal wellbeing are essential. There should also be attention to non-cardiac aspects of pregnancy, such as advice against smoking, detection of diabetes and monitoring for pre-eclampsia. Timing of delivery may need to be planned. Vaginal delivery is usually indicated. Lower segment caesarean section (LSCS) should be for obstetric or fetal indications and for cardiac reasons in certain conditions only. Prolonged labour should be avoided and careful, regional anaesthesia can help ameliorate the cardiovascular changes associated with pain and anxiety. Close monitoring should occur, with particular care around the administration of intravenous fluids and medications and the timing of anticoagulation. The need for invasive cardiac monitoring will depend on the severity of the maternal cardiac condition.

Antibiotic prophylaxis for endocarditis should follow local guidelines. Avoidance of significant postpartum haemorrhage is

important. Oxytocin should be used with care to avoid hypotension – ergotamine should be avoided. The haemodynamic changes of pregnancy persist for a number of days to weeks post delivery and ongoing regular review by clinicians should occur. Postpartum thrombo-prophylaxis should also be considered.

Acquired heart disease

Rheumatic valvular heart disease

In general, women tolerate regurgitant valvular lesions better than stenotic lesions. Severe mitral stenosis (valve area < 1 cm²) is associated with a high risk of deterioration in pregnancy. Tachycardia, increased blood volume and increased cardiac output lead to raised left atrial pressures and risk of atrial fibrillation and pulmonary oedema. Treatment includes beta blockers and diuretics. Percutaneous balloon valvotomy is preferable to surgical valvotomy. Surgery is associated with an increased risk of fetal and maternal morbidity and mortality (15 to 30 per cent fetal mortality and five per cent maternal mortality).

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Significant aortic stenosis can be life-threatening due to cardiac ischaemia and associated arrhythmias (for example, ventricular tachycardia). Pre-pregnancy function is not predictive and surgical treatment is often required with the associated maternal and fetal risks.

Prosthetic cardiac valves

Prosthetic heart valves are problematic in pregnancy. These women generally have good cardiac function so rarely develop cardiac compromise. Tissue valves avoid the issues surrounding anticoagulation in pregnancy and should be the valve of choice in young women. The optimal management of women with mechanical heart valves in pregnancy is controversial. Warfarin is best for the mother, as the pro-coagulant state of pregnancy confers an increased risk of valve thrombosis and therefore stroke, valve failure with subsequent cardiac compromise, and even death. The highest risk is associated with the older mechanical valves (for example, Starr Edwards), mitral position, atrial fibrillation, more than one mechanical valve and previous thrombotic complication. Warfarin crosses the placenta and is associated with an increased risk of miscarriage. There is a four to eight per cent incidence of embryopathy with warfarin use between six to 12 weeks gestation and an increased rate of fetal loss in the second and third trimesters due to intra-cerebral haemorrhage from fetal anticoagulation. Heparins do not cross the placenta but are associated with increased risk of valve thrombosis for the mother. The safest option for the mother is to continue warfarin through the pregnancy.

Other management options include replacing warfarin with therapeutic doses of low molecular weight heparin (LMWH) for the first trimester, returning to warfarin for the second and third trimester, or continuing LMWH throughout pregnancy. Anti-Xa levels should be monitored if using LMWH and adjunctive aspirin is recommended with all regimens. Warfarin needs to be stopped

and substituted for heparin at least ten days prior to delivery to allow clearance of warfarin from the fetal circulation. Heparin anticoagulation needs to be interrupted at delivery for safety of the mother. Balancing the risks of obstetric haemorrhage against the risks of thrombosis peripartum is challenging. Clearly, management of mechanical heart valves in pregnancy is an extremely high-risk situation and clinicians with expertise and knowledge in this area should be involved. Avoidance of pregnancy is the safest option.

Peripartum cardiomyopathy

Peripartum cardiomyopathy is the development of congestive heart failure in the last month of pregnancy or in the first five months postpartum. It is a diagnosis of exclusion. Typically, the left ventricular ejection fraction (LVEF) is less than 45 per cent with global dilatation. Risk factors include multiparity, increased age, obesity, pre-existing hypertension and pre-eclampsia. Standard treatment for heart failure should be instituted including anticoagulation, as there is a high rate of thrombo-embolism. Peripartum cardiomyopathy is associated with a seven per cent mortality. Failure to return to normal left ventricular (LV) function within six months is a poor prognostic sign and contraindication to further pregnancies.

Arrhythmia

Benign arrhythmias such as ventricular and atrial ectopy are common in pregnancy and usually require no treatment. More serious arrhythmias can present for the first time or be exacerbated by pregnancy. Treatment is usually the same as for non-pregnant women. In particular, intravenous adenosine and cardioversion are considered safe.

Ischaemic heart disease (IHD)

Acute myocardial infarction is a rare complication of pregnancy and is associated with significant maternal mortality. With increasing maternal age, obesity, diabetes and current smoking rates, the incidence is likely to increase. Coronary atherosclerosis is usually the main cause, although coronary artery dissection is often seen. Illicit drug use needs to be considered, along with congenital coronary anomalies. Management is similar as for non-pregnant women but will depend on gestation. Primary percutaneous coronary angioplasty will be preferable to thrombolysis in the peripartum period.

Congenital heart disease

Women with repaired, non-cyanosed congenital heart disease and good functional status are likely to do well in pregnancy. Predictors of poor outcome are well documented:

- pulmonary hypertension;
- maternal cyanosis;
- poor functional class (poor systemic ventricular function);
- maternal arrhythmias; and
- need for maternal anticoagulation.

Women with congenital heart disease may have limited cardiac reserve and poorly tolerate the cardiovascular changes of pregnancy. There is also an increased rate of thrombo-embolic complications. Maternal cyanosis is strongly associated with increased rates of fetal mortality, poor fetal growth and premature delivery. Oxygen saturations less than 85 per cent are associated with a 12 per cent live birth rate, compared with 92 per cent live birth rate when maternal oxygen saturations are greater than 90 per cent. Fetal morbidity and mortality can occur with maternal haemodynamic compromise from arrhythmias, hypotension and cardiac surgery.

There is also the issue of inheritance of congenital heart disease. The risk of inheritance varies from two to seven per cent depending on the specific maternal (or paternal) lesion and is not concordant. Routine fetal echo should be performed around 20 to 22 weeks gestation.

Atrial septal defects (ASD) and Ventricular septal defects (VSD)

Repaired ASD and VSD are common and are usually considered low-risk in pregnancy. Women with unrepaired ASD/VSD with left to right shunts generally tolerate pregnancy well. There is an increased incidence of atrial arrhythmias and a risk of paradoxical emboli. Emboli from leg or pelvic veins can cross to the systemic circulation via the defect and cause stroke. Low dose aspirin is often used. Large shunts with raised pulmonary pressures present increased risks of cardiac compromise. In particular, acute blood loss or fall in systemic pressures will exacerbate the left to right shunt. Echo is important to document degree of shunt, ventricular function and pulmonary pressures.

Eisenmenger's syndrome

When pulmonary vascular pressures supersede systemic vascular pressures, reversal of the shunt occurs resulting in cyanosis. Eisenmenger's syndrome is associated with a 50 per cent mortality in pregnancy despite intensive medical and obstetric care. Death usually occurs postpartum but can occur at anytime during pregnancy and even potentially at termination of pregnancy – so contraceptive counselling is vital. Death is usually due to thrombo-embolism or volume depletion exacerbating the shunt, resulting in worsening cyanosis and syncope. Pre-eclampsia can cause dramatic deterioration in cardiac function resulting in death. There is a high rate of fetal loss with a reported 30 per cent perinatal mortality.

Marfan's syndrome

Pregnancy is associated with an increased risk of aortic dissection and rupture. Serial echocardiography should be performed in pregnancy and prophylactic beta blockers considered. Pregnancy should be avoided if the aortic root is greater than 4.5 cm. Elective LSCS is usually recommended if the aortic root is greater than 4 cm or there is evidence of dilatation with pregnancy. Marfan's syndrome is an autosomal dominant condition, with a 50 per cent risk of inheritance.

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Complex congenital heart disease

There are many other types of congenital heart disease and various surgical repairs. In pregnancy, the underlying cardiac lesion should be documented with echo or MRI, along with systemic ventricular function, degree of shunt and pulmonary pressures. Maternal functional status should be assessed in conjunction with oxygen saturations, haemoglobin and documentation of previous cardiac events (pulmonary oedema, arrhythmia, stroke). Risk predication will vary depending on these factors and information is often only available from small case series of specific cardiac conditions in pregnancy. Maternal life expectancy may need to be discussed along with ability to physically care for a child.

High-risk woman should be aware of the likelihood of poor fetal outcome. As before, maternal functional status and cyanosis are major determinants of fetal mortality.

Summary

Women with cardiac disease can and do become pregnant. Most will have a successful outcome, however there are women with both acquired and congenital heart disease, where pregnancy confers significant risk. Pulmonary hypertension, cyanosis, cardiomyopathy, mechanical heart valves, obstructive valvular lesions, aortopathy and ischaemic heart disease are associated with poor maternal and fetal outcome. It is important that clinicians working in obstetrics can recognise and manage these women to ensure safe outcome for mother and baby.

KEY POINTS

- Cardiac disease is a leading cause of maternal morbidity and mortality in pregnancy.
- Pregnancy is associated with major cardiovascular changes.
- All women with cardiac disease should receive pre-pregnancy counselling.
- Congenital heart disease has increased in prevalence worldwide.
- Rheumatic heart disease is still common in indigenous and Pacific populations.
- Mechanical heart valves are problematic in pregnancy due to anticoagulation issues.
- The highest risk cardiac conditions are pulmonary hypertension, cardiomyopathy, stenotic valve lesions, aortic pathology, ischaemic heart disease and cyanotic congenital heart disease.
- Neonatal outcomes correlate with maternal cardiac status and maternal outcomes.

References

1. Ginsberg JS *et al.* Anticoagulation of pregnant women with mechanical heart valves. *Arch Intern Med* 2003;163:694-698.
2. Head CEG; Thorne SA. Congenital heart disease in pregnancy. *Postgrad Med J.* 2005; vol 81(955):292-298.
3. Lupton M *et al.* Cardiac disease in pregnancy. *Curr Opin Obstet Gynecol.* 2002; vol 14(2):137-143.
4. Nelson-Piercy C. Heart disease in pregnancy. *Acta Anaesth Belg.* 2002; 53:321-326.
5. Siu SC, Colman JM. Heart disease and pregnancy. *Heart* 2001; vol 85(6):710-715.
6. Thorne SA. Pregnancy in heart disease. *Heart* 2004; vol 90(4):450-456.

'Heart transplant and pregnancy'.

List of references continued from page 35.

5. Metcalfe J, McAnulty JH, Ueland K. Cardiovascular Physiology. *Clin Obstet Gynecol* 1981; 24:693-5.
6. Hunt SA. Pregnancy in Heart Transplant Recipients: A Good Idea? *J Heart Lung Transplant* 1991; 10:499-503.
7. Lowenstein BR, Vain NW, Perrone SV. Successful pregnancy and vaginal delivery after heart transplantation. *American Journal Obstetrics and Gynecology* 1988; 158: 589-90.
8. Miniero R, Tradivo I, Centofanti P, Goggi C, Mammana C, Parisi F, Dall'Omo AM. Pregnancy in Heart Transplant Recipients. *The Journal of Heart and Lung Transplantation* 2004; 23:898- 901.
9. McKay DB, Josephson MA. Reproduction and Transplantation: Report on the AST Consensus Conference on Reproductive Issues and Transplantation. *American Journal of Transplantation* 2005, 5:1592-1599.
10. Sibanda N, Briggs JD, Davison JM, Johnson RJ, Rudge CJ. Pregnancy After Organ Transplantation. *Transplantation* 2007; Vol.83 No.10, 27 May 2007.