Diagnosis and management of Rheumatic Heart Disease

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Learning Objectives

• Understand the best approaches to prevention, diagnosis and management of RHD
• Identify the aims and function of control programs
• Know where to locate information on best-practice approaches to the prevention, diagnosis and
• Management of RHD
• The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)
Introduction

• Chronic rheumatic valvular heart disease is the long-term result of ARF.
• It is a disease of poverty and disadvantage.
• In Australia, the burden of RHD is confined almost exclusively to Aboriginal and Torres Strait Islanders.
• Most live in remote areas of Western Australia, the Northern Territory, Queensland, South Australia and New South Wales.
• Virtually eliminated among the non-Indigenous population.
Background and management principles

- Major implications for the healthcare services of Aboriginal people and Torres Strait Islanders, especially in rural and remote regions.
- In addition to access to culturally-appropriate primary care services, best practice for RHD requires:
  - secondary prevention with penicillin prophylaxis
  - adequate monitoring of anticoagulation therapy in patients with AF and/or mechanical prosthetic valves
  - access to oral healthcare
  - access to echocardiography
  - access to a specialist physician, paediatrician and/or cardiologist, preferably the same specialist, for regular follow up visits
  - access to cardiothoracic and interventional cardiology services.
Secondary prevention with penicillin prophylaxis

• The fundamental goal is to prevent ARF recurrences and
• To prevent progression, and in many cases, to allow for the resolution of heart disease.
• This can be achieved by the register-based delivery of secondary prophylaxis with long-acting intramuscular penicillin administered every 28 days.
• Carditis following the first episode of ARF is often mild.
• On secondary prophylaxis, the majority with mild disease at diagnosis have no detectable disease within 5–10 years.
Secondary prevention with penicillin prophylaxis

• Those with moderate to severe disease and those who suffer from recurrent attacks of ARF have poorer long-term outcomes,

• They have a greater need for cardiac surgical intervention

• Many with severe heart disease at presentation can avoid cardiac surgery with high level of compliance with secondary prophylaxis
Secondary prevention

• Secondary prevention of rheumatic fever is defined as the continuous administration of specific antibiotics to patients with a previous attack of rheumatic fever, or well-documented rheumatic heart disease.

• The purpose is to prevent colonization or infection of the upper respiratory tract with group A beta-hemolytic streptococci and the development of recurrent attacks of rheumatic fever’. World Health Organization 2001125
Major elements of secondary prevention of ARF/RHD

- **Individual level**
  Accurate and timely diagnosis of ARF
  Appropriate delivery of secondary prophylaxis
  Prevention of infective endocarditis
  Routine review, structured care planning and coordinated multidisciplinary care
  Health education for individuals, families and the community

- **Screening for undiagnosed RHD**

- **Organisational level**
  RHD control programs

- **Societal level**
  Advocacy for improvements in social, economic, environmental and health service determinants of ARF/RHD incidence and adverse outcomes
Secondary prophylaxis

- Penicillin (compared to no therapy) is beneficial in the prevention of recurrent ARF
- Intramuscular BPG is superior to oral penicillin in the reduction of both recurrent ARF (87–96% reduction) and streptococcal pharyngitis (71–91% reduction)
- Secondary prophylaxis also reduces the severity of RHD
- 50-70% regression of heart disease with adequate adherence over a decade
- Reduces mortality
Secondary prophylaxis

• 1,200,000 U of BPG should be used for secondary prophylaxis for all persons weighing 20 kg or more, and 600,000 U for those weighing less than 20 kg
• BPG is most effectively given as a deep intramuscular injection, into the upper outer quadrant of the buttock or the anterolateral thigh.
• While BPG is usually administered every 4 weeks, serum penicillin levels may be low or undetectable 28 days following a dose of 1,200,000 U
• Fewer streptococcal infections and ARF recurrences have been documented among patients receiving three-weekly BPG, compared to four-weekly BPG
• The three-weekly regime resulted in a greater resolution of MR in a long-term, randomised study in Taiwan (66% vs 46%)
• But in the years between 2002 and 2009, no patients in the Northern Territory who received 100% of their four-weekly injections had a recurrent episode of ARF.
3 weekly BPG injections

- The benefits of three-weekly BPG injections are offset by the difficulties of achieving good adherence.
- The use of four-weekly BPG is currently the treatment of choice, except in patients considered to be at ‘high risk’, for whom three-weekly administration is recommended.
- High-risk patient groups include:
  - those with moderate or severe carditis, or a history of valve surgery, who demonstrate good adherence to less frequent injections
  - those who have confirmed breakthrough ARF, despite full adherence to four-weekly BPG
- Prolonging the dosing interval beyond 4 weeks may increase the risk of breakthrough ARF.
- Monthly, rather than four-weekly administration of BPG, is an acceptable alternative.
Alternatives to intramuscular benzathine penicillin G

- Oral penicillin is less efficacious than BPG in preventing GAS infections and subsequent recurrences of ARF.
- Twice-daily oral regimens are also likely to result in poorer rates of adherence over long periods of time and less predictable serum penicillin concentrations, when compared to intramuscular BPG.
- Oral penicillin should be reserved for patients who experience bleeding problems following injection, and for those who refuse intramuscular BPG.
- If a patient is offered oral penicillin, the consequences of missed doses must be emphasised, and adherence carefully monitored.
- Using an alternative injectable penicillin during a shortage in 2007 resulted in an increase in the number of reported cases of ARF in the Northern Territory, almost doubling the previous 3-year average.
- Organisational approaches to secondary prevention should therefore ensure consistent supply at the national, regional and local levels.
Penicillin allergy

• The benefits of long-term BPG administration outweigh the rare risk of serious allergic reactions to penicillin and fatality as a result of anaphylaxis.

• The rates of allergic and anaphylactic reactions to monthly BPG are 3.2% and 0.2%, respectively, and fatal reactions are exceptionally rare.

• There is no increased risk with prolonged BPG use.

• Carefully questioning about known allergies to penicillin and other beta-lactam antibiotics.

• If confirmed a non-beta-lactam antimicrobial (e.g. erythromycin) should be used instead.

• If there is no unequivocal evidence, they should be investigated for a penicillin allergy, preferably in consultation with an allergist.

• The options include skin testing or a supervised challenge test.

• Most of these patients are not truly allergic.
Duration of secondary prophylaxis

• All persons with ARF or RHD
• Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer).
Status after initial period has relapsed

- No RHD
- No pathological mitral or aortic regurgitation, but may have minor morphological changes to mitral or aortic valves on echocardiography
- Discontinue at that time
Mild RHD

- Mild mitral or aortic regurgitation clinically and on echocardiography, with no clinical evidence of heart failure, and no evidence of cardiac chamber
- Discontinue at the that time
Moderate RHD

• Any valve lesion of moderate severity clinically (e.g. mild–moderate cardiomegaly and/or mild–moderate heart failure) or on echocardiography
• Mild mitral regurgitation, together with mild aortic regurgitation clinically or on echocardiography
• Mild or moderate mitral or aortic stenosis
• Any pulmonary or tricuspid valve lesion co-existing with a left-sided valve lesion
• Continue until 35 yrs
Severe RHD

• Any severe valve lesion clinically (e.g. moderate to severe cardiomegaly or heart failure) or on echocardiography
• Any impending or previous cardiac valve surgery for RHD
• 40 yrs or longer
Potential strategies to improve the delivery of secondary prophylaxis

- Evaluate the local health service environment to identify specific barriers to injection delivery.
- Based on the outcome of the evaluation the following strategies may be useful:
  - Identify local, dedicated staff members to deliver secondary prophylaxis and coordinate routine care
  - Focus on improving relationships between health staff and patients/families
  - Support and use the expertise, experience, community knowledge and language skills of Aboriginal health workers
  - Develop and implement recall and reminder systems (based on a local ARF/RHD register where established) to accommodate the high mobility of individuals and groups:
Potential strategies to improve the delivery of secondary prophylaxis

- Ensure that recall systems extend beyond community boundaries
- Establish networks for timely communication between health clinics
- Use a centralised coordinator and register to assist in monitoring movement
- Minimise staff turnover in remote and rural primary healthcare centres and regional hospitals, or minimise the impact of staff turnover where possible
- Promote the importance of secondary prophylaxis in preventing recurrent ARF and the development or worsening of RHD
- Improve quality and delivery of ongoing health education and support for staff, patients and families
- Implement measures to reduce pain of injections where indicated
- Base routine care on standardised, evidence-based guidelines.
Measures that may reduce the pain of BPG injections

• Use a 21-gauge needle
• Warm syringe to room temperature immediately before using
• Allow alcohol from swab to dry before inserting needle
• Apply pressure with thumb for 10 sec before inserting needle
• Deliver injection very slowly (preferably over at least 2–3 min)
• Distract patient during injection
Procedures requiring endocarditis prophylaxis in patients with RHD

• Dental
• dental extractions
• periodontal procedures including surgery, subgingival scaling and root planning
• replanting avulsed teeth
• other surgical procedures (e.g. implant placement, apioectomy)
• supragingival cleaning, rubber dam placement with clamps; restorative matrix band/strip placement; endodontics beyond the apical foramen; orthodontic bands; interdental wedges; subgingival placement of retraction cords, antibiotic fibres
Respiratory tract

- Any procedure involving incision or biopsy of mucosa, such as: tonsillectomy/adenoidectomy
- Flexible or rigid bronchoscopy (with incision or biopsy)
- Surgery of the bronchial, sinus, nasal or middle ear mucosa (including tympanoplasty)
Genitourinary and gastrointestinal tract

- Any procedure where antibiotic prophylaxis is indicated for surgical reasons:
- Lithotripsy
- Vaginal delivery with prolonged labour
- Any genitourinary procedure in the presence of genitourinary infection
- Any gastrointestinal procedure in the presence of intra-abdominal infection
Others

• Incision and drainage of local abscess
• Surgical procedure through infected skin
Organisational approach-Aims of a RHD control programme

• Identify and register known cases of ARF and RHD
• Improve uptake of and adherence to secondary prophylaxis
• Increase awareness of diagnosis and management among healthcare providers
• Improve clinical care and follow up in line with best practice
• Support education and health promotion for individuals, families and the community
• Promote primary prevention aimed at preventing initial episodes of ARF
• Use data to monitor patient outcomes and improve program strategies
RHD registers

- Registers of people with RHD or a history of ARF are a key element of RHD control at an individual, community and national level
- Register-based programs:
  - improve case detection
  - increase adherence to secondary
  - reduce recurrences of ARF
  - decrease hospitalisation
National coordination unit for rheumatic heart disease

• RHD Australia was established as the national coordination unit in 2009 to support the control of RHD in Australia.
• RHD Australia, as part of the Rheumatic Fever Strategy, supports efforts to address ARF and RHD by providing:
  • national education, training and self-management resources for primary health care to assist with the detection and treatment of ARF and RHD
  • a performance management system for current activities in the detection and management of acute rheumatic fever and rheumatic heart disease.
Public health approaches to acute rheumatic fever and rheumatic heart disease control

- **Surveillance**
- Passive surveillance of ARF usually depends on case identification from healthcare providers.
- Underestimated the burden of disease due to inaccuracies and incompleteness.
- The APSU notification mechanism relies on voluntary reporting from clinicians working in paediatrics and child health who are registered with the APSU.
- The voluntary nature of reporting, together with the lack of core data for some reported cases, results in an underestimate of the true incidence of ARF.
Active surveillance

• Data includes
• hospital separations
• specialist and radiological reports
• automated alerting of registered patients on presentation to hospital
• review of patients with presenting complaints, possibly due to ARF
• community and staff education aimed at improving case identification.
Legislated notification of acute rheumatic fever and rheumatic heart disease

- In Australia, ARF became notifiable in the Northern Territory in 1994, in Queensland in 1999 and in Western Australia in 2007.
- RHD is not notifiable anywhere in New Zealand or Australia but should be strongly considered.
Screening programmes

• Echo criteria for RHD- separate talk at this forum
Monitoring anticoagulation therapy

• At this time warfarin (a vitamin K antagonist) is the only oral anticoagulant approved for the management of patients with RHD (mechanical valve replacements or valvular AF).
• In the near future, alternatives to warfarin may become available for patients with RHD.
• The major limitation of warfarin is the requirement for monitoring of its therapeutic effect (INR) in the form of regular blood tests.
• Both underanticoagulation and overanticoagulation can lead to a life-threatening event.
Monitoring anticoagulation therapy

• Dosing requirements are variable, as warfarin interacts with many commonly-used medications and food items.
• Difficulties may also arise because of language and cultural barriers, mobility of the population and remoteness from pathology services.
• Achieving satisfactory anticoagulation is often a challenge.
• Point of care INR testing is available, may help
Access to oral healthcare

• Meticulous dental and oral hygiene minimises the risk of infective endocarditis.
• Oral health remains suboptimal in many Aboriginal communities.
• All patients with RHD should undergo annual oral health review
Access to echocardiography

• With portable echocardiography all RHD patients in Australia, regardless of location, have access to this diagnostic imaging tool.

• Obtaining objective evidence and quantification of valvular disease is very important as many barriers prevent accurate history taking/examination.

• All patients with murmurs suggestive of possible valve disease, or a history of ARF, require echocardiography.

• This will detect any valvular lesion and allow assessment of its severity and of LV systolic function.
Access to echocardiography

• Serial echocardiography plays a crucial role in the diagnosis and follow up of rheumatic valve disease

• Allows objective monitoring of any change in the severity of valve lesions, LV chamber size, LV function and any increase in pulmonary artery pressure.

• Objective echocardiographic data are essential in helping to determine the timing of any possible intervention.
Access to cardiothoracic and interventional cardiology services

• As the number of patients undergoing rheumatic valvular procedures is low, few cardiothoracic and cardiology services are experienced in this field.

• Adherence to long-term anticoagulation with warfarin, means that mechanical valve replacement is not always the preferred surgical option.

• Significantly poorer outcome for Aboriginal patients who have undergone valve surgery difficulties in providing follow up specialist care to rural or remote communities, inadequate health literacy and cultural and language barriers.
Access to cardiothoracic and interventional cardiology services

• Early engagement with cardiac surgery and interventional cardiology is essential in determining the appropriate timing of valve surgery and balloon valvuloplasty

• In younger patients, especially children, it is highly desirable that the mitral and even the aortic valve be repaired, rather than these patients receiving prosthetic valves.

• Biological valves are a less desirable alternative if the valve cannot be repaired, as this increases the likelihood of redo surgery later in life because of bioprosthetic degeneration.
Access to cardiothoracic and interventional cardiology services

- In general, mechanical valves should be reserved only for adult patients who are likely to be compliant with warfarin.
- This knowledge is best obtained by direct contact with healthcare providers who work in the patient’s community.
Access to cardiothoracic and interventional cardiology services

• Repairing rheumatic valves is technically more difficult than non-rheumatic valves, particularly MR and AR.
• There is increasing interest in conservative surgery for aortic valve disease-limited success.
• Appropriate resources to accommodate families and Aboriginal liaison staff.
• Ensure discussions regarding the risks and implications of surgery and balloon valvuloplasty.
• The patient, their family and the surgical service understand the affect of the agreed treatment on future child bearing, activity and the need for anticoagulation.
A close partnership between the primary healthcare team, physician/cardiologist and cardiac surgeon is the prerequisite for the optimal care of patients with RHD.
Echocardiographic changes

- Separate talk
Specific Valve lesions and their management

- Mitral regurgitation
- MR is the most common valvular lesion in RHD
- Frequent in young patients who have not yet developed scarred and stenotic valves from persistent or recurrent valvulitis.
- In Aboriginal patients with RHD in the Northern Territory, 40% of the overall patient cohort, and 90% of children aged under 10 years, had pure MR.
Mitral regurgitation

• **Symptoms**
  • May be asymptomatic for many years
  • Exertional dyspnoea and fatigue

• **Examination**
  • Pan-systolic murmur at LV apex
Echocardiography

• Overriding or prolapse of AMVL
• Thickened ‘dog leg’ AMVL, especially if associated with mitral stenosis
• Retrograde colour (mosaic) regurgitant jet into left atrium, often posteriorly directed
• Severity graded by area of colour regurgitant jet in left atrium LV chamber dimensions enlarged if moderate or greater MR Assess LV systolic function
Investigations

- **Cardiac catheterisation**
  Patients aged more than 25–30 years, because of the premature onset of coronary artery disease in this population
Medical management

• In chronic, stable MR (regardless of severity), there is no role for vasodilators, diuretics or ACE inhibitors unless clinical heart failure is present
Choice of operation

- Mitral valve repair operation of choice
- Mitral valve replacement with biological or mechanical prosthesis
- Avoid mechanical prostheses, if concerns about warfarin adherence or future pregnancy
Moderate / severe MR - Indications for surgery

1. NYHA FC II-IV symptoms OR
2. Impaired LV systolic function EF <60% OR
3. LVESD ≥ 40 mm in adults or enlarged LVSED Z-score in children OR
4. PAS hypertension >50 mmHg OR
5. New onset atrial fibrillation
Choice of operation

- Mitral valve repair operation of choice
- Mitral valve replacement with biological or mechanical prosthesis
- Avoid mechanical prostheses, if concerns about warfarin adherence or future pregnancy
Rheumatic mitral stenosis

• **Symptoms**
  • May be asymptomatic
  Exertional dyspnoea, fatigue, palpitations

• **Examination**
  • Low-pitched mid diastolic ‘rumble’ at LV apex
Echocardiography

- Thickened restricted ‘dog leg’ AMVL
- Restricted posterior leaflet
- Measure mean mitral diastolic gradient from continuous-wave Doppler signal
- Calculate MVA from slope of Doppler mitral inflow velocity
- Calculate PAS pressure
Cardiac catheterisation

• Only to exclude coronary artery disease
Atrial fibrillation

• Common
• Rate control using beta-blockers or digoxin
• Consider cardioversion, if recent onset
• Need anticoagulation to prevent thromboembolic complications (only warfarin indicated)
Medical management

• Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic pulmonary venous congestion or pulmonary oedema

• All symptomatic patients should be referred for cardio-surgical assessment
Indications for intervention

- Symptoms NYHA FC II–IV
- MVA <1.5 cm²
- OR PAS pressure >50 mmHg
Procedure of choice

- PBMV by experienced operator/centre
- Mitral valve repair or replacement if morphology is not suitable for PBMV (e.g. valve is heavily calcified) or if moderate or greater MR is present
Rheumatic aortic regurgitation

• **Symptoms**
  • May be asymptomatic for many years
  • Exertional dyspnoea and fatigue

• **Signs**
  • Diastolic blowing and/or decrescendo murmur at left sternal border, usually associated with systolic ejection murmur
Echocardiography

• Retrograde diastolic regurgitant colour jet in LVOT and LV chamber
• Area of jet in LVOT correlates with severity
• LV chamber dimensions enlarged, if moderate or greater aortic regurgitation
• Associated mitral valve disease is common
• Pan-diastolic reversed diastolic flow in descending thoracic aorta, if moderate/severe aortic regurgitation (Doppler)
• Assess LV systolic function
Rhuematic Aortic Regurgiationation

• **Cardiac catheterisation**
  • Only to exclude coronary artery disease

• **Medical management**
  • All symptomatic patients should be commenced on an ACE inhibitor and referred for cardio-surgical evaluation
  • Consider ACE inhibitors or vasodilator therapy with dihydropyridines (e.g. nifedipine) in asymptomatic patients with moderate or greater aortic regurgitation
Indications for surgery

• Moderate/severe aortic regurgitation with symptoms NYHA FC II–IV
• Asymptomatic moderate/severe aortic regurgitation if:
  LVEF <55% OR
  LVESD ≥55 mm OR
• LVEDD >70 mm OR
• Enlarged LVESD or LVEDD Z-score (in children only)
Choice of surgery

- Bioprosthetic or homograft valve replacement: no anticoagulation needed
  - need for redo surgery
  - may allow pregnancy
- Mechanical valve replacement: needs anticoagulation
- Aortic valve repair: limited experience
- Ross procedure (replacement of the aortic valve with a pulmonary autograft and replacement of the pulmonary valve with a homograft) – needs experienced surgeons
Rheumatic aortic stenosis

- **Symptoms**
  - May be asymptomatic
  - Exertional dyspnoea, angina, syncope

- **Signs**
  - Low-pitched, systolic ejection murmur in aortic area

- **Cardiac catheterisation**
  - Only to exclude coronary artery disease
Echocardiography/Medical management

• Thickened, restricted aortic valve leaflets
• Measure peak and mean systolic gradient from Doppler velocity across aortic valve
• Assess left ventricular systolic function
• **Medical management**
  • Medical therapy is not indicated in asymptomatic patients
  • Symptomatic patients require surgery and do not benefit from medical therapy
Indications for surgery

• Symptoms plus mean systolic gradient > 40-50 mmHg or AVA <1.0 cm² OR

• Impaired cardiac function (EF < 50%) plus mean systolic gradient > 40-50 mmHg or AVA <1.0 cm²
Aortic stenosis

• **Choice of surgery**
  • Bioprosthetic or homograft valve replacement:
    • limited durability
  • No anticoagulation needed
  • Mechanical valve replacement:
    • Anticoagulation needed
Rheumatic tricuspid regurgitation

• **Symptoms**
  • Exertional dyspnoea and fatigue, usually secondary to left sided rheumatic valve disease

• **Examination**
  • Elevated jugular venous pressure with prominent v wave in jugular pulse
  • Pansystolic murmur left sternal border
  • Hepatomegaly, may be pulsatile
  • Ascites
  • Peripheral oedema
Echocardiography

- Thickened leaflets
- Retrograde colour jet into right atrium
- Severity graded by area of colour jet
- Dilated IVC
- Retrograde flow in hepatic veins
- Right ventricular chamber enlargement if moderate or greater TR
Medical/Surgical Management

• Symptoms are generally related to the left sided valve lesions
• Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic right and/or left heart failure
• **Indications for surgery**
  • Moderate/severe TR usually in association with symptomatic MVD
  • Progressive symptomatic right heart failure
Choice of surgery

- Tricuspid valvuloplasty
- Tricuspid valve replacement with mechanical or biological prosthesis if valvuloplasty not possible
Rheumatic tricuspid stenosis

Symptoms
Secondary to left sided disease

Examination

- Elevated jugular venous pressure
- Prominent a wave in jugular pulse
- Presystolic and mid diastolic murmur at the left sternal border
Echocardiography

- Thickened, restricted tricuspid valve leaflets with doming
- Diastolic gradient measured across tricuspid valve as per MS

**Medical management**
- Diuretics (e.g. frusemide, spironolactone) are only indicated in patients with symptomatic right and/or left heart failure
Rheumatic tricuspid valve

- **Indications for surgery**
  - Moderate/severe TS in association with symptomatic MVD
  - Progressive right heart failure

- **Choice of surgery**
  - Percutaneous balloon valvuloplasty or surgical commissurotomy operation of choice
  - Tricuspid valve replacement with mechanical or biological prosthesis, if repair or PBTV not possible
Multivalvular disease

• In patients with RHD, both the mitral and aortic valve may be involved (e.g. AR and MS or AR and MR).
• The management is usually that of the dominant lesion.
• However, the proximal valve lesion may modify the effects of the distal lesion; for example, severe MS may prevent the development of significant LV dilation secondary to AR.
• The combination of significant MR and AR is a surgical challenge, and carries a higher risk of ventricular dysfunction.
• In many patients, one valve lesion will be more dominant than the other. Indications for surgery are as for the dominant lesion described in earlier sections.
Multivalvular disease

• What to do with the less severe valve lesion?
• Mild to moderate MR in the presence of significant AR poses a difficult surgical management problem.
• The progression of the milder valve lesion is variable.
• Double valve surgery carries a higher surgical risk than single valve surgery, particularly in the presence of LV dysfunction.
• It is important that the severity of the less severe valve lesion be accurately quantified preoperatively with transoesophageal echocardiography.
• Thank you