South Australian Perinatal Practice Guideline

Cardiac Disease in Pregnancy

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

Note: The words woman/women/mother/she/her have been used throughout this guideline as most pregnant and birthing people identify with their birth sex. However, for the purpose of this guideline, these terms include people who do not identify as women or mothers, including those with a non-binary identity. All clinicians should ask the pregnant person what their preferred term is and ensure this is communicated to the healthcare team.

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 woman. 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 respectfully 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 (PPG)

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.

Table 1: Modified WHO classification of maternal cardiovascular risk

	mWHO I	mWHO II	mWHO II-III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	Uncomplicated or mild – pulmonary stenosis – patent ductus arteriosus – mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats, isolated	Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Most arrhythmias (supraventricular arrhythmias) Turner syndrome without congenital cardiac disease	Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or bioprosthetic valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilatation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation without residua (non-Turner's) Atrioventricular septal defect	Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated Unrepaired cyanotic heart disease Other complex congenital heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm/m2, tetralogy of Fallot <50 mm) Ventricular tachycardia	Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m^2, tetralogy of Fallot >50 mm) Vascular Ehlers–Danlos Severe (re)coarctation Fontan with any complication
Risk	No detectable increased risk of maternal mortality and no/mild increased risk in morbidity	Small increased risk of maternal mortality or moderate increase in morbidity	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity	Significantly increased risk of maternal mortality or severe morbidity	Extremely high risk of maternal mortality or severe morbidity
Maternal cardiac event rate	2.5 – 5 %	6 – 10 %	11 – 19 %	20 – 27 %	> 27%
Counselling	Preconception + early pregnancy specialist consultation with cardiologist / obstetric physician	Preconception + early pregnancy specialist consultation with cardiologist / obstetric physician / MFM / anaesthetist	Preconception + early pregnancy with specialist consultation with cardiologist / obstetric physician / MFM / anaesthetist	Preconception + early pregnancy with specialist consultation with cardiologist / obstetric physician / MFM / anaesthetist	Specialist consultation: **Pregnancy contraindicated**. If pregnancy occurs, termination should be discussed
Minimum cardiologist / obstetric physician follow-up visits during pregnancy	Either cardiologist or obstetric physician in early pregnancy and at 36 weeks if stable	Either cardiologist or obstetric physician in early pregnancy and 4-12 weekly thereafter based on maternal condition	Obstetric physician 4-8 weekly from early pregnancy and Cardiologist as needed (e.g. once per trimester)	Obstetric physician 4 weekly from early pregnancy and Cardiologist as needed	Obstetric physician 2-4 weekly from early pregnancy and Cardiologist as needed (e.g. 4 weekly)
Care during pregnancy and place of birth	Local hospital (level 3-4)	Local hospital (min level 4) following obstetric physician & anaesthetic review	Referral hospital (level 5-6)	Referral hospital (level 5-6)	Tertiary hospital with onsite CCU and ICU (level 6)

ASI = aortic size index; EF = ejection fraction; HTAD = heritable thoracic aortic disease; mWHO = modified World Health Organization classification; NYHA = New York Heart Association; WHO = World Health Organization

Modified from mWHO Classification of Maternal Cardiovascular Risk[1(p3175)]

Table 2: New York Heart Association (NYHA) functional classification

Risk stratification tool for use in patients with known ventricular dysfunction; not to be generally applied in pregnancy.

Class	Symptoms	Objective Assessment
I	No limitation of physical activity. Exercise does not cause fatigue, palpitations or dyspnoea	No objective evidence of cardiovascular disease. No symptoms or limitations.
II	Comfortable at rest. Slight limitation of physical activity as symptomatic (fatigue, palpitation and dyspnoea) with exercise	Objective evidence of minimal cardiovascular disease. Mild symptoms and slight limitation during ordinary activity
III	Comfortable at rest. Marked limitation of physical activity as light exercise or ordinary activity results in symptoms	Objective evidence of moderately severe cardiovascular disease. Marked limitation in activity due to symptoms
IV	Symptomatic at rest. If any physical activity undertaken, symptoms worsen	Objective evidence of severe cardiovascular disease. Activity severely limited with symptoms at rest

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Summary of Practice Recommendations

Preconception Care

- Preconception counselling by MFM / obstetrician / obstetric physician / cardiologist

Antenatal Care

Low risk (mWHO Class I and Class II)

- Tertiary referral hospital (Level 5, 6)
  OR
- Local hospital (level 3-4) after obstetric physician or cardiology review and plan

Intermediate (mWHO Class II-III) **or high risk** (mWHO Class III and Class IV)

- Tertiary referral hospital (level 5-6) with multidisciplinary care from obstetrician / Maternal Fetal Medicine (MFM), obstetric physician, anaesthetist, intensivist, neonatologist, midwifery and nursing

Antenatal Management

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

Intrapartum Management

Low and intermediate risk

- Epidural anaesthesia
- Cardiac telemetry and oximetry if required
- Deep venous thrombosis (DVT) prophylaxis (see *Thromboprophylaxis and Thromboembolic Disease in Pregnancy* PPG available at www.sahealth.sa.gov.au/perinatal)

High Risk

- Multidisciplinary approach including decision on place of birth.
- Planned birth at term
- Aim for vaginal birth except in specific instances (e.g. Marfan syndrome, critical aortic stenosis)
- Care in labour and birth unit, coronary or intensive care unit if indicated.
- Continued careful observation using special observation charts including systems such as 'Early warning scores' for signs of clinical deterioration in the 4th stage

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
MFM	Maternal Fetal Medicine
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

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 defects[2]

Specific hazards for women with cardiac disease in pregnancy include:

- Pulmonary oedema (main cause of death)
- Pulmonary hypertension
- Infective endocarditis
- Thromboembolism
- Peripartum cardiomyopathy
- Aortic dissection
- Primary arrhythmogenic disorders

The most frequent causes of cardiac disease in pregnancy are:

- Rheumatic heart disease
- Congenital heart disease
- Arrhythmias
- Cardiomyopathy

Fetal risks:

- Congenital heart disease
- Intrauterine growth restriction
- Prematurity

Preconception care

Assessment of the woman's general physical condition, medications and cardiac function:

- An exercise test (with VO_2 max measurements) and echocardiogram provide essential information on pre-pregnancy cardiac status and reserve[3]
- Assess lifestyle risk factors e.g. smoking, illicit drug use and education to optimise exercise tolerance
- Stablise complications such as arrhythmia and fluid overload where relevant
- 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 for use in pregnancy. Information is also available from SA Pharmacy Medicines Information Service, telephone 81617555
- Optimise management of other co-morbidities including hypertension, diabetes, hyperlipidaemia. 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 indicated by Modified World Health Organization (mWHO) classification of maternal cardiovascular risk[1] (see Table 1)
- 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)[2,4]

Antenatal management

Women who identify as Aboriginal should be offered the choice to have an Aboriginal Health Professional (Aboriginal health practitioner, Aboriginal midwife, Aboriginal maternal infant care worker or Aboriginal liaison officer) present to ensure cultural safety, appropriateness of care and support.

Early review at 6-10 weeks

Women with more complex cardiac lesions require early review (6-10 weeks gestation) by their current cardiologist or an obstetric physician to enable cessation of contra-indicated medications, updated assessment of cardiac function and planning of antenatal care.

Routine booking visit around 10 to12 weeks with medical input

Women should be managed by a multi-disciplinary team, which might include an obstetrician / MFM, cardiologist or obstetric physician, anaesthetist and intensivist. These clinicians should all be familiar with the management of cardiac problems in pregnancy.

Women should be involved in discussions about the management of the pregnancy:

- Decision about the most appropriate facility for the woman to birth and criteria for changing the place of birth (see Table 1)
- Advice on avoiding anaemia – iron supplementation, prenatal vitamins, dietary counselling and regular haemoglobin checks

Ongoing antenatal care

- Antenatal visits in medical clinic (frequency and care provider to be decided by severity of disease)
- Regular review of maternal adaptation to the pregnancy and fetal growth and wellbeing review at each visit
- Regular review of functional cardiac status at each visit
- Echocardiograms with timely follow up by obstetrician / MFM and cardiologist / obstetric physician
- Anaesthetic review before 28 weeks
- Ongoing discussion regarding work schedules and levels of activity

Clinical assessment

History

- Maternal age, gestational age, parity status, mWHO category, 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 if no appropriate recent study available; timeframe depends on lesion and stability of lesion. Aim to determine:
  - Systolic and diastolic ventricular function
  - Doppler quantification of inflow or outflow obstruction
  - Doppler quantification of valvular function, including width and gradient
  - Right heart pressures and systolic function if measurable[5]

Fetal wellbeing

Women with lesions of mWHO Class II-III or higher, especially cyanotic heart disease and left heart obstruction, in addition to current cigarette use, 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
Congenital heart disease

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 disease.[1] 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 and channelopathies have a 50 % chance of being inherited, but may not be associated with any structural abnormalities in the fetus.

Early diagnosis of congenital heart defect in the fetus allows appropriate counselling and informed decision making, including neonatal medical or surgical treatment, palliation or termination of the pregnancy (TOP). Under current South Australian legislation, termination is permissible up to 22^{+6} weeks gestation. This will change under current legislative reform. See *Perinatal Loss* PPG available at www.sahealth.sa.gov.au/perinatal.

Consideration should be given to tertiary level morphology ultrasound +/- fetal echocardiogram with referral to Maternal Fetal Medicine sub-specialist.

Affected fetuses should be born in a tertiary care centre (Level 5, 6) with paediatric cardiac specialist support and a Paediatric Intensive Care Unit as per the SA *Maternal & Neonatal Services Standards* available at www.sahealth.sa.gov.au/perinatal. This is particularly important if they have a duct dependent cardiac anomaly requiring a prostaglandin infusion.

Intrauterine Growth Restriction and Prematurity

The two main determinants of fetal prognosis (apart from congenital anomaly), are maternal functional status and the degree of maternal cyanosis. When the mother is in mWHO Class II/III or higher (e.g. has a high risk disease such as severe aortic stenosis or Eisenmenger syndrome or is cyanosed), monitoring of fetal growth is very important.[1]

In Australia and New Zealand, neonatal survival ranges from 68 % (approximately 50 % without permanent impairment) at 24 weeks gestation to 94 % (approximately 70 % without permanent impairment) at 27 weeks gestation.[6] MFM input to assist in optimising gestation at birth, particularly when fetal growth issues or fetal compromise are present may be useful.

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 birth as long as possible in collaboration with the MFM or obstetric team.[1]

The choice between corrective procedures and planned birth may be difficult between 28 and 32 weeks, and decisions must be individualised.

Administer antenatal corticosteroids if birth is likely to occur between 23^{+0} and 34^{+0} weeks or up to 37^{+0} weeks if birth is via elective caesarean section. See *Preterm Labour and Birth* PPG. Administer magnesium sulphate if birth is between 23^{+0} and 32^{+0} weeks. See *Magnesium Sulphate for Neuroprotection of the Fetus* PPG. PPGs available at www.sahealth.sa.gov.au/perinatal.

Level of hospital care (see **Table 1**)

Initial assessment of mWHO classification, functional status, echocardiographic assessment results and frequency of complications such as arrhythmia and pulmonary oedema will determine if the level of hospital care required is:

- Hospital of choice / 'low risk' (Level 3)
- Hospital with 'moderate risk' capability (Level 4)
- Tertiary centre (Level 5 or 6)
- Tertiary centre with onsite specialist adult cardiac service 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 birth at a higher level unit is required. This should be negotiated on a case-by-case basis with a documented plan of care in both the woman's medical record and SA Pregnancy Record.

Women with mWHO Class I lesions may be suitable for care at a Level 3 hospital with obstetric physician or cardiology support. Referral for higher level care may be required if her cardiac status deteriorates.

Women with mWHO Class II lesions may be suitable for care at a Level 4 hospital with obstetric physician or cardiology support and following anaesthetic review. A plan for birth (including escalation of care) should be documented in both the SAPR and medical record. Referral for higher level care may be required if her cardiac status deteriorates.

Women with mWHO Class II-III lesions or higher should be referred to their cardiologist and obstetric physician for discussion regarding an antenatal care plan and appropriate site for birth, with view to birthing in a level 5 or 6 hospital. This discussion should include a multidisciplinary team approach for more complicated lesions.

If the woman's cardiac condition is stable:

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

SPECIFIC CONDITIONS

Benign arrhythmias

Sinus tachycardia, premature atrial and ventricular contractions (mWHO Class I)

- The majority of women with arrhythmias during pregnancy have a benign increased rate of atrial or ventricular premature beats.[9] 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 with exercise: Holter monitoring and echo may be useful in excluding more sinister arrhythmia and providing reassurance.
- Haemodynamically stable women do not usually require treatment
- Advise woman to avoid precipitating factors such as caffeine, alcohol, tobacco, fatigue and anxiety
- Management of iron deficiency, anaemia and gastro-oesophageal reflux can reduce the frequency of episodes.

Other arrhythmias

Women with more significant atrial arrhythmias (**supraventricular tachycardia, atrial fibrillation and flutter – mWHO Class II**) should be managed in collaboration with a cardiologist or obstetric physician, usually using the same agents that would be chosen in the non-pregnant woman, including electrical cardioversion when necessary.[7]

- 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 is the first line agent to terminate an SVT. This must be given as a rapid intravenous bolus: initial dose 6mg, repeat dose 6-12mg. Intravenous administration of adenosine can cause a sinus pause: the patient should be connected to continuous rhythm monitoring with at least one doctor and one nurse / midwife present during administration
- 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 non-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 in cases of drug refractory and poorly tolerated SVT.[1]

- 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 torsdae 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
- Flecainide is recommended for prevention of SVT in patients with Wolf Parkinson White Syndrome (WPW)[1]
- 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 (SAPR).
- The presence of a pacemaker or ICD should not alter the mode of birth. 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 woman should have continuous ECG monitoring and external defibrillation available at all times

Rheumatic heart disease (RHD) and mitral stenosis

mWHO Class II / III and above, depending on severity of mitral stenosis

The incidence of rheumatic heart disease is decreasing in developed countries. Mitral stenosis is the most common sequelae. 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 disease.[8]

Rheumatic fever and RHD are both notifiable conditions in South Australia. For more information, including reporting forms, fact sheets for patients/families and health professionals, go to the SA Health website or contact the SA RHD program direct on email HealthRHD@sa.gov.au or phone (08) 7425 7146.

Acute rheumatic fever and RHD disproportionally affects Aboriginal people with acute rheumatic fever now endemic (2 % of Aboriginal people in SA), with the majority of admissions in the younger population (< 35 years).[9]

A cardiovascular risk assessment should be obtained in Aboriginal women, including a targeted cardiovascular history and examination, with careful auscultation of the woman's heart for abnormal rhythm, sounds and murmurs. Direct questioning should be made regarding a history of penicillin injections and/or prolonged hospital admissions as a child, other family members with RHD, or if she is symptomatic with physical activity. There should be a low threshold for performing a screening echocardiogram if suspicion is raised from this medical history and examination.

Women who identify as Aboriginal should be referred to an Aboriginal health professional to support their care. Perinatal service providers need to consider cultural sensitivity within a non-judgemental environment when planning care with Aboriginal women and their families.

Mitral stenosis

Classification of mitral stenosis

	Mild	Moderate	Severe
mWHO Class	Class II/III	Class III	Class IV
Valve area	>1.5cm²	1 – 1.5cm²	<1cm²
Mean gradient	<5mmHg	5 – 10mmHg	>10mmHg
Pulmonary artery systolic pressure	<30mmHg	30-50mmHg	>50mmHg
Assumes sinus rhythm 60 – 80bpm			

Table adapted from ACC/AHA 2006 Guidelines for the Management of Patients with Valvular Hear Disease[10]

Women with mild 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. Women with moderate or severe mitral stenosis may have symptoms prior to pregnancy, and are likely to become more symptomatic during pregnancy. Ideally, women with severe mitral stenosis should have their valve lesion repaired prior to pregnancy.[2,1]

The risk of complications during pregnancy is increased in the presence of atrial fibrillation or in women with a history of thromboembolic events or pulmonary oedema.

- Manage according to symptoms and with the guidance of echocardiography, which is the key to the assessment of valvular severity.[2,1]

Medical treatment

- Limitation of exercise and fluid and salt restriction should be used in women with moderate to severe stenosis
- 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)[8]
- 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 of atrial fibrillation 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 birth is determined by obstetric indications only
- Women with mild, non-complicated mitral stenosis can be managed in the delivery suite / labour ward of a tertiary centre without invasive monitoring. ECG monitoring to monitor maternal heart rate would be preferable and should be available
- Women with moderate to severe mitral stenosis should be referred to a hospital with specialist adult cardiac service and capacity for invasive monitoring in labour. These women should be managed in an intensive care setting with invasive monitoring if required, 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, with attention to fluid administration (do not overload) is necessary

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 after the first trimester and therefore contraindicated or only used as a last resort
- Previously asymptomatic women may worsen immediately after birth because of sudden increases in systemic vascular resistance: avoid fluid overload in labour and for 24 hours post-partum (e.g. fluid restriction of 3L in 24 hours unless requires resuscitation)
- 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. mWHO Class depends on severity of stenosis and presence of symptoms.

	Mild	**Moderate**	**Severe**
mWHO Class	Class II / III	Class II / III	Class III or IV, depending on symptoms
Vmax (m/s)	2.0 – 2.9	3.0 – 3.9	≥ 4.0
Mean gradient (mmHg)	<30	30 - 49	≥50
Aortic valve area (cm²)	>1.5	1.0 – 1.5	<1.0
Indexed AVA (cm²/m² BSA)	≥1.0	0.6-0.9	<0.6

Table adapted from ACC/AHA 2006 Guidelines for the Management of Patients with Valvular Hear Disease[10]

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.
- 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
- 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 function[8]

Aortic regurgitation

mWHO Class II-III

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. 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
- Cardiac surgery for aortic regurgitation (valve replacement) is high risk to mother and fetus and can usually be deferred until after birth (aortic regurgitation is well tolerated in pregnancy)

Congenital heart disease

mWHO classification depends on the lesion; many women may remain largely asymptomatic. May be organised into acyanotic and cyanotic types:

Acyanotic congenital heart lesions include:

- Atrial / ventricular septal defects (mWHO Class I if successfully repaired, Class II if unrepaired),
- Patent ductus arteriosus(mWHO Class I)
- 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

The risk of a cardiac event in pregnancy is increased for women with:

- A prior cardiac event or arrhythmia
- mWHO Class II or higher lesions
- 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.[1,11] Women with a documented aortic root diameter of < 40 mm without an abnormal aortic valve have a mortality rate of < 5 % and generally tolerate pregnancy well. Over 100 pregnancies have been reported in the literature with no deaths. Worsening of systemic ventricular function during or shortly after pregnancy occurred in 10 % of the reported cases.[1]

Risk of aortic dissection during pregnancy or shortly thereafter is approximately 10 % with the incidence appearing to increase with increasing aortic root diameter.[1]

Aortic or mitral regurgitation is also seen in 60 % of women with Marfan syndrome and may complicate pregnancy.[11]

- 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 when pregnancy is confirmed[1]
- 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 cervical insufficiency
- Physical, echocardiographic, and ophthalmologic examination of newborns is recommended
- Counsel woman regarding the risk of autosomal dominant inheritance and need for follow- up for their offspring[11]
- Intra-atrial repair for transposition of great arteries (TGA)

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 complications.[1]

Fontan procedure

mWHO Class III and IV

Careful patient selection is important. The successful Fontan Procedure with a small right atrium or total cavopulmonary connection (TCPC) in mWHO Class III 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 dissection.[12]

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 disorders[13]

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 murmur[13]

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
    - Gadolinium crosses the placenta and a case controlled study found a slightly higher incidence of stillbirths and early neonatal deaths. Furthermore, gadolinium exposure in pregnancy was associated with an increased risk of rheumatological, inflammatory, and infiltrative skin conditions in the offspring. MRI in the first trimester, without gadolinium, is not associated with increased risk of harm to the fetus.
- The risks of ionizing radiation to the pregnancy from a contrast CT of the chest are small, particularly with the newer CT scanners. Therefore, a contrast CT Aortogram should not be withheld if aortic dissection is suspected.

Treatment

The 2017 ACC/AHA/AATS guidelines[14] 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 section 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. Beta blockers are highly effective delivered intravenously to reduce wall stress and prevent hypertension. Most recent guidelines recommend titrating to a target heart rate of 60 beats per minute. If beta blockers are contraindicated, non-dihydropyridine calcium channel blocking agents are recommended for second line use. 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 pregnancy[7]
- Women with mechanical valves require continuous anticoagulation prophylaxis and should receive a detailed discussion of the advantages, disadvantages and timing of the anticoagulant options (warfarin, and low molecular weight heparin [LMWH]) (see Anticoagulation below)
- Despite the known fetal risks of warfarin, this can still be considered in pregnancy because of increased risk of valve thrombotic complications while on LMWH. A detailed discussion of the options should be held with the patient.
- 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 trimester[15]
- **Women on warfarin cannot attempt normal birth if warfarin has been used in the 10-14 days prior to birth** due to the risk of haemorrhage in the neonate. Women must have a caesarean section or transition to LMWH at 36 weeks or 2 weeks prior to planned birth. An INR should be performed to confirm normalisation prior to birth

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 death).[16]

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 failure.[16]

Women with Eisenmenger syndrome are at particular risk of increased right-to-left shunting. Hypoxia can lead to syncope and sudden death.

- When pregnancy occurs and TOP 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 ICU[16]

Ischaemic heart disease

Includes **acute myocardial infarction (MI), unstable angina, stable angina**

Rare in pregnancy: Ischaemic heart disease occurs on average in 1 in 10,000 pregnancies.

Myocardial infarction (MI) in pregnancy occurs on average in 7.5 in 100,000 pregnancies with mortality highest in the third trimester in women under 35 years of age.[17]

There is a higher incidence of non-atherosclerotic MI in pregnant women with MI than in aged-matched non-pregnant women with MI

CEMCH 2015-2017[18] 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 history

Women with ischaemic heart disease require care in a tertiary centre or a tertiary centre with specialist adult cardiac service care depending on echocardiograph findings and symptoms.

Myocardial infarction (MI) management

Similar to management of the non-pregnant woman with MI and includes:

- Oxygen if hypoxia is present, nitrates, morphine, and most anti-arrhythmics, calcium channel blockers, β-blockers, low-molecular weight heparin, low-dose aspirin, and invasive hemodynamic monitoring if necessary
- Percutaneous interventions and systemic and local thrombolytic therapy have also been successful.[17] Therefore the woman should be considered for coronary angiography and stenting as in the non-pregnant woman, with an awareness that the risk of coronary artery dissection is greater in pregnant women than non-pregnant women. PTCA should be undertaken by the most highly experience operator available.

MI is not an indication for immediate birth (mortality is higher in women who birth within 2 weeks of MI).

- Caesarean section only for obstetric 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, peripartum cardiomyopathy, anaemia or previously undiagnosed rheumatic or congenital heart disease.[17]

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 peripartum 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 birth.

- Counsel women with persisting left ventricular dysfunction to avoid pregnancy / consider termination of pregnancy (mWHO Class IV)

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 & Signs

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

Investigations

- Complete blood count, electrolytes, renal function and arterial blood gases
- Electrocardiogram
- Chest X-ray
- 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 furosemide
- In the antepartum 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
- Consider bromocriptine which improve LV recovery and clinical outcome in women with acute severe PPCM. Bromocriptine (2.5 mg once daily) for at least 1 week may considered in uncomplicated cases. Prolonged treatment (2.5 mg twice daily for 2 weeks, then 2.5 mg once daily for 6 weeks) may be considered in patients with EF <25% and/or cardiogenic shock.
- 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 after a functional cardiac assessment

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 children.[19]

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 %)
- Preterm birth (up to 50 %)

Preconception counselling

Ideally, preconception counselling by both an obstetric/MFM specialist and the cardiology team should be introduced at the pre-transplant stage and continue throughout the post-transplant process.

Consider 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 and COVID 19.

Discuss possible consequences of preterm birth, intrauterine growth restriction with both the woman and her partner.

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
- Monitor fetal growth

Antibiotic prophylaxis

Unless active infection is suspected, routine endocarditis antibiotic prophylaxis is not recommended for women with congenital heart disease or prosthetic valves undergoing uncomplicated vaginal birth or caesarean section.[20] However, for women with complex congenital heart disease, heart transplant or with a significant amount of prosthetic material, the decision regarding prophylaxis should be determined on a case-by-case basis in consultation with the woman's cardiologist.

Intrapartum antibiotic prophylaxis is recommended for vaginal birth complicated by chorioamnionitis (suspected or proven) or prelabour rupture of membranes (PROM), and other suspected infections where women are at highest risk for intrapartum endocarditis.

Antibiotic prophylaxis should be administered for women with suspected or proven infection with the following conditions as they are at highest risk for intrapartum endocarditis:

- Any prosthetic valve, including a transcatheter valve, or those in whom any prosthetic material was used for cardiac valve repair.
- Previous episode of infective endocarditis.
- Congenital Heart Disease (CHD) if it involves:
    - Unrepaired cyanotic defects, including palliative shunts and conduits

- Completely repaired defects with prosthetic material or devices, whether placed surgically or by percutaneous techniques, up to 6 months after the procedure or lifelong if residual shunt or valvular regurgitation remains.

- Cardiac transplantation with the subsequent development of cardiac valvulopathy
- Rheumatic heart disease in Aboriginal women and individuals at significant social disadvantage
- If uncertain, contact the woman's cardiologist or an infectious diseases specialist
- Refer to the *Antibiotics in the Peripartum Period* PPG for recommended antibiotic treatment for chorioamnionitis, PROM and endocarditis prophylaxis, (at www.sahealth.sa.gov.au/perinatal). Also, See SA Health Surgical Antimicrobial Prophylaxis Clinical Guideline for more information.

Anticoagulation

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. Valve thrombosis can occur in 4.7% of pregnancies and appear to be higher with LMWH use throughout pregnancy (5.8-7.4%) than with INR adjusted warfarin (0-4%).[1] Maternal mortality after valve thrombosis has been reported as 10-20%.

Subcutaneous UFH carries a high risk of valve thrombosis (9-33%), and should not be used for anticoagulation in women with prosthetic valves.[1] Continuous intravenous infusion of UFH may be appropriate for anticoagulation around delivery or procedures with a high risk of bleeding.

Although INR adjusted warfarin appears more effective in prevention of valve thrombosis, it comes at greater fetal risk. Miscarriage rates with the use of warfarin in the first trimester can be up to 28.6%. Fetal loss rates of a combined heparin/warfarin regimen is 22.7% compared to 12.2% with use of LMWH throughout pregnancy.

Warfarin use in the first trimester results in embryopathy (limb defects, nasal hypoplasia) in 0.6 - 10% and is dose related. Early morphology ultrasound at 16 weeks gestation may be helpful to identify issues. There is also the risk of ocular and CNS haemorrhage in the second and third trimesters with warfarin use.

There are no definitive recommendations about optimal antithrombotic treatments for women with mechanical heart valves in pregnancy.[1] Due to the challenge of striking a balance between maternal safety and fetal safety as well as the risk of maternal haemorrhage, discussion around anticoagulation for women with prosthetic valves should be undertaken by clinicians with experience in the area.

A clinically appropriate anticoagulation regimen might include:

1. Dose-adjusted LMWH throughout pregnancy
    a. Administer twice a day in dose to achieve anti-Xa levels of 1.0 to 1.2 U / mL 4 hours after subcutaneous injection and >0.6 immediately prior to injection
    b. Regular anti-Xa levels are required because of the increased renal clearance of LMWH in pregnancy; weight-based LMWH regimes carry a higher risk of valvular thrombosis than dose-adjusted regimes. This regime requires a high degree of patient compliance with monitoring and access to an appropriate collection centre. The staff at the collection centre must be aware of the timing sensitivity and appropriate transport of the samples for processing
2. Dose-adjusted LMWH between 6 weeks and 12 weeks (or 14 weeks in women with a history of miscarriage) and 10-14 days prior to birth only.
    a. Use warfarin at other times (despite fetal risks)
    b. Adjust dose to attain a target INR of 2.0 - 3.0 depending on the site and nature of the valve
    c. Intravenous unfractionated heparin should be used in the 24-36 hours prior to planned birth, in preference to LMWH (see "Managing anticoagulation around birth" in the *Thromboprophylaxis and Thromboembolic Disease in Pregnancy* PPG available at www.sahealth.sa.gov.au/perinatal)

Note: Aspirin 100 mg / day may offer additional protection against thromboembolism, but is likely to increase the risk of haemorrhagic complications. If indicated for placental reasons, it could be ceased after 16-20 weeks.

Cardiac conditions associated with high maternal and / or fetal risk during pregnancy

Risk of maternal mortality and morbidity increases according to the mWHO classification, with women classified as mWHO IV at extremely high risk of maternal mortality or severe morbidity and associated fetal complications. See Table 1.

Management in labour

Ensure that adult resuscitation equipment is available. In very high risk cardiac conditions there should be the ability to perform a perimortem caesarean section within 4 minutes of Cardiac Arrest.

Management team includes:

- Obstetrician
- Midwife
- Cardiologist / Obstetric 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 at www.sahealth.sa.gov.au) for regional anaesthesia / analgesia considerations after antenatal administration of intravenous unfractionated heparin or subcutaneous low molecular weight heparin.
    - Women on warfarin for prosthetic valves should not be given Vitamin K for reversal, FFP should be used instead
- Use anti-embolic stockings

Intrapartum antibiotic prophylaxis

- See Antibiotic Prophylaxis section or the *Antibiotics in the Peripartum Period* PPG (available at www.sahealth.sa.gov.au/perinatal)

Preterm labour

- Notify cardiologist / obstetric physician if any cardiac advice or assistance with anticoagulation is required quickly.

Term Labour

Most women with cardiac disease can labour spontaneously:

- Oxytocin infusion for induction or augmentation of labour or prostaglandin E_2 may be used if indicated
    - In women with risk of fluid overload use low volume oxytocin infusion (see *Induction and Augmentation of Labour* PPG available at www.sahealth.sa.gov.au/perinatal)
- In fixed cardiac output disorders or great vessel or valvular pathologies, 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.
- Neuraxial anaesthesia (epidural or spinal) may be hazardous in women with:
    - A fixed cardiac output
    - Severe Aortic stenosis
    - Severe Mitral stenosis

  - Hypertrophic obstructive cardiomyopathy
  - Fontan circulation
- Neuraxial anaesthesia (epidural or spinal) is contraindicated during therapeutic 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)
- In many cases, restriction of total fluid intake to ≤ 3L/24 hours during labour and for 24 hours post-birth (unless resuscitation is required) is sensible

Oxygen treatment

- Oxygen saturation should be continuously monitored with pulse oximetry
- Regulate oxygen flow (via mask or nasal specs) to maintain oxygen saturations > 95 %
- Oxygen is not required for women with normal oxygen saturations

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 a serious risk in women with some forms of complex congenital heart disease, valve replacements and previous history of endocarditis. 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 (obstetric physician or cardiologist)

Electronic fetal monitoring

- Continuous electronic fetal monitoring during labour is recommended

Observations

Take and record the following observations every fifteen minutes:

- Pulse
- Respiration

Take and record the following observations every hour:

- Intake and output (maintain accurate fluid balance)
- 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 (MFM / obstetrician, physician, cardiologist and anaesthetist) should discuss and agree on the time and mode of birth
- Document plan in medical record and SA Pregnancy Record
- The time and mode of the birth is ultimately the MFM / 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 obstetric physician, anaesthetist and paediatrician before birth
- Consider endocarditis antibiotic prophylaxis
- Cardiac drugs during labour should be planned in consultation with microbiologist, obstetrician and cardiologist / obstetric physician
- Maintain a strict record of the woman's fluid balance
- Consider the liberal use of neuraxial anaesthesia if not contraindicated (see Pain management and anaesthesia above)

Birth

To shorten second stage and avoid excessive maternal expulsive efforts:

- Allow the fetal head to descend into the maternal pelvis (ideally under epidural block) with uterine contractions only
- If maternal and fetal condition allow, delay maternal pushing until the fetal head is on view. Avoid excessive Valsalva efforts by the woman
- Consider episiotomy and forceps delivery on usual obstetric indications
    - Do not use local anaesthetic with adrenaline for episiotomy infiltration

To minimise cardiac compromise:

- Prior to the use of stirrups, discuss the possible increase in cardiac perlaod generated to the woman with the attending anaesthetist
- Report a pulse rate > 130 or < 40 bpm and perform a 12-lead ECG
- Report respiratory rate > 24 or < 5 rpm
- Report rapidly changing pulse or respiration rate (even if within threshold)

Management of third stage

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.

Avoid ergometrine.

Oxytocic for active management of the third stage

- Oxytocin 10 units diluted with 9 mL with sodium chloride 0.9 % in a 10 mL syringe
- Give 2 x 1 mL aliquots (i.e. oxytocin 1 unit) 30 seconds apart to produce uterine contraction following delivery of the anterior shoulder
- Await classic signs of separation of the placenta (uterine fundal rising, lengthening of cord and gush of blood PV before attempting to deliver placenta
- If no signs of separation after 60 seconds give a third aliquot of 1 unit (1mL) of oxytocin
- Consider tranexamic acid 1g prophylactically
- Consider oxytocin infusion for prevention or control of PPH
    - Oxytocin 40 units diluted in 100 mL bag of sodium chloride 0.9 %, run at 25 mL/hour
    - The oxytocin is delivered at the normal dose but in 1/10th the volume

Management of PPH

- Prompt treatment is 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, ensure that the uterus is empty, insert a urinary catheter and administer 1 mL of dilute oxytocin solution (10 units in 10 mL)
- Administer misoprostol (Cytotec®) 200 micrograms orally (1 tablet) and 800 micrograms rectally (4 tablets)
- Administer tranexamic acid 1g
- Judicious resuscitation with assistance of intensive care unit and anaesthetic colleagues
- Vitamin K should not be given to women on warfarin for anticoagulation in the context of prosthetic valves; reversal should be with FFP if required

Postpartum

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.

Fluid restriction of 3L (or less) in 24 hours unless resuscitation is required may be indicated during labour and for 24 hours to reduce risk of pulmonary oedema. This applies to any valvular lesion (including mitral valve prolapse) and any compromise of myocardial function.

- 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

- Sterilisation 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
  - Women with significant atrial or ventricular dilation and reduced ventricular function have increased risk of intracardiac thrombosis
  - Cardiac disease with associated hypertension
    - Progesterone-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 progesterone-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. There is no longer a requirement for antibiotic prophylaxis with insertion or removal of IUCD. See SA Health Surgical Antimicrobial Prophylaxis Clinical Guideline for more information.

Follow-up

Postnatal follow-up should be with an obstetrician, a high-risk medical clinic / obstetric physician and with the woman's cardiologist. For rural women it may be possible to have follow-up with their local GP with the cardiologist linking via telehealth. Suitability for this option should be assessed on an individual basis with a documented plan communicated with the GP.

Aboriginal women should be consulted and included in decisions regarding postnatal follow-up plans, supported by their nominated Aboriginal health professional. Where possible Aboriginal Community Controlled Health Services or Local Health Networks should be involved in follow-up care and plans if transferring back to rural/remote communities.

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Appendices

Appendix 1: Cardiovascular Drugs in Pregnancy and Breastfeeding

Medication	Use in pregnancy	Potential fetal / neonatal effects	Use in breastfeeding
Adenosine	Maternal supra-ventricular arrhythmias	No adverse fetal effects reported; data on use during first trimester are limited	Limited data. Unlikely to have any adverse neonatal effects due to short duration of action and limited oral bioavailability.
Amiodarone	Maternal arrhythmias	IUGR, prematurity, congenital goiter, hypothyroidism and hyperthyroidism, transient bradycardia, and prolonged QT in the newborn, premature birth	Not recommended for chronic use. If used briefly (3-7 days) a 24-48 hour interruption is advised before re-instating breastfeeding. Monitor infant for cardiovascular and thyroid function.[22]
Angiotensin-converting enzyme inhibitors / Angiotensin receptor II antagonists	Hypertension, heart failure	Oligohydramnios, IUGR, renal or tubular dysplasia, anemia, skull ossification defect, lung hypoplasia, limb contractures, IUFD Cease in the first trimester	Captopril Enalapril Perindopril Compatible. Limited data on other ACEIs and ARAs
Apixaban	Direct oral anticoagulant	Not recommended. Limited human data	Not recommended, consider alternative. Excreted in milk in animal studies No human data[23]
Beta blockers	Hypertension, maternal arrhythmias, myocardial ischemia, mitral stenosis, hypertrophic cardiomyopathy, hyperthyroidism, Marfan syndrome, migraine prophylaxis	Fetal bradycardia, low placental weight Labetalol: first line anti-hypertensive Atenolol: use in pregnancy associated with IUGR	Metoprolol, labetalol and propranolol are preferred to atenolol as they are more extensively protein bound and therefore less likely to pass into breast milk.
Digoxin	Maternal and fetal arrhythmias, heart failure	No evidence for unfavorable effects on the fetus	Compatible
Diltiazem	Myocardial ischemia	Limited human data, no increased risk of congenital malformations. High doses have resulted in embryo and fetal toxicity in animal studies	Compatible

Medication	Use in pregnancy	Potential fetal / neonatal effects	Use in breastfeeding
Diuretics	Congestive heart failure, Postpartum pulmonary oedema	Hypovolemia leads to reduced uteroplacental perfusion, , hyponatremia, hypokalemia. Fetal thrombocytopenia has been reported with thiazide diuretics. Diuretics do not have a role in the antepartum treatment of hypertensive disorders of pregnancy.	Compatible Unlikely to suppress lactation[23]
Flecainide	Maternal and fetal arrhythmias	Limited human data; no teratogenic effects in rats or mice; delayed sternebral and vertebral ossification at high doses in rats	Compatible
Heparin	Anticoagulation	Maternal bleeding	Compatible
Hydralazine	Hypertension	No fetal effects unless precipitous blood pressure fall	Compatible
Ivabradine	Heart failure	Limited human data. IUGR, neonatal bradycardia and hypotension, hypoglycaemia	Excretion in breast milk in animal studies. No human data. Not recommended, consider alternatives.
Lignocaine	Local anesthesia, maternal arrhythmias	Not usually associated with adverse fetal effects, High serum levels may cause central nervous depression at birth, fetal bradycardia, acidosis	Compatible
Methyldopa	First line anti-hypertensive	No evidence of adverse fetal or neonatal effects	Compatible
Nifedipine	First line anti-hypertensive in the second and third trimesters, tocolysis	Fetal distress related to precipitous maternal hypotension reported	Compatible
Nitrates	Myocardial infarction and ischemia, hypertension, pulmonary edema	Limited data; use is generally safe, few cases of fetal heart rate deceleration and bradycardia have been reported	Limited data
Procainamide	Maternal and fetal arrhythmias	Limited data; no fetal side effects reported	Limited data, Low levels in breastmilk.[22]
Rivaroxaban	Direct oral anticoagulant	Not recommended. Limited human data	Limited data indicates that 30 mg daily produces low levels in milk. Consider alternatives if possible.
Sodium nitroprusside	Hypertension, aortic dissection	Limited data; potential thiocyanate fetal toxicity, fetal mortality reported in animals	Limited data. Caution advised if mother has received sodium nitroprusside for more than 24 hours.[22]

Medication	Use in pregnancy	Potential fetal / neonatal effects	Use in breastfeeding
Sotalol	Maternal arrhythmias, hypertension, fetal tachycardia	Limited data; cases of fetal death and cases of neurological morbidity in newborns reported, as well as bradycardia in newborns. Monitor maternal QT length	Compatible Monitor infant for drowsiness, poor feeding, adequate weight gain, bradycardia[22]
Verapamil	Maternal and fetal arrhythmias, hypertension, tocolysis	Well tolerated, no teratogenicity in animal studies	Compatible
Warfarin	Anticoagulation	Crosses placental barrier; fetal hemorrhage in utero, embryopathy, central nervous system abnormalities	Compatible

Appendix 2: Maternal mortality CEMACH (UK) 2015-2017[18(p24)]

Type and cause of death	Number	Percentage
Aortic Dissection	9	11
Essential Hypertension	1	1
Ischaemic Heart disease	20	24
Sudden Arrhythmic Cardiac Death with morphologically normal heart (SADS/MNH)	15	18
Myocardial Disease / Cardiomyopathy	22	27
Peripartum Cardiomyopathy	3	*Included in figures above*
Myocarditis	2	*Included in figures above*
Valvular heart disease	3	4
Infectious endocarditis	3	4
Pulmonary arterial hypertension	4	5
Undetermined cardiovascular disease	5	6
Congenital heart disease	4	*Included in figures above*
Total	**82**	**100**

Acknowledgements

The South Australian Perinatal Practice Guidelines gratefully acknowledge the contribution of clinicians and other stakeholders who participated throughout the guideline development process particularly:

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Endorsed by: Commissioning and Performance, SA Health
Next review due: 16/09/2026
ISBN number: 978-1-76083-439-5
CGSQ reference: **PPG003**
Policy history: Is this a new policy (V1)? **N**
Does this policy amend or update and existing policy? **Y**
If so, which version? **V4.1**
Does this policy replace another policy with a different title? **N**
If so, which policy (title)?

Approval Date	Version	Who approved New/Revised Version	Reason for Change
16/09/2021	V5	Clinical Governance, Safety & Quality Domain Custodian	Formally reviewed
5/07/2018	V4.1	SA Health Safety and Quality Strategic Governance Committee	Review date extended to 5 years following risk assessment. New template.
19/12/2014	V4	SA Health Safety and Quality Strategic Governance Committee	Formally reviewed in line with 3 year scheduled timeline for review.
20/11/2012	V3	SA Maternal and Neonatal Clinical Network	Formally reviewed in line with 3 year scheduled timeline for review.
20/04/2010	V2	SA Maternal and Neonatal Clinical Network	Minor amendment for SA Health process
6/04/2010	V1	SA Maternal and Neonatal Clinical Network	Original SA Maternal and Neonatal Clinical Network approved version.