



6. Rheumatic heart disease in pregnancy

Ideally, patients with known rheumatic valvular disease should be properly assessed before pregnancy. Discussion regarding fertility planning should be undertaken with all women with more than mild valvular disease, even if immediate pregnancy is not planned.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). Recurrences of ARF may cause further cardiac valve damage. Thus, RHD steadily worsens in people who have multiple episodes of ARF.

What is RHD?

RHD is damage to the heart that remains after the acute ARF episode has resolved. It is caused by an episode or recurrent episodes of ARF, where the heart has become inflamed; the heart valves remain stretched and/or scarred, and normal blood flow is interrupted. Recurrences of ARF may cause further valve damage, leading to worsening of RHD. Preventing recurrences of ARF by using prophylactic treatment with penicillin is therefore of great importance in controlling RHD.

Who gets RHD?

In Australia, the vast majority of people with RHD are Aboriginal people and Torres Strait Islanders, many of whom live in remote areas of central and northern Australia. Pacific Islanders, and migrants from high-prevalence countries, are also at high risk.

Pregnancy in women with RHD

Normal pregnancy is associated with a 30–50% increase in blood volume, reduction in systemic vascular resistance and corresponding increase in cardiac output. These changes begin during the first trimester, peaking at 28–30 weeks of pregnancy, and are then sustained until term. The increase in blood volume is associated with an increase in heart rate by 10–15 beats per min. Because of the hyperdynamic circulation, innocent, soft mid-systolic murmurs are common during pregnancy, particularly along the left sternal border. These circulatory changes of pregnancy will exacerbate any pre-existing valvular disease. Sometimes RHD, especially mitral stenosis, is first diagnosed during pregnancy or soon after delivery when a woman develops symptoms, usually dyspnoea.

Assessment of women with RHD

Ideally, patients with known rheumatic valvular disease should be properly assessed before pregnancy. Discussion regarding fertility planning should be undertaken with all women with more than mild valvular disease, even if immediate pregnancy is not planned. Assessment should include a full history and examination, and an echocardiogram. If patients are already symptomatic, due to significant rheumatic valvular disease, serious consideration should be given to interventional therapy or surgery prior to pregnancy to avoid life-threatening complications, which may occur in these patients. In these patients, the use of contraception with a low failure rate (etonogestrel implant; Implanon, Organon International, Oss, the Netherlands) should be strongly encouraged if there is a risk of pregnancy, while more definitive treatment is being undertaken.

Risk factors

The predictors of increased maternal and fetal risk in the pregnant patient with rheumatic valvular disease are:

- reduced left ventricle (LV) systolic function
- significant aortic or mitral stenosis
- moderate or severe pulmonary hypertension
- a history of heart failure
- symptomatic valvular disease before pregnancy
- atrial fibrillation, especially when anticoagulation is required
- pregnant women with mechanical valves are a very high-risk group, in whom all anticoagulation options carry maternal and/or fetal risks.

Management of women with RHD

Women with more than mild RHD should be identified as having a higher than normal risk of complications in pregnancy, and should receive antenatal care at an appropriate referral centre with an experienced obstetrician, in collaboration with an obstetric physician and/or cardiologist. The most severe cases should be seen at a referral centre with cardiology and intensive care facilities. Discussions with the woman regarding timing, nature and site of planned delivery should occur before or early in pregnancy.

During pregnancy, women with valvular heart disease should be reviewed regularly by a cardiac specialist, and cardiac status should be reviewed whenever there is a change in symptoms. A multidisciplinary approach to management is an important principle for care of the pregnant patient with rheumatic valvular disease. It is often necessary to advise women with heart disease to cease work earlier in pregnancy for medical reasons, as cardiac demands increase significantly as pregnancy proceeds. Most women with valvular heart disease become more symptomatic in the third trimester.

Key points in the management of pregnancy in women with RHD

Predictors of increased maternal and fetal risk	<p>Decreased LV systolic function</p> <p>Significant aortic and mitral stenosis</p> <p>Moderate or severe pulmonary hypertension</p> <p>Heart failure</p> <p>Symptoms before pregnancy</p> <p>Mechanical valve prostheses</p> <p>Atrial fibrillation requiring warfarin</p>
Cardiac assessment	<p>Early comprehensive assessment with echocardiography to assess valves and LV function</p> <p>Plan multidisciplinary management</p>
Mitral/aortic regurgitation	<p>Usually well tolerated</p> <p>Treat medically with diuretics, vasodilators (no ACE inhibitors/angiotensin II receptor blockers) for clinical heart failure</p>
Mitral stenosis	<p>Mild to moderate mitral stenosis: manage medically moderate to severe mitral stenosis (MVA $<1.5 \text{ cm}^2$)—consider PBMV during late second trimester, if patient remains symptomatic and PAS pressure $>50 \text{ mmHg}$</p> <p>Beta-blockers or digoxin for rate control of atrial fibrillation</p>
Aortic stenosis (rare)	<p>Mild to moderate aortic stenosis: well-tolerated. Diuretics for heart failure</p> <p>Consider PTAV if severe symptoms</p> <p>Beta-blockers or digoxin for rate control of atrial fibrillation. Avoid cardiac surgery, as high risk of fetal loss</p>
Mechanical/prosthetic valves and anticoagulation in pregnancy	<p>High maternal and fetal risk</p> <p>Risk of warfarin embryopathy in first trimester</p> <p>Embryopathy may be avoided if warfarin dose $\leq 5 \text{ mg}$</p>
Choice of 3 antithrombotic regimens	<ol style="list-style-type: none"> 1. LMWH throughout pregnancy, weight-adjusted dose with anti-Xa level monitoring 2. Warfarin throughout pregnancy, if can keep warfarin $\leq 5 \text{ mg}$, e.g. INR 2–3 in aortic prosthesis, sinus rhythm; change to LMWH or unfractionated heparin at 36 weeks 3. LMWH until 13 weeks, and then warfarin and aspirin until 36 weeks; change to LMWH or UFH until labour. Monitor anti-Xa levels with LMWH
Labour	<p>Haemodynamic monitoring: non-invasive, if mild to moderate valve disease</p> <p>Antibiotic prophylaxis, if prolonged labour and/or ruptured membranes</p> <p>Aim for short second stage and multidisciplinary management approach, with low threshold for obstetric intervention</p>

ACE, angiotensin-converting enzyme; anti-Xa, antifactor Xa; INR, international normalised ratio; LMWH, low-molecular weight heparin; LV, left ventricle; MVA, mitral valve area; PAS, pulmonary artery systolic; PBMV, percutaneous balloon mitral valvuloplasty; PTAV, percutaneous transluminal aortic valvuloplasty; UFH, unfractionated heparin.



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The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)

Quick reference guides include:

- Primary prevention of ARF
- Diagnosis of ARF
- Management of ARF
- Secondary prevention of ARF
- Management of RHD
- RHD in pregnancy
- RHD control programs

RHDAustralia

Ph: 08 8922 8196

Email: info@rhdaustralia.org.au

www.rhdaustralia.org.au

