Rheumatic heart disease and pregnancy

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

Background

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
</table>
| **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*\(^6\)  
  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

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
</table>
| **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

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Care pathways</strong></td>
<td>• Establish and promote local antenatal pathways for accessing:&lt;br&gt;  ○ Echocardiography&lt;br&gt;  ○ Face to face and virtual/telehealth specialist care (including cardiology, maternal fetal medicine and obstetric medicine)&lt;br&gt;  ○ Structure care so it is woman centred, culturally safe, multidisciplinary, coordinated and community based wherever possible&lt;br&gt;  ○ Promote continuity of care models and nurse navigator services where available&lt;br&gt;  ○ Coordinate multiple appointments where possible to reduce travel burden and time away from community and family</td>
</tr>
<tr>
<td><strong>Model of care</strong></td>
<td>• Individualise care and consider:&lt;br&gt;  ○ Severity of RHD&lt;br&gt;  ○ Co-morbidities&lt;br&gt;  ○ Preference for birthing location&lt;br&gt;  ○ Engagement with health services and adherence with recommended treatments&lt;br&gt;  ○ Access to services&lt;br&gt;  ○ Social, cultural and financial risk factors</td>
</tr>
<tr>
<td><strong>Culturally safe care</strong></td>
<td>• Promote local community based care that incorporates and respects cultural values (e.g. Birthing on Country or Birthing in Our Community)&lt;br&gt;  ○ 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&lt;br&gt;  ○ Refer to:&lt;br&gt;    ○ Queensland Health: <em>Aboriginal and Torres Strait Islander Patient care guideline</em>&lt;br&gt;    ○ Queensland Health: <em>Making tracks towards closing the gap in health outcomes for Indigenous Queenslanders by 2033: Policy and accountability framework</em>&lt;br&gt;    ○ Queensland Clinical Guidelines: <em>Standard care</em>&lt;br&gt;    ○ Queensland Health: <em>Making tracks together: Queensland’s Aboriginal and Torres Strait Islander health equity framework</em>&lt;br&gt;    ○ Migrant and Refugee Women’s Health Partnership: <em>Culturally responsive clinical practice: Working with people from migrant and refugee backgrounds</em></td>
</tr>
</tbody>
</table>

### Risk assessment and stratification

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

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
</table>
| **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, arrhythmias)  
• 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  |
| **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  
• Monitor for pre-eclampsia  
  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  
  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  
• 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  |
| **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  
• Caesarean section is indicated if:  
  o Unplanned labour and warfarinised  
  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*  |

© State of Queensland (Queensland Health) 2022
## Intrapartum care

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
</table>
| **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 |
### Postpartum

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
</table>
| **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  
• Signs and symptoms of underlying cardiovascular disease may occur up to five months postpartum |
| **Breastfeeding**  | • Refer to Queensland Clinical Guideline: *Establishing breastfeeding*  
• Review safety of cardiac medications during lactation |
| **Anticoagulation** | • Management of anticoagulation for women with MHVs is complex  
• 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

<table>
<thead>
<tr>
<th>Aspect</th>
<th>Consideration</th>
</tr>
</thead>
</table>
| **Discharge planning** | • Confirm next dose of secondary prophylaxis treatment (if applicable)  
• 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  
• Referral to multicultural, rural or RHD nurse navigators where available |
| **Conception counselling** | • Counsel about the importance of contraception and the planning of future pregnancies  
• Recommended interval between pregnancies is 24 months  
• For girls and women with high risk cardiovascular disease and risk of unplanned pregnancy:  
• Recommend long acting reversible contraceptives  
• Intra-uterine contraceptive devices (IUCD) including levonorgestrel (Mirena®) and copper are safe  
• Etonogestrel implant (e.g. Implanon®)  
• Avoid oestrogen containing contraceptives as associated with elevated risk of thrombosis  
• Barrier and ‘natural’ methods of contraception have unacceptable failure rates  
• If further children not desired, explore tubal ligation |
| **Follow up**          | • Follow-up cardiac review according to priority  
• Arrange prior to discharge wherever possible  
• High risk: referral to main cardiac clinic for management and possible cardiothoracic surgery  
• Recommend  
• Designated GP or primary care provider for home or centre based therapy and education following discharge  
• Sharing information on treatment, medications, future management plans and conception planning with primary care providers/referring hospital within 48 hours of discharge |
### 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.*

<table>
<thead>
<tr>
<th>Level</th>
<th>Risk category</th>
<th>General&lt;sup&gt;5&lt;/sup&gt;</th>
<th>Left ventricular impairment&lt;sup&gt;5,7&lt;/sup&gt;</th>
<th>Valves&lt;sup&gt;5,7,18&lt;/sup&gt;</th>
<th>Mitral stenosis&lt;sup&gt;5&lt;/sup&gt;</th>
<th>Aortic stenosis&lt;sup&gt;5&lt;/sup&gt;</th>
<th>Suggested actions&lt;sup&gt;5,35&lt;/sup&gt;</th>
</tr>
</thead>
</table>
| IV    | Extreme risk  | Pulmonary arterial hypertension or NYHA class III/IV | Severe (EF < 30%) | MV or AV disease with pulmonary hypertension | Severe symptomatic (MVA < 1.0 cm<sup>2</sup>) | Severe symptomatic (AVA < 1.0 cm<sup>2</sup>) | • Preconception: advise against pregnancy  
• If pregnant, continuation of may not be recommended  
• Discussion about ToP may be required |
| III   | Very high     | Severe symptomatic mitral regurgitation or aortic regurgitation | Moderate (EF 30–45%)<sup>3</sup> | Mechanical heart valve | Moderate (MVA 1.5–2.0 cm<sup>2</sup>) | Severe asymptomatic (AVA < 1.0 cm<sup>2</sup>) | • Urgent cardiac review  
• Cardiac review then monthly or more frequently as required  
• Birth at CSCF level 5 or 6 |
| II    | High          | Severe asymptomatic mitral regurgitation or aortic regurgitation | Bioprosthetic valves or Previous PBMV | Mild (MVA > 2.0 cm<sup>2</sup>) | Mild (AVA 1.0–1.5 cm<sup>2</sup>) | Moderate (AVA 1.0–1.5 cm<sup>2</sup>) | • Cardiac review ASAP  
• Cardiac review then 2<sup>nd</sup> monthly  
• Birth at CSCF level 5 or 6 |
| I     | 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<sup>2</sup>) | Moderate (AVA 1.0–1.5 cm<sup>2</sup>) | • Cardiac review at next opportunity  
• Cardiac review then each trimester (minimum)  
• *Birth at CSCF level ≥ 4 |
|       | Low           | Mild or suspected RHD and  
• ≥ 20 weeks at booking  
• BMI < 35 kg/m<sup>2</sup> and  
• 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.*
References


Rheumatic heart disease and pregnancy

© State of Queensland (Queensland Health) 2022
Page 7 of 7