

South Australian Perinatal Practice Guideline

Cardiac disease in pregnancy

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Note:

This guideline provides advice of a general nature. This statewide guideline has been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach. The guideline is based on a review of published evidence and expert opinion.

Information in this statewide guideline is current at the time of publication.

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Health practitioners in the South Australian public health sector are expected to review specific details of each patient and professionally assess the applicability of the relevant guideline to that clinical situation.

If for good clinical reasons, a decision is made to depart from the guideline, the responsible clinician must document in the patient's medical record, the decision made, by whom, and detailed reasons for the departure from the guideline.

This statewide guideline does not address all the elements of clinical practice and assumes that the individual clinicians are responsible for discussing care with consumers in an environment that is culturally appropriate and which enables respectful confidential discussion. This includes:

- The use of interpreter services where necessary,
- Advising consumers of their choice and ensuring informed consent is obtained,
- Providing care within scope of practice, meeting all legislative requirements and maintaining standards of professional conduct, and
- Documenting all care in accordance with mandatory and local requirements

Explanation of the aboriginal artwork:

The aboriginal artwork used symbolises the connection to country and the circle shape shows the strong relationships amongst families and the aboriginal culture. The horse shoe shape design shown in front of the generic statement symbolises a woman and those enclosing a smaller horse shoe shape depicts a pregnant women. The smaller horse shoe shape in this instance represents the unborn child. The artwork shown before the specific statements within the document symbolises a footprint and demonstrates the need to move forward together in unison.



Australian Aboriginal Culture is the oldest living culture in the world yet Aboriginal people continue to experience the poorest health outcomes when compared to non-Aboriginal Australians. In South Australia, Aboriginal women are 2-5 times more likely to die in childbirth and their babies are 2-3 times more likely to be of low birth weight. The accumulative effects of stress, low socio economic status, exposure to violence, historical trauma, culturally unsafe and discriminatory health services and health systems are all major contributors to the disparities in Aboriginal maternal and birthing outcomes. Despite these unacceptable statistics the birth of an Aboriginal baby is a celebration of life and an important cultural event bringing family together in celebration, obligation and responsibility. The diversity between Aboriginal cultures, language and practices differ greatly and so it is imperative that perinatal services prepare to respectively manage Aboriginal protocol and provide a culturally positive health care experience for Aboriginal people to ensure the best maternal, neonatal and child health outcomes.

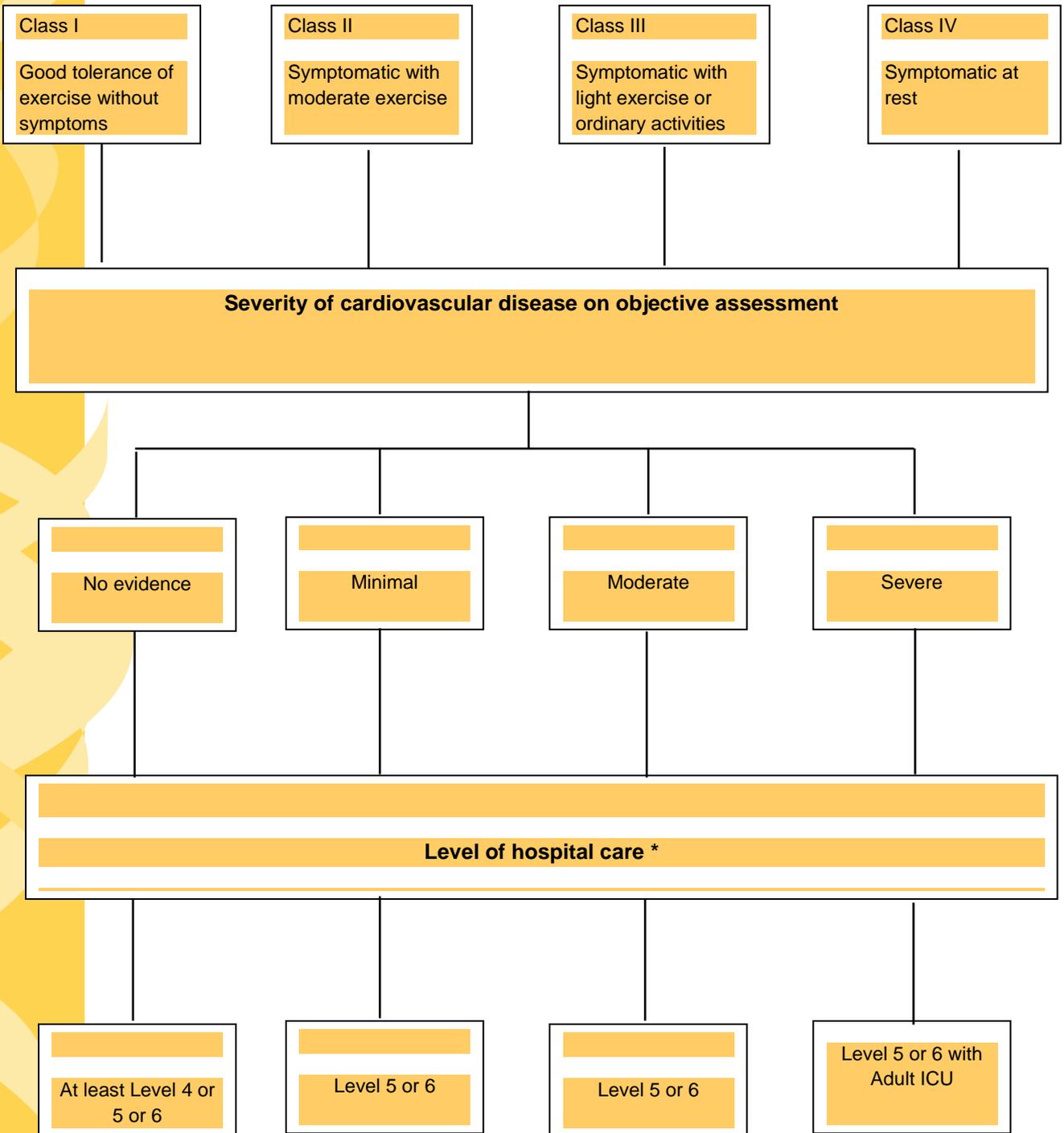
Purpose and Scope of Perinatal Practice Guideline

The purpose of this guideline is to provide clinicians with information on the management of cardiac disease in pregnancy. It includes details on pre-conception counselling and assessment and perinatal care allocation according to severity of cardiac disease. Specific cardiac conditions with associated care are outlined.



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Flowchart I: Level of hospital care according to New York Heart Association (NYHA) functional classification



* For more information on hospital service levels, see *Standards for Maternal and Neonatal Care in SA* available at www.sahealth.sa.gov.au/perinatal

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Table 1: Allocation of care according to severity of cardiac disease

Cardiac Condition	Tertiary centre	Tertiary centre with onsite CCU and ICU
Primary pulmonary hypertension		Yes
Eisenmenger syndrome		Yes
Uncorrected coarctation of the aorta		Yes
AS	Mild to moderate stenosis	Severe with or without symptoms Ao valve area < 1.0 cm ²
AR	NYHA functional class I-II	NYHA functional class III-IV
MS	NYHA functional class I	NYHA functional class II-IV Valve area < 1.5 cm ² PA systolic pressure > 50 mm Hg or > 75 % systemic pressure
MR	Yes NYHA functional class I-II	Yes NYHA functional class III-IV
Aortic and / or mitral valve disease resulting in severe pulmonary hypertension		Yes (pulmonary pressure > 75 % of systemic pressures)
Aortic and / or mitral valve disease with severe LV dysfunction		Yes (EF < 40 %)
Mechanical prosthetic valve requiring anticoagulation		Yes
Marfan syndrome		Yes with aortic complications, with or without AR
History of peripartum cardiomyopathy		Yes with persistent left ventricular dysfunction and NYHA functional class IV
Pacemakers	Yes	
Implanted Cardioverter Defibrillator management in pregnancy		Yes
Management of ventricular arrhythmias		Yes



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Table 2: Cardiovascular Drugs in Pregnancy

Drug	Use in pregnancy	Potential side effects	Breast feeding	Risk factors (* FDA classification)
Adenosine	Maternal and fetal arrhythmias	No side effects reported; data on use during first trimester are limited	Data NA	C
Amiodarone	Maternal arrhythmias	IUGR, prematurity, congenital goiter, hypothyroidism and hyperthyroidism, transient bradycardia, and prolonged QT in the newborn	Not recommended	C
Angiotensin-converting enzyme inhibitors / Angiotensin receptor II antagonists	Hypertension, heart failure	Oligohydramnios, IUGR, prematurity, neonatal hypotension, renal failure, anemia, death, skull ossification defect, limb contractures, patent ductus arteriosus	Compatible	D
Beta blockers	Hypertension, maternal arrhythmias, myocardial ischemia, mitral stenosis, hypertrophic cardiomyopathy, hyperthyroidism, Marfan syndrome	Fetal bradycardia, low placental weight, possible IUGR, hypoglycemia, no information on carvedilol	Compatible, monitoring of infant's heart rate recommended	Acebutolol: B Labetalol: C Metoprolol: C Propranolol: C Esmolol: C Atenolol: D
Digoxin	Maternal and fetal arrhythmias, heart failure	No evidence for unfavorable effects on the fetus	Compatible	C
Diltiazem	Myocardial ischemia, tocolysis	Limited data; increased incidence of major birth defects	Compatible	C
Diuretics	Hypertension, congestive heart failure	Hypovolemia leads to reduced uteroplacental perfusion, fetal hypoglycemia, thrombocytopenia, hyponatremia, hypokalemia; thiazide diuretics can inhibit labour and suppress lactation	Compatible	C



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Flecainide	Maternal and fetal arrhythmias	Limited data; 2 cases of fetal death after successful treatment of fetal SVT reported, but relation to flecainide uncertain	Compatible	C
Heparin	Anticoagulation	None reported	Compatible	C
Hydralazine	Hypertension	None reported	Compatible	C
Lignocaine	Local anesthesia, maternal arrhythmias	No evidence for unfavorable fetal effects; high serum levels may cause central nervous depression at birth	Compatible	C
Nifedipine	Hypertension, tocolysis	Fetal distress related to maternal hypotension reported	Compatible	C
Nitrates	Myocardial infarction and ischemia, hypertension, pulmonary edema, tocolysis	Limited data; use is generally safe, few cases of fetal heart rate deceleration and bradycardia have been reported	Data NA	C
Procainamide	Maternal and fetal arrhythmias	Limited data; no fetal side effects reported	Compatible	C
Sodium nitroprusside	Hypertension, aortic dissection	Limited data; potential thiocyanate fetal toxicity, fetal mortality reported in animals	Data NA	C
Sotalol	Maternal arrhythmias, hypertension, fetal tachycardia	Limited data; 2 cases of fetal death and 2 cases of significant neurological morbidity in newborns reported, as well as bradycardia in newborns	Compatible, monitoring of infant's heart rate recommended	B
Verapamil	Maternal and fetal arrhythmias, hypertension, tocolysis	Limited data; other than a single case of fetal death of uncertain cause, no adverse fetal or newborn effects have been reported	Compatible	C
Warfarin	Anticoagulation	Crosses placental barrier; fetal hemorrhage in utero, embryopathy, central nervous system abnormalities	Compatible	X

* See appendix for [FDA Classification](#) information



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Summary of Practice Recommendations

Antenatal Care

Low risk

- > Tertiary hospital (Level 5, 6)
- > Consultation with anaesthetists, cardiologist / obstetric physician in high risk clinic

Intermediate or high risk

- > High risk obstetric centre with multidisciplinary care from obstetrician, anaesthetist, intensivist, neonatologist, midwifery and nursing

Antepartum Management

- > Multidisciplinary approach
- > Serial cardiac assessments to identify and act upon cardiac deterioration
- > Fetal echo for mothers with congenital heart disease +/- genetic counselling
- > Optimise medication for pregnancy
- > Early admission for high risk women

Peripartum Management

Low and intermediate risk

- > Epidural anaesthesia
- > Cardiac telemetry and oximetry if required
- > Deep venous thrombosis (DVT) prophylaxis

High Risk

- > Multidisciplinary approach including decision on delivery in an intensive care unit (ICU) or not and level of invasive monitoring
- > Planned birth at term
- > Aim for vaginal birth except in specific instances (e.g. Marfan's, critical aortic stenosis)
- > Care in labour and delivery unit, coronary or intensive care unit if indicated. Continued careful observation using special observation charts and possibly including systems such as 'Early warning scores' for signs of clinical deterioration in the 4th stage



Cardiac disease in pregnancy

Abbreviations

ACC	American College of Cardiology
ACE	Angiotensin converting enzyme
AFI	Amniotic fluid index
AHA	American Heart Association
APTT	Activated Partial Thromboplastin Time
AR	Atrial regurgitation
AS	Atrial stenosis
AV	Atrio ventricular
BPM	Beats per minute
BSA	Body surface area
CEMACH	Confidential Enquiry into Maternal and Child Health (UK)
CCU	Coronary care unit
DC	Direct current
DVT	Deep venous thrombosis
ECG	Electrocardiogram
EDC	Estimated date of confinement
EF	Ejection fraction
e.g.	For example
g	Gram(s)
ICD	Internal cardiac defibrillator
ICU	Intensive care unit
INR	International normalised ratio
IUCD	Intrauterine contraceptive device
IUGR	Intrauterine growth restriction
IU	International unit(s)
LMWH	Low molecular weight heparin
LV	Left ventricular
mg	Milligram(s)
MI	Myocardial infarction
mL	Millilitre(s)
MS	Mitral stenosis
MR	Mitral regurgitation
NA	Not applicable
NYHA	New York Heart Association
PA	Pulmonary artery
RF	Radio frequency
RPM	Respirations per minute
SVT	Supraventricular tachycardia
TCPC	Total cavopulmonary connection
TOP	Termination of pregnancy
UFH	Unfractionated heparin



Cardiac disease in pregnancy

Introduction

- > Between 1 % and 3 % of women will have a form of cardiac disease diagnosed before or during pregnancy
- > Cardiac pathology may be:
 - > Congenital or acquired
 - > Functional or structural
 - > Cyanotic or non-cyanotic
 - > Or may include endocardial, myocardial or pericardial defects²
- > Specific hazards for women with cardiac disease in pregnancy include:
 - > The occurrence of pulmonary oedema (main cause of death)
 - > Pulmonary hypertension
 - > Infective endocarditis
 - > Thromboembolism
 - > Fulminating peripartum cardiomyopathy
 - > Aortic dissection
 - > Primary arrhythmogenic disorders
- > The most frequent causes of cardiac disease in pregnancy are:
 - > Rheumatic heart disease
 - > Congenital heart disease
 - > Arrhythmias
 - > Ischemic heart disease
 - > Cardiomyopathy
- > Fetal risks
 - > Congenital heart disease
 - > Intrauterine growth restriction
 - > Prematurity

New York Heart Association (NYHA) functional classification

- > Classification of the severity of heart disease can aid in the prediction of maternal and neonatal outcomes and in the counselling of prospective parents³

Class	Clinical Implications	Objective Assessment
I	Good tolerance of exercise without symptoms (chest pain, angina, dyspnoea, palpitations, fatigue)	No objective evidence of cardiovascular disease
II	Symptomatic with moderate exercise	Objective evidence of minimal cardiovascular disease
III	Symptomatic with light exercise or ordinary activities	Objective evidence of moderately severe cardiovascular disease
IV	Symptomatic at rest	Objective evidence of severe cardiovascular disease

Adapted from American Heart Association. In: Dobbenga-Rhodes YA, Privé AM. Assessment and evaluation of the woman with cardiac disease during pregnancy. J Perinat Neonat Nurs 2006; 20: 295-302.

Preconception care

- > Assessment of the woman's general physical condition, medications and cardiac function
- > An exercise test (with VO₂ max measurements) and echocardiogram provide essential information on pre-pregnancy cardiac status and reserve⁴
- > Optimise management of hypertension / arrhythmias
- > Referral to cardiologist to discuss current medications (e.g. Angiotensin-converting enzyme [ACE] inhibitors) and associated risks to the fetus in pregnancy and plan alternative treatments where the woman's condition permits. Information is also available from Medicines and Drug information service, telephone 81617222
- > Assess lifestyle risk factors e.g. smoking and education to optimise exercise tolerance
- > Ideally, discussions should occur between the woman, her family and a cardiologist, physician, obstetrician and / or anaesthetist familiar with the management of cardiac diseases in pregnancy to agree and decide on the best timing for pregnancy
- > Points to be discussed include:
 - > The natural history of the woman's disease, including the possibility of a successful pregnancy through medical treatment and optimising the woman's cardiac function
 - > Present clinical staging of cardiac disease as per New York Heart Association functional classification
 - > Optimising the woman's clinical condition by changes in medications or surgical procedures before conception
 - > Expected outcomes including chance of spontaneous miscarriage, live birth, death, proposed care plan of the pregnancy and implications of admissions, antenatal care, anaesthesia, medications and peripartum management
 - > The risk of transmitting the cardiac condition or syndrome to offspring (e.g. congenital heart disease, Marfan syndrome, hypertrophic cardiomyopathy)^{2,5}

Antenatal management

- > Routine booking visit around 10 to 12 weeks
- > Early ultrasound to establish estimated date of confinement (EDC)
- > These women should be managed by a multi-disciplinary team, which might include an obstetrician, cardiologist or obstetric physician, anaesthetist and intensivist. These clinicians should all be familiar with the management of cardiac problems in pregnancy
- > The woman should be involved in a discussion about the management of the pregnancy:
 - > Decision about the most appropriate facility to deliver the woman and criteria for changing the place of birth
 - > Antenatal visits in high risk medical clinic every 2 weeks if required because of severity of disease
 - > Regular review of functional cardiac status at each visit
 - > Echo cardiograms with timely follow up by Obstetrician and Cardiologist
 - > Avoid anaemia – iron supplementation, prenatal vitamins and dietary counselling and regular haemoglobin checks
 - > Ongoing discussion regarding work schedules and levels of activity
 - > Arrange anaesthetic review before 28 weeks



Clinical assessment

History

- > Maternal age, gestational age, parity status, NYHA functional classification, comorbid conditions, previous cardiac events / surgery / interventions, cardiac lesions, cyanosis (oxygen saturation < 90 %), medications, use of cigarettes and / or alcohol

Investigations

- > 12-lead electrocardiogram (ECG)
- > Echocardiographic assessment to determine:
 - > Systemic (left) and venous (right) ventricular systolic function,
 - > Doppler quantification of inflow or outflow obstruction
 - > Doppler quantification of valvular function
 - > Right heart pressures and systolic if measurable⁶

Fetal wellbeing

- > Women with cyanotic heart disease, NYHA III or IV, left heart obstruction, smokers, multiple gestation, beta-blocker use and anticoagulation are at high risk of having a fetus with growth restriction. These women should be offered regular ultrasound assessments of fetal wellbeing (growth, umbilical artery Doppler, AFI) and cardiotocography in the third trimester

Fetal risks

- > Fetal cardiac assessment is necessary because there is a 2 to 16 % risk of congenital heart disease in the fetus in pregnant woman with structural congenital heart disease⁷. The incidence of congenital heart disease in the offspring is more common in the fetus when the mother, rather than the father, is affected, particularly if the mother has a condition such as bicuspid aortic valve, which is more common in the male
- > Autosomal dominant conditions such as Marfan's syndrome, hypertrophic cardiomyopathy, mitral valve prolapse and channelopathies have a 50 % chance of being passed on, but may not be associated with any structural abnormalities in the fetus
- > Early diagnosis of congenital heart defect in the fetus (before 24 weeks of gestation) allows the possibility of termination of the pregnancy (TOP)
- > Affected fetuses benefit from delivery in a tertiary care centre (Level 5, 6) with Paediatric Cardiac Support
- > The two main determinants of fetal prognosis are maternal functional class and the degree of maternal cyanosis. When the mother is in functional class III to IV, or has a high risk disease such as severe aortic stenosis or Eisenmenger syndrome or is cyanosed, monitoring of fetal growth is very important⁷
- > In Australia and New Zealand, neonatal survival ranges from 69 % at 24 weeks gestation to 93 % at 27 weeks gestation. The risk of cerebral palsy in surviving neonates at < 28 weeks gestation is 9.1 %, varying from 7% to 15 % (27 to 24 weeks gestation). Of the overall 9.1 %, 27 % have grade 1 cerebral palsy (on a grading scale of 1 to 5)⁸
- > Therefore, when cardiac function is reduced to critical levels, surgery or percutaneous procedures to improve maternal cardiac function should be undertaken, if feasible, in order to postpone delivery as long as possible⁷
- > The choice between corrective procedures and delivery may be difficult between 28 and 32 weeks, and decisions must be individualised
- > Administer IM betamethasone in two doses of 11.4 mg (5.7 mg x 2) 24 hours apart to the woman if birth is likely to occur between 23⁺⁰ and 35⁺⁰ weeks



Level of hospital care (see [Flowchart 1](#) and [Table 1](#))

- > Echocardiographic assessment results and the on-going NYHA functional classification will determine if the level of hospital care required is:
 - > Hospital of choice care (must be at least Level 4)
 - > Tertiary centre care (Level 5 or 6)
 - > Tertiary centre with onsite specialist adult cardiac service care and adult intensive care (available at some Level 5 or 6 hospitals)
- > It may be possible for some women to have some antenatal visits with a geographically closer unit ("shared care") if delivery at a higher level unit is required. This should be negotiated on a case by case basis
- > Women with NYHA functional class I may be suitable for care at a Level 4 hospital with an onsite specialist cardiac and / or adult intensive care service/s; however, they will usually require care in a tertiary centre (Level 5 or 6)
- > Women with NYHA functional class II, III, IV may be referred to a hospital with onsite specialist adult cardiac service care and adult intensive care (Level 5 or 6), particularly if there is a history of:
 - > Severe cardiac condition (see specific cardiac conditions)
 - > Valve replacement or other cardiac surgery
 - > Any particular risk of endocarditis
- > Referral to a hospital with specialist adult cardiac service (Level 5 or 6) may be required if condition worsens

Cardiac condition stable:

- > Aim for vaginal birth at term (see individual cardiac diagnoses for specific contra indications for vaginal delivery)
- > Consider caesarean section only for obstetric indications
- > Inform the woman that decisions about her management may change at short notice
- > Maintain on-going communication within the high risk medical team
- > Further investigations may be indicated, e.g. echo, chest X ray etc

SPECIFIC CONDITIONS

Benign arrhythmias

- > Includes **sinus tachycardia, premature atrial and ventricular contractions, paroxysmal supraventricular tachycardia (SVT), atrial fibrillation, atrial flutter**
 - > The majority of women with arrhythmias during pregnancy have a benign increased rate of atrial or ventricular premature beats⁹. Both ectopic beats and sustained arrhythmias become more frequent during pregnancy. They may even develop for the first time. In general they are treated in the same way as non-pregnant patients, but as conservatively as possible. Definitive treatment is reserved for the postpartum period if it is safe to do so. Women who are worried about ectopic beats can usually be reassured unless the frequency increases on exercise
 - > Haemodynamically stable women do not usually require treatment
 - > Advise woman to avoid precipitating factors such as caffeine, alcohol, tobacco, fatigue and anxiety



Other arrhythmias

- > Women with more ominous atrial arrhythmias (**supraventricular tachycardia, atrial fibrillation and flutter**) should be managed in collaboration with a cardiologist, usually using the same agents that would be chosen in the non-pregnant woman, including electrical cardioversion when necessary⁹
- > All commonly used anti-arrhythmic drugs cross the placenta. The pharmacokinetics of drugs are altered in pregnancy and require close clinical monitoring to ensure maximum efficacy and avoid toxicity
- > Supraventricular tachycardias are corrected by vagal stimulation or, failing that, intravenous adenosine. If adenosine is unsuccessful at restoring sinus rhythm consider using β -blockers or verapamil. Electrical cardioversion is not contraindicated and should be used for any sustained tachycardia causing haemodynamic instability and therefore threatening fetal wellbeing. Beta blocking drugs with beta-1 selectivity are the first choice for prophylaxis. Verapamil is constipating but may be used if there is a contraindication to β -blocker use (such as asthma)
- > Both β -blockers and the dihydropyridone calcium antagonists can cause fetal bradycardia. If the arrhythmia is uncontrolled and severely compromising the health of the mother and fetus, radio frequency ablation for atrio ventricular (AV) nodal re-entry or certain atrio ventricular re-entry tachycardias can, if necessary, be performed during pregnancy with suitable lead shielding and maximal use of echo rather than X-ray fluoroscopy
- > Class I agents such as flecainide may be considered if the maternal left ventricle is normal. Information on its use in pregnancy is limited. Of the Class III agents, sotalol is much more preferable to amiodarone and has similar risks to β -blockers with the increased risk of torsades de pointes due to QT prolongation. Amiodarone should be avoided unless the maternal arrhythmia is life-threatening to the mother or fetus, other agents have failed and the arrhythmia is not manageable with radiofrequency (RF) ablation. Long term use of amiodarone can cause neonatal hypothyroidism (9 % of newborns), hyperthyroidism, and goitre
- > Potentially life-threatening **ventricular tachyarrhythmias** are much less common and should be terminated by electrical cardioversion if causing maternal haemodynamic compromise. Management requires the opinion of a cardiologist and should be managed as in the non-pregnant woman. This includes the use of antiarrhythmic medication in the short- to long-term and / or the likely insertion of an automatic internal cardiac defibrillator (ICD)

Pacemakers and automatic internal cardiac defibrillators (ICD)

- > The presence of a pacemaker or an ICD does not itself contraindicate pregnancy. The presence of any underlying structural heart disease determines the risk of pregnancy
- > Pregnancy in patients with an ICD does not cause increased ICD-related complications or adverse events in the mother or fetus; neither does it increase the number of ICD discharges
- > A pacemaker for the alleviation of symptomatic bradycardia, or ICD can be implanted at any stage of pregnancy using X-ray fluoroscopy with suitable lead shielding and echo guidance, to minimise the radiation dose delivered to the mother and fetus
- > Any woman with an ICD should be advised to carry identification information about the ICD type, date implanted, and location of implantation in her South Australian pregnancy record. A plan for deactivation in case of an emergency should be documented in her South Australian pregnancy record.

- > The presence of a pacemaker or ICD should not alter the mode of delivery. This is determined by obstetric indications
- > If Caesarean section is required, the ICD should be temporarily inactivated (preferably by reprogramming or in an emergency by application of a magnet over the generator) until the procedure is finished. While inactivated, the patient should have continuous ECG monitoring and external defibrillation available at all times

Rheumatic heart disease and mitral stenosis

- > The incidence of rheumatic heart disease is decreasing in developed countries. Mitral stenosis is the most common sequelae
- > RDH Registry in SA: email rhd@sa.gov.au; Ph (08) 7425 7146
- > In women with rheumatic heart disease, the mitral valve is most commonly affected. Mitral stenosis occurs in 90 % of cases, mitral regurgitation in 7 %, aortic regurgitation 2.5 % and aortic stenosis in 1 % of cases of rheumatic heart disease¹⁰

Mitral stenosis

- > Women with mild (NYHA class I) mitral stenosis may be asymptomatic until pregnancy – the increase in left atrial pressure may lead to subsequent symptoms of dyspnoea, and eventually tachypnoea, orthopnoea, and paroxysmal nocturnal dyspnoea
- > Manage according to symptoms and with the guidance of echocardiography, which is the key to the assessment of valvular severity. Mitral stenosis is considered severe and associated with significant increased maternal and fetal risk when the mitral valve area is estimated to be < 1.5 cm², the functional capacity is NYHA III or IV or the right heart pressures rise significantly (estimated PA systolic pressure > 50 mmHg or > 75 % of the systemic pressure)

Medical treatment

- > Limitation of exercise, fluid and salt restriction should be used when the functional class is NYHA II or more
- > In cases of poor functional tolerance, consider oral β -adrenergic blockers (decreases maternal tachycardia), and judicious use of diuretics, avoiding vigorous volume depletion to protect against uteroplacental hypoperfusion (caution in women with severe mitral stenosis – may need concurrent use of invasive haemodynamic monitoring)¹⁰
- > Aggressively cardiovert new onset atrial fibrillation back to sinus rhythm with β -blockers, verapamil and / or direct current (DC) cardioversion (see above)
- > Consider digoxin to control the ventricular response to chronic atrial fibrillation.
- > See anticoagulation prophylaxis for treatment recommendations

Intervention

- > Women who fail medical treatment during pregnancy with repeated episodes or persistent heart failure should be considered for percutaneous mitral valvotomy if suitable anatomy is present. After successful mitral valvotomy, pregnancy can often continue successfully to term
- > Surgical replacement of the mitral valve is used as a last resort with high fetal risk of peri-operative death. If possible, this should be delayed until the fetus is viable and can be delivered before the surgical intervention



Intrapartum

- > The mode of delivery is determined by obstetric indications only
- > Care in tertiary centre for NYHA functional class I, II. These patients can be managed in the delivery suite without invasive monitoring. ECG monitoring to monitor maternal heart rate would be preferable
- > NYHA functional class III, IV: Refer to hospital with specialist adult cardiac service and invasive monitoring in labour. These patients should be managed in an intensive care setting with invasive monitoring, avoiding haemodynamic stress and having careful control of heart rate. Tachycardia can be prevented with intravenous esmolol as necessary
- > Excellent pain control is important and choices should be planned in advance in consultation with the anaesthetist
- > Close monitoring for the first 48 hours after birth

Mitral regurgitation

- > Mitral valve prolapse is the most common cause of mitral regurgitation in women who are pregnant
- > May also be associated with mitral stenosis if the aetiology is of rheumatic fever
- > Pregnancy is generally well tolerated (It is theorised that mitral regurgitation may improve in pregnancy due to the physiologic reduction in systemic vascular resistance)

Medications

- > Pharmacological treatment is not often needed. Diuretics and digoxin can be used for pulmonary congestion. Vasodilator therapy with hydralazine should be used with caution only in those with associated hypertension. ACE inhibitors are considered unsafe for the fetus and are therefore contraindicated
- > Previously asymptomatic women may worsen immediately after delivery because of sudden increases in systemic vascular resistance
- > Increased risk of left atrial enlargement and subsequent atrial fibrillation

Aortic stenosis

- > The most common cause is congenital aortic valve disease (e.g. Bicuspid aortic valve). There is an association with bicuspid aortic valve and aortic root dilatation and co-arctation
- > May also be associated with mitral stenosis if the aetiology is rheumatic fever
- > Maternal mortality associated with severe aortic stenosis is 17 % with a fetal mortality of 32 %¹⁰
- > Echocardiography is essential in determining the severity of the stenosis. In pregnancy an aortic valve area < 1.0 cm² (0.6 cm² / m² BSA) is associated with a significant increased maternal and fetal risk
- > Decisions regarding anaesthesia and mode of delivery must be individualised on the basis of obstetric indications, severity of maternal symptoms and aortic valve area
- > In severe or symptomatic aortic stenosis, the intrapartum management should be carried out in a hospital with adult cardiac services and an adult intensive care. Invasive monitoring, especially intra-arterial pressure monitoring is of utmost importance. Avoidance of hypotension and hypovolaemia is essential to avoid haemodynamic collapse
- > Balloon aortic valvuloplasty (BAV) is a good option for severe aortic stenosis (usually preferred to be performed in 2nd trimester) to provide assistance in managing symptoms in pregnancy, defer definitive valve surgery and is low risk to mother and fetus

Cardiac disease in pregnancy

- > Despite aggressive medical therapy, some women will require aortic valve replacement in pregnancy. Case reports suggest that pulsatile perfusion at bypass may help preserve placental haemodynamic function¹⁰

Aortic regurgitation

- > May also be associated with mitral stenosis
- > Clinical course is determined more by the extent of their mitral valve disease than by their aortic regurgitation
- > When aortic regurgitation is the predominant lesion, pregnancy is usually well tolerated and may improve in pregnancy due to the decrease in systemic vascular resistance
- > Cardiac surgery for aortic regurgitation (valve replacement) is high risk to mother and fetus and can usually be deferred until after delivery (aortic regurgitation is well tolerated in pregnancy)
- > Also, the physiologic tachycardia of pregnancy may reduce regurgitant flow as diastolic filling times are shortened
- > Murmurs normally associated with both aortic and mitral regurgitation may be reduced in pregnancy
- > Women with severe aortic regurgitation and symptoms of left-sided heart failure should decrease their physical activity
- > Reduce sodium intake < 2 g per day
- > Consider diuresis or vasodilators (e.g. hydralazine or nifedipine) and inotropic therapy in difficult cases

Congenital heart disease

- > May be organised into acyanotic and cyanotic types
- > Acyanotic congenital heart lesions include:
 - > Atrial / ventricular septal defects,
 - > Patent ductus arteriosus
 - > Aortic coarctation
 - > Marfan syndrome
 - > Aortic / pulmonary stenosis
- > Cyanotic congenital heart lesions include:
 - > Cyanotic congenital heart disease,
 - > Tetralogy of Fallot,
 - > Post Fontan operation
- > Usually require referral to a tertiary centre with specialist adult cardiac service care
- > Fetal echocardiogram at 20 weeks of gestation
 - > If on warfarin, look for echogenic foci in the heels and coccyx, profile to assess nasal bridge
 - > Identified cardiac fetal anomaly carries a 4-5 % risk of associated chromosomal abnormality – recommend amniocentesis with antibiotic prophylaxis as above
- > Most women present as NYHA class I or II lesions and remain largely asymptomatic



Cardiac disease in pregnancy

- > The risk of a cardiac event in pregnancy is increased for women with:
 - > A prior cardiac event or arrhythmia
 - > NYHA functional class class III-IV
 - > Cyanosis
 - > Left heart obstruction
 - > Systemic ventricular dysfunction

Coarctation of the aorta

- > Coarctation of the aorta should be repaired before pregnancy. It is rare during pregnancy (9 % of all congenital defects). The management of hypertension is difficult in the unoperated pregnant woman. Fetal growth is usually normal and in contrast to essential hypertension, there is not an increased incidence of preeclampsia. Over enthusiastic treatment may cause hypotension in the distal segment. This may result in abortion or fetal death even though pressure in the proximal segment continues to rise on effort
- > Rupture of the aorta is the most common reported cause of death, and rupture of an aneurysm of the circle of Willis has also been reported during pregnancy. The increase in blood volume and cardiac output increases the risk of aortic dissection or rupture during pregnancy and a beta blocker should be prescribed
- > Restriction of physical activity is the only way of minimising potentially dangerous surges in blood pressure
- > Surgical correction is only very rarely indicated during pregnancy if systolic hypertension is uncontrolled or heart failure is present
- > Balloon angioplasty is contraindicated because of the risk of dissection or rupture. Whether this risk is avoidable with stenting is not known

Marfan syndrome

- > Rare - incidence is 1 in 5,000 ^{7,11}
- > Women with a documented aortic root diameter of < 40 mm without an abnormal aortic valve have a mortality rate of < 5 %
- > Risk of aortic dissection during pregnancy or shortly thereafter is approximately 10 % with the incidence appearing to increase with increasing aortic root diameter⁷
- > Aortic or mitral regurgitation is also seen in 60 % of women with Marfan syndrome and may complicate pregnancy¹¹
- > Regular echocardiography before, during and after pregnancy to monitor aortic root size and valvular function
- > Genetic testing through chorionic villus biopsy, amniocentesis cell culture or postnatal testing
- > Angiotensin receptor blockers are recommended before pregnancy to decrease increasing aortic root dilatation but must be stopped during pregnancy
- > B-blockers are strongly recommended before and during pregnancy to decrease the risk of aortic dilatation and dissection
- > Pregnancies complicated by Marfan syndrome are not associated with poor perinatal outcomes, though some suggest there is an increased risk of incompetent cervix
- > Physical, echocardiographic, and ophthalmologic examination of newborns
- > Counsel woman regarding the risk of autosomal dominant inheritance and need for follow-up for their offspring¹¹

Intraatrial repair for transposition of great arteries (TGA)

- > Over 100 pregnancies have been reported in the literature with no deaths. In women in NYHA functional class I-II pregnancy is usually well tolerated. Worsening of systemic ventricular function during or shortly after pregnancy occurred in 10 % of the reported cases⁷
- > Angiotensin-converting enzyme (ACE) inhibitors should be stopped as soon as pregnancy is confirmed⁷

Congenitally corrected transposition of the great arteries

- > Women without significant other cardiac defects usually do well, but problems can develop through failure of the systemic right ventricle with increasing regurgitation through its tricuspid atrioventricular valve. Supraventricular arrhythmias, embolism, and atrioventricular block are other potential complications⁷

Fontan procedure

- > Careful patient selection is important. The successful Fontan Procedure with a small right atrium or total cavopulmonary connection (TCPC) in functional class I or II can probably complete pregnancy with a normal live birth. Fontan patients with a large right atrium and some venous congestion have to be monitored very carefully. They need anticoagulant treatment and conversion to total cavopulmonary connection before pregnancy is considered

Aortic dissection

- > The Stanford system classifies Acute Aortic Dissection into type A (proximal to left subclavian artery origin) or type B (distal to left subclavian artery origin)¹²
 - > 80% of aortic dissections occurring in pregnancy are type A dissections and 87.5% of these occur in the antepartum period (usually in the third trimester)¹³
- > Uncontrolled hypertension is the most significant treatable risk factor for acute aortic dissection¹²

Diagnosis

- > Diagnosis and management during pregnancy are similar to that for aortic dissection in the non-pregnant population
- > Prompt and urgent diagnosis is the first objective
- > Transfer to a Level 6 unit capable of diagnosing and dealing with the complications associated with aortic dissection, namely diagnostic and therapeutic cardiology, cardio thoracic surgery, adult and neonatal ICU, and specialist obstetrics
- > History: consider the presence of underlying risk factors such as Marfan's Syndrome, bicuspid aortic valve (BAV) aortopathy or severe hypertension. Other causes include Turner syndrome, Ehlers-Danlos syndrome and familial-dissection disorders¹³



Signs and symptoms

Presenting symptoms vary and may include

- > Epigastric pain
 - > Unilateral blindness
 - > Central chest or interscapular pain (acute)
 - > A wide pulse pressure, mainly secondary to systolic hypertension
 - > A new cardiac murmur¹³
- > Thoracic aortic dissection is usually suspected clinically from the history and physical examination. The initial tear typically presents with severe, sharp or "tearing" anterior chest pain (in ascending aortic dissection) or posterior chest or back pain (in dissection distal to the left subclavian). If the dissection leads to impaired or absent blood flow to peripheral vessels, the physical examination may reveal a unilateral or bilateral pulse deficit, defined as a weak or absent carotid, brachial, or femoral pulse

Investigations

- > Trans oesophageal echocardiogram
- > Magnetic resonance imaging (MRI) aortic imaging if immediately available. MRI without gadolinium contrast is preferred (may be appropriate if it enhances the clarity of the diagnosis). Limited data suggest that the fetal risk of gadolinium may be minimal
- > Theoretically, Contrast Computed Tomography scan (CT) should be avoided due to concerns about ionizing radiation during pregnancy. However the availability, reliability and speed of CT often make this the test of choice in pregnant patients presenting with a high clinical suspicion for aortic dissection

Treatment¹⁴

- > The 2010 ACC/AHA/AATS guidelines include the following recommendations:
 - > For Type A (ascending aorta) dissection during the first or second trimester, urgent surgical repair with aggressive fetal monitoring when feasible is preferred. Fetal loss is common during hypothermia and prolonged cardiopulmonary bypass and there is an increased rate of teratogenesis
 - > For Type A dissection during the third trimester, urgent caesarean delivery followed by aortic repair is suggested
 - > For acute arch or Type B aortic dissection, medical therapy is preferred unless intervention (percutaneous stent or surgery) is required to treat malperfusion, aortic rupture, or subacute aortic leaking
 - > Pharmacologic interventions to reduce shear stress and blood pressure are needed but the administration of nitroprusside during pregnancy should be avoided when feasible in order to avoid possible fetal thiocyanate toxicity



Prosthetic heart valves

- > The majority of women with bioprosthetic valves do not require anticoagulation during pregnancy⁹
- > Women with mechanical valves require anticoagulation prophylaxis and should receive a detailed discussion of the advantages and disadvantages of the three anticoagulant options (warfarin, unfractionated heparin and low molecular weight heparin) (see anticoagulation prophylaxis below)
- > Despite valve replacement there remains a mild functional stenosis across these valves due to the prosthesis itself
- > Women with valve replacements are at high risk of bacterial endocarditis
- > Perform blood cultures if any signs of bacterial endocarditis
- > Observe for signs of cardiac failure e.g. dyspnoea (respirations > 24 per minute), tachycardia (> 130 beats per minute), fatigue, oedema, hypoxia / cyanosis

Warfarin

- > Warfarin crosses the placenta, is teratogenic and may cause haemorrhage in the fetus
- > The teratogenic effects appear to result from inhibition of vitamin K and / or arylsulphatase E activity during skeletal development
- > The fetal warfarin syndrome comprises nasal hypoplasia, short fingers with hypoplastic nails, low birth weight, stippling of epiphyses on X-ray and intellectual disability
- > Recent estimates indicate the risk of fetal warfarin syndrome in babies of women who require warfarin throughout pregnancy is around 5 % with the risk increasing with higher doses of warfarin
- > The period of greatest embryonic susceptibility is between 6 and 9 weeks of gestation. It is recommended that, wherever possible, the drug is avoided throughout the first trimester¹⁴

Pulmonary hypertension

- > Rare in pregnancy – women with pulmonary hypertension have a high morbidity and mortality during pregnancy e.g. right heart failure, pulmonary thromboembolism, sudden death¹⁶
- > Existing pulmonary vascular disease prevents the fall in pulmonary vascular resistance, leading to a rise in pulmonary artery pressure with increased cardiac output. Ultimately, the increase in cardiac output cannot be achieved resulting in right heart failure¹⁶
- > When pregnancy occurs and termination is declined, or pulmonary hypertension is newly diagnosed during pregnancy, management must be by the multidisciplinary team, including a pulmonary hypertension specialist, in a Level 6 tertiary centre with Adult ICU¹⁶
- > Women with Eisenmenger syndrome are at particular risk of increased right-to-left shunting. Hypoxia can lead to syncope and sudden death



Ischaemic heart disease

- > Includes acute myocardial infarction (MI), unstable angina, stable angina
- > Requires care in a tertiary centre or care in a tertiary centre with specialist adult cardiac service care depending on echocardiograph findings and symptoms according to NYHA functional classification
- > Rare in pregnancy - ischaemic heart disease occurs on average in 1 in 10,000 deliveries
- > Myocardial infarction (MI) in pregnancy occurs on average in 7.5 in 100,000 deliveries with mortality highest in the third trimester in women under 35 years of age¹⁷
- > CEMCH 2003-2005 noted an increase in maternal deaths from ischaemic heart disease. The following risk factors were identified:
 - > Obesity
 - > Older age
 - > Higher parity
 - > Smoking
 - > Diabetes
 - > Pre existing hypertension
 - > Family history¹⁸

Myocardial infarction management

- > Similar to management of the non-pregnant woman with MI and includes:
 - > Oxygen as required to avoid hypoxia, nitrates, morphine, and most antiarrhythmics, calcium channel blockers, β -blockers, heparin, low-dose aspirin, and invasive hemodynamic monitoring if necessary
 - > Case reports using percutaneous interventions and systemic and local thrombolytic therapy have also been successful¹⁷
- > MI is not an indication for immediate delivery (mortality is higher in women who deliver within 2 weeks of MI)
- > Caesarean section only for obstetrical indications (does not protect women from the dramatic changes in stroke volume and cardiac output associated with the immediate postpartum period)
- > Electrocardiographic monitoring in labour

Heart failure and cardiomyopathy

- > Includes left sided, right heart failure and biventricular heart failure
- > Common causes for heart failure in the puerperium include iatrogenic fluid therapy overload, pre-eclampsia, thyrotoxicosis, peri-partum cardiomyopathy, anaemia or previously undiagnosed rheumatic or congenital heart disease¹⁷

Hypertrophic cardiomyopathy

Clinical assessment

- > Echocardiography
- > electrocardiography (ECG), including ambulatory ECG
- > Exercise testing and stress echocardiography



Treatment

- > Genetic counselling
- > β -blockers
- > Diuretics
- > Manage atrial fibrillation with a preference for rhythm control and maintenance of sinus rhythm and anticoagulation
- > Implantation of an internal cardiac defibrillator for documented sustained ventricular arrhythmias and amiodarone if frequent shocks occur due to frequent sustained VT
- > Normal vaginal birth

Peripartum dilated cardiomyopathy

- > Several risk factors but no specific cause
- > Excludes women with unrecognised pre-existing dilated cardiomyopathy and women with acute viral myocarditis during pregnancy
- > Women with past peri-partum dilated cardiomyopathy should receive counselling as to the risk of recurrence in subsequent pregnancies. The risk varies according to the completeness and speed of recovery of left ventricular function after delivery
- > Counsel women to avoid pregnancy / consider termination of pregnancy

Clinical assessment

- > Echocardiography

Diagnosis criteria

- > Development of cardiac failure in the last month of pregnancy or within five months postpartum
- > Global left ventricular systolic dysfunction on echocardiography
- > Lack of another cause for the woman's cardiac failure
- > Lack of cardiac disease preceding the month before delivery

Risk factors

- > Older, multiparous women and women with preeclampsia or twins

Symptoms

- > Shortness of breath, lung field crackles, oedema within the first 5 months postpartum

Investigations

- > Complete blood count, electrolytes and renal function, electrocardiogram, chest X-ray, arterial blood gases, and an echocardiogram

Treatment

- > Treat as for other causes of decompensated heart failure with LV systolic dysfunction
- > Reduce preload and afterload and maximise ventricular contractility
- > Hospitalisation to medically stabilise decompensated heart failure
- > Reduce salt intake < 2 g per day
- > Fluid restriction 1.5 – 2L per day



Medical treatment

- > Diurese with frusemide
- > In the prepartum period, reduce afterload by using hydralazine and nitrates
- > β -blockers may improve left ventricular systolic function and should be used if not contraindicated once the heart failure is in a compensated state
- > Angiotensin-converting enzyme inhibitors may be safely used to reduce afterload in the postpartum period
- > Digoxin may improve the symptoms of heart failure
- > Avoid or treat severe anaemia to prevent decompensated heart failure
- > Prophylactic anticoagulation while dilation of the heart persists (see anticoagulation prophylaxis)
- > Counsel regarding recurrence risk in future pregnancies. If the woman recovers fully and has normal ventricular function, future pregnancies may be considered

Heart transplant recipients

- > After heart transplant, approximately 90 % of otherwise healthy young female recipients survive the first year and almost two-thirds will survive the next ten years with an excellent quality of life. These women may desire pregnancy and children¹⁹
- > These women require high risk care by both transplant physicians and obstetric specialists in a tertiary centre with onsite specialist adult cardiac service care and adult intensive care
- > Pregnancy risks for heart transplant recipients include:
 - > Rejection
 - > Hypertension, pre-eclampsia
 - > Psychological stress
 - > Fetal growth restriction (20 %)
 - > Pre-term birth (up to 50 %)

Preconception counselling

- > Ideally, preconception counselling should be introduced at the pre-transplant stage and continue throughout the post-transplant process
- > Individual factors such as
 - > History of rejection in the first year
 - > Advanced maternal age
 - > Comorbidities (see below)
 - > Medical non-compliance should be considered when counselling
- > Comorbidities that may influence pregnancy outcome include
 - > Risk of recurrent disease
 - > Chronic allograft dysfunction
 - > Cardiovascular or pulmonary disease
 - > Diabetes mellitus
 - > Hypertension
 - > Inherited diseases-maternal or paternal (genetic versus chromosomal)
 - > Hepatitis B, hepatitis C, cytomegalovirus
 - > Obesity



- > Where possible, vaccinate pre-transplant or pre-pregnancy for influenza, hepatitis B, tetanus, pneumococcus
- > Discuss possible consequences of preterm birth, intrauterine growth restriction with both the woman and father of the baby

Timing of pregnancy

Before conception ensure:

- > No rejection in the past year
- > Adequate and stable graft function
- > Try to defer pregnancy for at least one year after transplantation
- > No acute infections that might impact the fetus
- > Maintenance immunosuppression is at stable doses

Antenatal care

- > Graft dysfunction during pregnancy warrants appropriate investigation (by biopsy if necessary)
- > Immunosuppression must be maintained during pregnancy to avoid rejection
- > Future studies need to address optimal selection and dosing of these agents
- > Hyperemesis gravidarum may lead to decreased absorption or inadequate immunosuppression
- > Caesarean section indicated only for obstetric reasons

Treatment goals:

- > Ensure that patients maintain graft function using appropriate immunosuppressive dosing during gestation and immediately after delivery
- > Optimise maternal health including graft function
- > Maintain a normal metabolic environment
- > Minimise complications associated with preterm birth
- > Detect and manage hypertensive complications especially preeclampsia
- > Ensure adequate fetal growth

Antibiotic prophylaxis

- > Generally, routine antibiotic prophylaxis is not recommended for women with valvular heart disease undergoing uncomplicated vaginal birth or caesarean section unless infection is suspected²⁰
- > Intrapartum antibiotic prophylaxis is recommended for vaginal birth complicated by amnionitis (suspected or proven) or prelabour rupture of membranes, and other suspected infections where one of the following cardiac conditions is present:
 - > Prosthetic heart valves
 - > Complex congenital heart disease
 - > Past history of endocarditis
 - > Surgically constructed systemic-pulmonary shunts, or conduits



Recommended antibiotic treatment

- > Ampicillin [or amoxicillin] 2 g IV as a stat dose as close as practical to the time of birth. Repeat dose after 8 hours if birth has not occurred

Allergy to penicillin

- > Vancomycin 25 mg / kg (up to 1.5 g) IV, administered slowly (over at least one hour) and repeated after 12 hours if birth has not occurred

Anticoagulation prophylaxis

- > Anticoagulation is indicated for women who have mechanical heart valves, atrial fibrillation or atrial flutter in the presence of structural heart disease, past documented cardiac embolic events, a past history of multiple pulmonary emboli, cyanotic congenital heart defects and Fontan circulation
- > Women with mechanical heart valves are at very high risk of thromboembolism in pregnancy and require continued anticoagulation
- > There are no definitive recommendations about optimal antithrombotic treatments for women with mechanical heart valves in pregnancy
- > Substantial concern remains about the fetal safety of warfarin, the efficacy of subcutaneous unfractionated heparin (UFH) and low molecular weight heparin (LMWH) in preventing thromboembolic complications, and the risk of maternal bleeding with the various regimens

A clinically appropriate anticoagulation regimen would be:

- > Warfarin should be avoided between 6 weeks and 12 weeks of gestation (to avoid embryopathy) and close to term (to avoid delivery of an anticoagulated fetus). The association of neuro-developmental problems with mid pregnancy use of warfarin should also be considered
- > Use of one of the following three regimens after explaining the associated risks with the woman:
 1. Either LMWH or UFH between 6 weeks and 12 weeks and close to term only. Use vitamin K antagonists e.g. warfarin at other times (despite fetal risks)
 - > If warfarin is used, adjust dose to attain a target INR of 3.0 (a lower therapeutic range of 2.0 to 3.0 can be used in women with mechanical bileaflet aortic valves)
 2. Dose-adjusted subcutaneous UFH throughout pregnancy
 - > Initiate subcutaneous UFH in high doses (start at 17,500-20,000 units every 12 hours) and adjust to prolong a 6 hour post injection APTT into the therapeutic range (at least twice control) or to attain an anti-Xa heparin level of 0.35 to 0.70 U / mL
 3. Dose-adjusted LMWH throughout pregnancy
 - > Administer twice a day in dose to achieve anti-Xa levels of 1.0 to 1.2 U / mL 4 to 6 hours after subcutaneous injection or according to weight.
- > Aspirin 100 to 150 mg / day may offer additional protection against thromboembolism



Cardiac conditions associated with high maternal and / or fetal risk during pregnancy (See [Table 1](#))

- > Primary pulmonary hypertension
- > Eisenmenger syndrome
- > Uncorrected coarctation of the aorta
- > Severe aortic stenosis (AS) (valve area < 1 cm²) with or without symptoms
- > Aortic regurgitation (AR) with NYHA functional class III-IV symptoms
- > Mitral stenosis (MS) with NYHA functional class II-IV symptoms and / or valve area < 1.5 cm²
- > Mitral regurgitation (MR) with NYHA functional class III-IV symptoms
- > Aortic and / or mitral valve disease resulting in severe pulmonary hypertension (pulmonary pressure > 75 % of systemic pressures)
- > Aortic and / or mitral valve disease with severe LV dysfunction (EF < 40 %)
- > Mechanical prosthetic valve requiring anticoagulation
- > Marfan syndrome with aortic complications with or without AR
- > History of peripartum cardiomyopathy with persistent left ventricular dysfunction and / or NYHA functional class IV

Management in labour

- > Ensure that adult resuscitation equipment is available, including the ability to perform a perimortem Caesarean Section within 5 minutes of Cardiac Arrest

Management team includes:

- > Obstetrician
- > Midwife
- > Cardiologist / physician
- > Anaesthetist
- > Intensive care specialist
- > Other allied health professionals as required

Thromboembolism prophylaxis

- > See 'Managing anticoagulation around birth' in the Thromboprophylaxis and Thromboembolic Disease in Pregnancy PPG (available www.sahealth.sa.gov.au/perinatal) for regional anaesthesia / analgesia considerations after antenatal administration of intravenous unfractionated heparin or subcutaneous low molecular weight heparin
- > Use anti-embolic stockings

Intrapartum antibiotic prophylaxis

- > Follow link to antibiotic prophylaxis in cardiac disease

Preterm labour

- > Notify cardiologist / physician if any cardiac advice is required quickly



Term Labour

- > Most women with cardiac disease should be encouraged to labour spontaneously
- > Oxytocin infusion for induction or augmentation of labour or prostaglandin E₂ may be used if indicated
- > In women with risk of fluid overload use low volume oxytocin infusion (Follow link to oxytocin regimen)
- > In fixed cardiac output disorders / aortic aneurysm women may be offered elective caesarean section
- > Ensure intravenous access
- > Maintain continuous midwifery presence and care

Pain management and anaesthesia

- > Nitrous oxide and other pain relief should be provided only after consultation with the anaesthetist
- > An epidural block and assisted vaginal birth may be advised after discussion between the woman, anaesthetist, obstetrician and others
- > Epidural anaesthesia may be hazardous in women with:
 - > A fixed cardiac output
 - > Aortic stenosis
 - > Mitral stenosis
 - > Hypertrophic cardiomyopathy
 - > Fontan circulation
- > Consider combined spinal-epidural anaesthesia (decreases the preload and afterload with a positive effect on most cardiac lesions)²
- > Regional anaesthesia (epidural or spinal) is contraindicated during anticoagulation treatment (risk of spinal haematoma)

Fluid management

- > Closely monitor fluid management in labour and maintain accurate fluid balance chart (increased risk of pulmonary oedema)

Oxygen treatment

- > Oxygen saturation should be continuously monitored with pulse oximetry. Regulate oxygen flow (via mask or nasal specs) to maintain oxygen saturations > 95 %

Signs of cardiac failure:

- > Observe for signs of cardiac failure e.g. dyspnoea (respirations > 24 per minute), tachycardia (> 130 beats per minute [bpm]), fatigue, oedema, cyanosis
- > Position woman in a supported upright sitting position

Continuous electrocardiographic (ECG) monitoring

- > Required in moderate to high risk women
- > Maternal cardiac monitoring should occur throughout labour and after birth for 24 hours, according to the severity of maternal condition
- > High risk women may require invasive cardiac monitoring in labour

Infection risks

- > Limit vaginal examinations to reduce risk of infection and possible subacute bacterial endocarditis
- > Bacterial endocarditis is a very rare, but serious risk in women with valvular abnormalities and valve replacements. Particular attention should be paid to preventing infection e.g. regular changing of intravenous lines in addition to bacterial endocarditis prophylaxis
- > Report a pulse rate of > 130 bpm and respiratory rate of > 24 respirations per minute (rpm) to senior responsible clinician (physician or cardiologist)

Electronic fetal monitoring

- > Continuous electronic fetal monitoring during labour

Observations

- > Take and record the following observations every fifteen minutes:
 - > Pulse
 - > Respiration
- > Take and record the following observations every hour:
 - > Intake and output
 - > Oedema
 - > Colour
 - > Blood pressure

High risk cardiac management in labour

- > Women with severe or barely controlled heart failure present significant problems during labour and birth
- > Labour and birth should occur in a hospital with an adult intensive care or cardiac unit

Timing of labour and birth

- > Increased risk of preterm labour
- > Before 30 weeks gestation, in partnership with the woman, the high risk management team (obstetrician, physician, cardiologist and anaesthetist) should discuss and agree on the time and mode of birth
- > Document plan in case notes
- > The time and mode of the birth is ultimately the obstetrician's decision
- > Aim to birth at term if cardiac state is well stabilised
- > Inform the woman of possible management change at short notice if cardiac function and reserve alter rapidly

During labour

- > Contact the cardiologist or physician, anaesthetist and paediatrician before birth
- > Consider Endocarditis antibiotic prophylaxis
- > Cardiac drugs during labour should be planned in consultation with microbiologist, obstetrician and cardiologist / physician



Birth

- > To shorten second stage and avoid excessive maternal expulsive efforts:
 - > Consider episiotomy
 - > Consider forceps delivery
 - > Do not use local anaesthetic with adrenaline
- > To minimise cardiac compromise
 - > Do not use stirrups without discussion between obstetrician, anaesthetist and cardiologist / physician
 - > Report a pulse rate > 130 or < 40 bpm
 - > Report respiratory rate > 24 or < 5 rpm
 - > Report rapidly changing pulse or respiration rate (even if within threshold)

Postpartum

Management of the third stage, including complications

- > This is potentially the time of most risk for haemodynamic compromise or decompensated heart failure
- > Many of these women are intolerant of changes in vascular resistance and venous return. Therefore care is required to avoid vasodilatation from bolus doses of oxytocin, whilst avoiding excessive bleeding from uterine atony

Oxytocic for active management of the third stage

- > Oxytocin 10 IU diluted to 9 mL with sodium chloride 0.9 % in a 10 mL syringe
- > Give 2 x 1 mL aliquots (i.e. oxytocin 1 IU) 30 seconds apart to produce uterine contraction following delivery of the anterior shoulder
- > Await classic signs of separation of the placenta before attempting to deliver placenta
- > if no signs of separation after 60 seconds give a third aliquot of 1 IU (1mL) of oxytocin

Oxytocin Infusion for prevention or control of PPH

- > Oxytocin 40 IU diluted in 100 mL bag of sodium chloride 0.9 % and run at 25 mL / hour
- > The oxytocin is delivered at the normal dose but in 1/10th the volume

Management of PPH

- > Prompt treatment important to avoid hypovolaemia and reduced preload
- > Large boluses of oxytocin are contraindicated.
- > Ergometrine and Prostaglandin F₂ alpha are contraindicated
- > Rub up a contraction manually and administer 1 mL of dilute oxytocin solution (10 IU in 10 mL)
- > Administer misoprostol (Cytotec®) 200 micrograms orally (1 tablet) and 800 micrograms rectally (4 tablets)
- > Judicious resuscitation with assistance of intensive care unit and anaesthetic colleagues



Observations

- > Observe woman very closely for 24 hours postpartum. This is a high risk period for the woman since blood volume increases from auto transfusion and after load increases as placental bed circulation ceases
- > Refer and report any signs of cardiac failure:
 - > Dyspnoea
 - > Cyanosis / pallor
 - > Tachycardia
 - > Cold clammy extremities
 - > Haemoptysis
 - > Oedema of the face and hands
- > Take and record the woman's respiration when pulse is taken
- > Continue to position the woman in a semi-upright position
- > Encourage deep breathing and passive leg exercises
- > The clinician should realise that maternal mortality associated with pulmonary hypertension and Eisenmenger syndrome may occur up to several weeks postpartum

Contraception

- > Sterilization of the male partner carries the least risk in monogamous couples who have completed their family
- > Oral contraceptives can be used with several exceptions:
 - > Women with right to left shunts
 - > Cardiac disease with associated hypertension
- > Progestin-only oral contraceptives or medroxy-progesterone acetate (Depro-Provera®) may be used by these women, as the thromboembolic risk of oral contraceptives is thought to be due to the oestrogen component.
- > The progestin-releasing intrauterine contraceptive device (IUCD) has a failure rate of < 1 % and in a monogamous couple the risk of endocarditis associated with the use of an IUCD is rare. Prophylaxis with oral amoxicillin (2 g taken one hour before insertion or removal) is recommended

Follow-up

- > Postnatal follow up should be at a high risk medical clinic and with the woman's cardiologist



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Additional Resources

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Appendix 1:

Table 2: Maternal mortality CEMACH (UK) 2003-2005

Type and cause of death	Indirect	Late
Acquired		
Aortic Dissection	9	0
Myocardial Infarction	12	4
Ischaemic Heart disease		0
Sudden Adult Death Syndrome	4	9
Peripartum Cardiomyopathy	3	12
Cardiomyopathy	0	4
Myocarditis or myocardial fibrosis	1	0
Mitral valve disease	5	0
Infectious endocarditis	3	2
Right or left ventricular hypertrophy or hypertensive heart failure	2	1
Congenital		
Pulmonary hypertension	3	0
Congenital heart disease	3	2
Total	47	34



Cardiac disease in pregnancy

Appendix 2:

Table 3: CEMACH (UK) 2003-2005 Indirect maternal deaths

Triennium	Congenital	Acquired		Total	Rate	95 per cent CI	
		Ischaemic	Other				
	n (%)	n (%)	n (%)	n (%)			
1985-1987	10 (43)	9 (39)	4 (17)	23 (100)	1.01	0.68	1.52
1988-1990	9 (50)	5 (28)	4 (22)	18 (100)	0.76	0.48	1.21
1991-1993	9 (24)	8 (22)	20 (54)	37 (100)	1.60	1.16	2.20
1994-1996	10 (26)	6 (15)	23 (59)	39 (100)	1.77	1.30	2.43
1997-1999	10 (29)	5 (14)	20 (57)	35 (100)	1.65	1.19	2.29
2000-2002	9 (20)	8 (18)	27 (61)	44 (100)	2.20	1.64	2.96
2003-2005	4 (8)	16 (33)	28* (58)	48 (100)	2.27	1.67	2.96



Appendix 3: FDA classification

Category B:

Either animal reproduction studies have not demonstrated a fetal risk but there are no controlled studies in pregnant women, or animal reproduction studies have shown an adverse effect that was not confirmed in controlled studies in women.

Category C:

Either studies in animals have revealed adverse effects on the fetus and there are no controlled studies in women, or studies in women and animals are not available. Drugs should be given only if potential benefits justify the potential risk to the fetus.

Category D:

There is positive evidence of human fetal risk, but the benefits from use in pregnant woman may be acceptable despite the risk.

Category X:

Studies in animals or human beings have demonstrated fetal abnormalities. The risk of the use of the drug in pregnant women clearly outweighs any possible benefit. The drug is contraindicated in women who are or may become pregnant.

Source: Drug Information for the Health Care Professional (USDPI Vol 1); Micromedex; 23rd ed (January 1, 2003). Adapted from Elkayam U. Pregnancy and cardiovascular disease. In: Zipes DP, Libby P, Bonow RO, Braunwald E, editors. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. 7th ed. Philadelphia, PA: Elsevier, 2005: 1965 (763).

Adapted from: Bonow RO, Carabello BA, Chatterjee K, de Leon AC Jr., Faxon DP, Freed MD, et al. ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology /American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Patients With Valvular Heart Disease).¹



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Cardiac disease in pregnancy

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