MANAGEMENT OF RHEUMATIC HEART DISEASE

Quick Reference Guide for Health Professionals

This quick reference guide is derived from 'National Heart Foundation of Australia (NHFA) and the Cardiac Society of Australia and New Zealand (CSANZ) Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia — an evidence-based review. 2006'

What is rheumatic heart disease?

Rheumatic heart disease (RHD) results from damage to the heart valves caused by an episode or recurrent episodes of acute rheumatic fever (ARF). ARF is an auto-immune inflammatory response to bacterial infection with group A streptococcus (GAS).

Recurrences of ARF may cause further valve damage, leading to steady worsening of RHD. Secondary prevention of recurrent ARF — through prophylactic treatment with penicillin — is therefore of great importance in controlling RHD.

Who gets rheumatic heart disease?

In Australia, the vast majority of people with chronic RHD are Aboriginal and Torres Strait Islander people.

It is difficult and expensive for Aboriginal and Torres Strait Islander people to travel to major centres for cardiac services, which are often hospital based. Although specialist outreach services are improving in many regions, access to specialist care is suboptimal in rural and remote areas.

Best practice in RHD management

The implementation of guidelines for chronic RHD has major implications for Aboriginal and Torres Strait Islander health care services, especially in rural and remote regions. In addition to access to appropriate primary care services, best practice for RHD requires:

- access to a specialist physician and/or cardiologist (preferably the same specialist over a long time);
- access to echocardiography;
- adequate monitoring of anticoagulation therapy in patients with atrial fibrillation and/or mechanical prosthetic valves; and
- secondary prevention with benzathine penicillin G (BPG) prophylaxis.

The fundamental goal in long-term management of chronic RHD is to avoid, or at least delay, valve surgery. Therefore, prophylaxis with BPG to prevent recurrent ARF is a crucial strategy in managing patients with chronic RHD. Where adherence to secondary prevention is poor, there is greater need for surgical intervention and long-term surgical outcomes are not as good.

VALVULAR LESIONS IN RHD

Specific valvular lesions in chronic RHD include:

- *mitral regurgitation*, in which volume overload of the left ventricle and left atrium occurs — in the more severe cases this may result in a progressive decline in systolic contractile function;
- *mitral stenosis*, where progressive obstruction to left ventricular inflow develops, due to fibrosis and partial fusion of the mitral valve leaflets;
- *aortic regurgitation*, where left ventricular volume overload occurs and there is an increase in left ventricular end diastolic volume, eventually leading to left ventricular contractile dysfunction in the more severe cases; and
- *aortic stenosis*, which results from fibrosis and fusion of the valve cusps, causing progressive obstruction to left ventricular outflow.

In patients with multiple valve lesions, management usually focuses on the most severe valve lesion.

KEY POINTS IN MANAGEMENT OF RHEUMATIC MITRAL REGURGITATION

Symptoms	May be asymptomatic for many years Exertional dyspnoea and fatigue
Examination	Pansystolic murmur at left ventricular apex
Echocardiography	Over-riding or prolapse of anterior mitral valve leaflet Thickened "dog leg" anterior mitral valve leaflet, especially if associated mitral stenosis Retrograde colour (mosaic) regurgitant jet into left atrium, often posteriorly directed Severity graded by area of colour regurgitant jet in left atrium Left ventricular chamber dimensions enlarged if moderate or greater mitral regurgitation Assess left ventricular systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	No role for vasodilators (eg nifedipine) Diuretics and ACE inhibitors if heart failure
Indications for surgery	Moderate/severe mitral regurgitation with symptoms NYHA FC II, III, IV Asymptomatic severe mitral regurgitation in children Asymptomatic severe mitral regurgitation in adults when: • reduced LVEF (<60%) • LVESD ≥40mm • pulmonary hypertension (PAS >50mmHg)
Choice of operation	Mitral valve repair — operation of choice Mitral valve replacement only in older patients with very calcified leaflets Avoid mechanical prostheses if concerns about warfarin adherence or future pregnancy

Notes: LVEF=left ventricular ejection fraction; LVESD=left ventricular end systolic diameter (echo); NYHA FC=New York Heart Association Functional Class; MR=mitral regurgitation; PAS=pulmonary artery systolic

KEY POINTS IN MANAGEMENT OF RHEUMATIC MITRAL STENOSIS

Symptoms	May be asymptomatic Exertional dyspnoea, fatigue, palpitations
Examination	Low-pitched mid-diastolic "rumble" at left ventricular apex
Echocardiography	Thickened, restricted "dog leg" anterior mitral valve leaflet Restricted posterior leaflet Measure mean mitral gradient from continuous wave Doppler signal Calculate MVA from slope of Doppler mitral inflow velocity Calculate PAS pressure from peak tricuspid regurgitant jet velocity ($4V^2$)
Cardiac catheterisation	Only to exclude coronary artery disease
Atrial fibrillation	Common Rate control using beta-blockers or digoxin, or consider cardioversion if recent onset Need anticoagulation to prevent thromboembolic complications
Indications for intervention	Symptoms NYHA FC II–IV MVA <1.5cm^2 or PAS >50mmHg No left atrial thrombus Mild or no mitral regurgitation
Procedure of choice	Percutaneous balloon mitral valvuloplasty by high-volume operator/centre 65% free of restenosis after 10 years Mitral valve repair or replacement if valve leaflets heavily calcified

Notes: MVA=mitral valve area; NYHA FC=New York Heart Association Functional Class; PAS=pulmonary artery systolic; V=velocity

KEY POINTS IN MANAGEMENT OF RHEUMATIC AORTIC REGURGITATION

Symptoms	May be asymptomatic for many years Exertional dyspnoea and fatigue
Signs	Diastolic blowing, decrescendo murmur at left sternal border, usually associated with systolic ejection murmur
Echocardiography	Retrograde diastolic regurgitant colour jet in LVOT and left ventricular chamber Area of jet in LVOT correlates with severity Left ventricular chamber dimensions enlarged if moderate or greater aortic regurgitation May have associated mitral valve disease Pan-diastolic reversed diastolic flow in descending thoracic aorta if moderate/severe aortic regurgitation (Doppler) Assess left ventricular systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Medical management	Vasodilator therapy with dihydropyridines (eg nifedipine), especially if systolic hypertension in asymptomatic, moderate or greater aortic regurgitation Diuretics and ACE inhibitors if heart failure
Indications for surgery	Moderate/severe aortic regurgitation with symptoms NYHA FC II–IV Asymptomatic moderate/severe aortic regurgitation if: • LVEF <55% • LVESD ≥55mm • LVEDD >70mm
Choice of surgery	Valve replacement: • bioprosthesis or homograft – no warfarin if in sinus rhythm – limited durability in younger patients • mechanical valve – warfarin required Aortic valve repair: • limited experience Ross procedure: • aortic autograft (pulmonary valve) and pulmonary homograft replacement Ross procedure and aortic valve repair only in selected cases with experienced surgeons

Notes: LVEDD=left ventricular end diastolic diameter; LVEF=left ventricular ejection fraction; LVESD=left ventricular end systolic diameter; NYHA FC=New York Heart Association Functional Class; LVOT=left ventricular outflow tract

KEY POINTS IN MANAGEMENT OF RHEUMATIC AORTIC STENOSIS

Symptoms	May be asymptomatic Exertional dyspnoea, angina, syncope
Signs	Low-pitched systolic ejection murmur in aortic area
Echocardiography	Thickened, restricted aortic valve leaflets Measure peak and mean systolic gradient from Doppler velocity across aortic valve ($4V^2$) Assess left ventricular systolic function
Cardiac catheterisation	Only to exclude coronary artery disease
Indications for surgery	Symptoms plus mean systolic gradient >50mmHg or AVA <1.0cm^2
Choice of surgery	Valve replacement • bioprosthesis or homograft – limited durability – no warfarin if in sinus rhythm • mechanical valve – long-term warfarin required

Notes: AVA=aortic valve area; V=velocity

KEY POINTS IN MANAGEMENT OF PREGNANCY IN PATIENTS WITH CHRONIC RHD

During pregnancy	Blood volume increases 20–100% Will exacerbate any pre-existing rheumatic valvular heart disease
Factors that put pregnancy and mother at increased risk	Decreased left ventricular systolic function Significant aortic and mitral stenosis Pulmonary hypertension Heart failure Symptoms before pregnancy
Cardiac assessment	Early comprehensive assessment with echocardiography to assess valves and left ventricular function Plan multidisciplinary management
Mitral/aortic regurgitation	Usually well tolerated Treat medically with diuretics, vasodilators (no ACE inhibitors/angiotensin II receptor blockers) for heart failure
Mitral stenosis	Mild to moderate mitral stenosis — manage medically Moderate to severe mitral stenosis (MVA <1.5cm^2) — consider percutaneous balloon mitral valvuloplasty during late second trimester if patient remains symptomatic and PAS pressure >50mmHg Beta-blockers, digoxin for rate control of atrial fibrillation
Aortic stenosis (rare)	Mild to moderate — well tolerated Diuretics for heart failure Beta-blockers, digoxin for rate control of atrial fibrillation Severe aortic stenosis (AVA >50mmHg mean gradient): • percutaneous aortic valvuloplasty if severely symptomatic Avoid cardiac surgery as high risk of foetal loss
Mechanical/prosthetic valves and anticoagulation in pregnancy	High maternal and foetal risk Risk of warfarin embryopathy in first trimester Embryopathy may be avoided if warfarin dose ≤5mg
Choice of three anti-thrombotic regimens	1 LMWH throughout pregnancy, weight-adjusted dose with anti-Xa level monitoring 2 Warfarin throughout pregnancy if can keep warfarin ≤5mg (eg INR 2.0–3.0 in aortic prosthesis, sinus rhythm); change to LMWH or UFH 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	Haemodynamic monitoring — non-invasive if mild to moderate valve disease Antibiotic prophylaxis if prolonged labour and/or ruptured membranes Aim for short second stage, multidisciplinary management Approach with low threshold for obstetric intervention

Notes: anti-Xa=anti-factor Xa; AVA=aortic valve area; INR=international normalised ratio; LMWH=low molecular weight heparin; MVA=mitral valve area; PAS=pulmonary artery systolic; UFH=unfractionated heparin

FURTHER INFORMATION

The full evidence-based review from which this quick reference guide is derived provides detailed information on the diagnosis and management of ARF, secondary prevention and RHD control programs, and diagnosis and management of RHD.

Other quick reference guides are:

- *Diagnosis of Acute Rheumatic Fever*
- *Management of Acute Rheumatic Fever*
- *Secondary Prevention of Acute Rheumatic Fever*
- *Rheumatic Heart Disease Control Programs*.

These publications are available from the National Heart Foundation of Australia through:

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