Rheumatic heart disease and pregnancy

IMPORTANT: *Consider individual clinical circumstances. Read the full disclaimer at www.health.qld.gov.au/qcg*

Background

Aspect	Consideration
Aetiology	• Rheumatic heart disease (RHD) is caused by damage to the heart resulting from previous acute rheumatic fever (ARF)[1-3] • ARF is caused by *Group A streptococcal* infection (throat and/or skin)[3,4] • ARF is associated with poor living conditions, overcrowding and socioeconomic deprivation[3,4]
High risk populations	• Maintain awareness that high risk groups for ARF/RHD include women who[6]: o Live in an ARF endemic setting o Are Aboriginal and/or Torres Strait Islander, Maori and Pacific Islander peoples o Are immigrants, or children of immigrants from low/middle income countries and countries with continuing high ARF and RHD prevalence o Live in crowded households and/or are of lower socioeconomic status o Are refugees or have spent time in refugee camps o Have a personal history of ARF and are aged less than 40 years
Signs and symptoms of RHD (known or unknown cases)	• Maintain a high index of suspicion about women in high risk populations who present with[5]: o Breathlessness, cough, wheeze or worsening fatigue o Orthopnoea o Significant reduction in exercise tolerance o Syncope or presyncope o Tachycardia o Leg oedema o Undiagnosed cardiac murmur
Classification	• ARF is classified as *definite, probable* or *possible* using Revised Jones Criteria[5] o There is no specific diagnostic laboratory test o Diagnosis relies on clinical recognition of major and minor clinical manifestations • RHD is classified as *borderline* or *definite*[5] o Diagnosis is based on echocardiographic features o Lesion severity is classified as *mild* or *none, moderate* or *severe*
RHD Register	• ARF and RHD are notifiable diseases in Queensland[6] • The Queensland RHD Register and Control Program is a statewide patient register and recall system for ARF and RHD • If known or new RHD, contact the Queensland RHD register (1300 135 854) and update with pregnancy status

Preconception

Aspect	Consideration
Assessment	• Preconception care is key to optimising pregnancy and perinatal outcomes[7,8] • Identify at risk groups to target information sharing and assessment o Historical diagnosis of ARF and/or RHD may have been missed • If known RHD, refer to cardiology and obstetric medicine services for individualised counselling about: o Risks of ARF/RHD and pregnancy o Management of medications and anticoagulation where indicated o Contraceptive options available until full counselling completed
Pregnancy not recommended	• Pregnancy is not recommended if[7]: o Significant pulmonary arterial hypertension o Severe symptomatic mitral stenosis (mitral valve area less than 1.0 cm^2) o Severe symptomatic aortic stenosis (aortic valve area less than 1.0 cm^2) o Severe left ventricular impairment (ejection fraction less than 30%) o New York Heart Association (NYHA) class III/IV • If pregnancy not recommended, facilitate: o Multidisciplinary team review including maternal fetal medicine specialist o Psychological and emotional support

Clinical standards

Aspect	Consideration
Care pathways	• Establish and promote local antenatal pathways for accessing: o Echocardiography o Face to face and virtual/telehealth specialist care (including cardiology, maternal fetal medicine and obstetric medicine) • Structure care so it is woman centred, culturally safe, multidisciplinary, coordinated and community based wherever possible • Promote continuity of care models and nurse navigator services where available • Coordinate multiple appointments where possible to reduce travel burden and time away from community and family
Model of care	• Individualise care and consider[5]: o Severity of RHD o Co-morbidities o Preference for birthing location o Engagement with health services and adherence with recommended treatments o Access to services o Social, cultural and financial risk factors
Culturally safe care	• Promote local community based care that incorporates and respects cultural values (e.g. Birthing on Country or Birthing in Our Community)[5] • If woman identifies as Aboriginal and/or Torres Strait Islander, offer support from advanced health care workers and multicultural nurse navigators at entry point to service where available • Refer to: o Queensland Health: *Aboriginal and Torres Strait Islander Patient care guideline*[9] o Queensland Health: *Making tracks towards closing the gap in health outcomes for Indigenous Queenslanders by 2033: Policy and accountability framework*[10] o Queensland Clinical Guidelines: *Standard care*[11] o Queensland Health: *Making tracks together: Queensland's Aboriginal and Torres Strait Islander health equity framework*[12] o Migrant and Refugee Women's Health Partnership: *Culturally responsive clinical practice: Working with people from migrant and refugee backgrounds*[13]

Risk assessment and stratification

Aspect	Consideration
Context	• RHD in pregnancy is associated with higher rates of perinatal morbidity and mortality[14] • Early diagnosis and/or management offers best opportunity for optimal outcomes[14]
Significant cardiac risk factors	• Prior cardiovascular event or symptoms before pregnancy (e.g. congestive cardiac failure, systemic embolism) • Dyspnoea with minimal exertion or at rest (NYHA class III/IV) o Requires immediate evaluation by cardiology/physician and/or intensive care team • Atrial arrhythmias • Left ventricular dysfunction • Severe mitral regurgitation (MR) or aortic regurgitation (AR) • Moderate or severe pulmonary hypertension[8] • Multiple or stenotic valvular lesions • Mechanical prosthetic heart valve
Risk stratification	• Refer to Table: Risk categories and care pathways • If late booking, or limited or no antenatal care, expedite cardiac review to enable risk assessment and stratification[15] • Maternal risk is based on a combination of history, current symptoms and echocardiography o Multiple risk classification schema have been developed and these have been modified and adapted by various bodies and organisations[7,16-18] • Assessment of risk level requires expert clinical judgement in a multidisciplinary context • Determine level of risk in consultation with multidisciplinary team • If continuation of pregnancy not recommended due to severity of RHD, termination of pregnancy may be offered o Refer to Queensland Clinical Guideline: *Termination of pregnancy*[19]

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Antenatal care

Aspect	Consideration
Unknown RHD	• First presentation of RHD may be during pregnancy • Symptoms of RHD/ARF may be difficult to distinguish from normal pregnancy changes • Take history for prior cardiac events (e.g. heart failure, arrythmias)[5] • Perform cardiovascular examination • For high risk populations without known RHD, explore history for RHD (e.g. penicillin injections, previous echocardiograms) • Maintain low threshold for echocardiography and cardiac referral in at-risk populations[5,15]
Known RHD	• All routine antenatal assessments are indicated • Perform cardiovascular examination including baseline electrocardiogram (ECG) and echocardiogram • If secondary prophylaxis (antibiotics for prevention of recurrent ARF) is currently prescribed, continue during pregnancy[5] • Monitor for pre-eclampsia[20] o Can exacerbate valvular heart disease and is associated with increased risk of heart failure • Strongly recommend: o Oral health assessment to decrease risk of infective endocarditis[5] o Early anaesthetic review • Review and modify medications as required • Surveillance for infection and ARF recurrence o Decreased immune function increases susceptibility o Wide variation in presentations in pregnancy • Monitoring of fetal growth and wellbeing in the setting of a maternal cardiac condition
Mechanical and/or prosthetic heart valves	• Bioprosthetic valves are usually well tolerated in pregnancy[21] • Women with a mechanical heart valve (MHV) are at a high bleeding and embolic risk, and of poor maternal and perinatal outcomes compared to women with a bioprosthetic heart valve[18,22]
Anticoagulation	• Is required for all women with MHVs o Management is complex; liaise with or refer to an expert practitioner for management options • May be required for atrial fibrillation or venous thromboembolism (VTE) prophylaxis
Cardiac review and place of birth	• If known RHD, arrange cardiology review as early as possible in pregnancy • Refer to Table: Risk categories and care pathways for guidance on frequency of cardiac review and recommended place of birth • Individualise birth plan (including place of birth) according to level of risk and in consultation with multidisciplinary team
Planning birth	• Vaginal birth preferred unless specific indications for caesarean birth[5] • Caesarean section is indicated if: o Unplanned labour and warfarinised[5,7] o Severe heart failure, haemodynamic instability or pulmonary hypertension • Induction of labour may be indicated: o To allow optimisation of anticoagulation in fully anticoagulated women o To facilitate access to specialist medical staff at the birth o If deteriorating maternal cardiac function o Refer to Queensland Clinical Guideline: *Induction of labour*[23]

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Intrapartum care

Aspect	Consideration
Context	• Peripartum is potentially the highest risk period as[5]: o Cardiac output increases as heart rate and blood pressure rise during labour o Inability to increase cardiac output secondary to moderate/severe RHD may lead to pulmonary oedema o Auto transfusion occurs from the release of caval compression and sustained uterine contraction immediately postpartum
General principles	• Individualise intensive and/or invasive monitoring according to severity of disease • Neuraxial blockade may be beneficial • Maintain clinical surveillance for signs of cardiovascular deterioration (e.g. tachycardia, dyspnoea, fatigue, oedema, cyanosis) • Monitor fluid balance because of increased risk of pulmonary oedema o Avoid excessive intravenous fluids • Recommend continuous electronic fetal monitoring in labour • Continuous oxygen saturation monitoring may be indicated • Paediatric or neonatal teams for birth according to individual clinical circumstances
Antibiotics	• Administer antibiotic prophylaxis according to obstetric indications, and local protocols and regimens (e.g. instrumental birth)[5,7,24] • Insufficient evidence to recommend additional antibiotic prophylaxis for endocarditis for women with RHD, including those with prosthetic valves[5,7,25]
Second stage	• As indicated by condition, consider: o Shortening active phase of second stage (e.g. episiotomy, instrumental vaginal birth) o Limiting active maternal pushing to reduce additional load on the cardiovascular system • Pushing in left lateral, rather than supine, lessens cardiovascular changes and improves preload • Avoid prolonged lithotomy positioning
Third stage	• Limited high quality evidence to guide management in women with cardiac disease[26] • Balance management strategies against maternal risk and life threatening bleeding[5] • Recommend modified active management of third stage and prophylactic uterotonics: o Oxytocin 10 units intramuscularly (IM) immediately following birth o An oxytocin infusion can be given prophylactically or for treatment of postpartum haemorrhage ▪ To minimise cardiovascular disturbance, preferentially administer oxytocin slowly by infusion in small volumes of diluent (50–250 mL) in accordance with local protocol ▪ Avoid administering bolus intravenous doses of oxytocin[27] • Avoid ergometrine and carboprost where possible[5] • Misoprostol and tranexamic acid are not contraindicated[28]
Moderate risk	• Labour and birth o Notify anaesthetist at onset of labour (especially if anticoagulated) o Establish intravenous (IV) access o Close surveillance of haemodynamic status and fluid balance o If required, administer vasoactive medication o Recommend early neuraxial blockade to help minimise tachycardia[5], limit pain and prevent hypertensive responses that may lead to heart failure o If obstetric considerations allow, assisted vaginal birth (forceps or vacuum) preferred to caesarean section • Post birth o IV fluids to replace vaginal blood loss only o Monitor in birthing suite, intensive care (ICU) or high dependency unit (HDU) for 12 hours or overnight o Monitor pulse, blood pressure, oxygen saturations, blood loss and dyspnoea
High risk	• Labour and birth as for moderate risk and: o Consider invasive monitoring (arterial line) for labour and birth, with critical care nursing support if managed in birthing suite o Consider birth in operating room suite • Regular multidisciplinary clinical assessment • Post birth as for moderate risk and: o Monitor in HDU/ICU for at least 12 hours (risk of pulmonary oedema) o Strict fluid balance chart and indwelling catheter

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Postpartum

Aspect	Consideration
Clinical surveillance	• Routine postnatal observations and care according to clinical condition • Signs and symptoms of RHD may appear for the first time postpartum • If new onset postpartum dyspnoea or cough, investigate promptly o Signs and symptoms of underlying cardiovascular disease may occur up to five months postpartum[29]
Breastfeeding	• Refer to Queensland Clinical Guideline: *Establishing breastfeeding*[30] • Review safety of cardiac medications during lactation
Anticoagulation	• Management of anticoagulation for women with MHVs is complex[31] o Liaise with or refer to an expert practitioner for management options • For other anticoagulation indications, discuss anticoagulation recommencement with multidisciplinary team • Review safety of anticoagulant medications during lactation

Discharge planning

Aspect	Consideration
Discharge planning	• Confirm next dose of secondary prophylaxis treatment[15] (if applicable) o Facilitate access to secondary prophylaxis if away from local community (e.g. if baby is in neonatal intensive care) • Promote health literacy with discharge plan, consider interpreter as required • Take into consideration if woman has had RHD, other children will often have an increased risk for ARF/RHD[15] • Referral to multicultural, rural or RHD nurse navigators where available
Conception counselling	• Counsel about the importance of contraception and the planning of future pregnancies o Recommended interval between pregnancies is 24 months[32,33] • For girls and women with high risk cardiovascular disease and risk of unplanned pregnancy[34]: o Recommend long acting reversible contraceptives o Intra-uterine contraceptive devices (IUCD) including levonorgestrel (Mirena®) and copper are safe o Etonogestrel implant (e.g. Implanon®) o Avoid oestrogen containing contraceptives as associated with elevated risk of thrombosis[5] o Barrier and 'natural' methods of contraception have unacceptable failure rates o If further children not desired, explore tubal ligation
Follow up	• Follow-up cardiac review according to priority o Arrange prior to discharge wherever possible o High risk: referral to main cardiac clinic for management and possible cardiothoracic surgery • Recommend o Designated GP or primary care provider for home or centre based therapy and education following discharge o Sharing information on treatment, medications, future management plans and conception planning with primary care providers/referring hospital within 48 hours of discharge[15]

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Risk categories and care pathways

If in doubt or borderline, refer to category with higher risk. Individualise risk stratification and actions according to clinical circumstances and consultation with experts.

Level	Risk category	General[5]	Left ventricular impairment[5,7]	Valves[5,7,18]	Mitral stenosis[5]	Aortic stenosis[5]	Suggested actions[5,35]
IV	Extreme risk	Pulmonary arterial hypertension or NYHA class III/IV	Severe (EF < 30%)		Severe symptomatic (MVA < 1.0 cm^2)	Severe symptomatic (AVA < 1.0 cm^2)	• Preconception: advise against pregnancy • If pregnant, continuation of may not be recommended • Discussion about ToP may be required
	Very high	Severe symptomatic mitral regurgitation or aortic regurgitation		MV or AV disease with pulmonary hypertension	Severe (MVA < 1.5 cm^2)		• Urgent cardiac review • Cardiac review then monthly or more frequently as required • Birth at CSCF level 5 or 6
III	High	Severe asymptomatic mitral regurgitation or aortic regurgitation	Moderate (EF 30–45%)[5]	Mechanical heart valve	Moderate (MVA 1.5–2.0 cm^2)	Severe asymptomatic (AVA < 1.0 cm^2)	• Cardiac review ASAP • Cardiac review then 2nd monthly • Birth at CSCF level 5 or 6
II	Moderate	Mild/suspected asymptomatic RHD *and* • < 20 weeks at booking • Moderate mitral regurgitation or aortic regurgitation • Not mWHO III or IV	Mild (EF > 45%) without severe regurgitation or stenosis and good functional capacity	Bioprosthetic valves or Previous PBMV	Mild (MVA > 2.0 cm^2)	Moderate (AVA 1.0–1.5 cm^2)	• Cardiac review at next opportunity • Cardiac review then each trimester (minimum) • *Birth at CSCF level ≥ 4
I	Low	• Mild or suspected RHD *and* o ≥ 20 weeks at booking *and* o BMI < 35 kg/m^2 *and* o No significant co-morbidities • History of ARF with no carditis, or mild MR or AR • History of atrial arrhythmias in absence of significant valvular disease					• Cardiac review including echocardiography and ECG • Cardiac review then 1–2 times (minimum) • Birth at CSCF level ≥ 3

*Consider cultural significance for Aboriginal and Torres Strait Island women and families of birthing on homelands. CSCF level 3 may be appropriate in some circumstances in consultation with local experts, obstetric and cardiac specialists, and individualised risk assessment for each woman.

Abbreviations: > greater than; **<** less than; **≤** less than or equal to; **AR** aortic regurgitation; **ARF** acute rheumatic fever; **ASAP:** as soon as possible, **AV** aortic valve **AVA** aortic valve area; **CSCF** clinical services capability framework; **ECG** electrocardiogram; **EF** ejection fraction; **MR** mitral regurgitation, **MV** mitral valve, **MVA** mitral valve area; **mWHO** modified World Health Organization; **NYHA** New York Heart Association; **PBMV** percutaneous balloon mitral valvuloplasty; RHD rheumatic heart disease, **ToP** termination of pregnancy.

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