An Australian guideline for rheumatic fever and rheumatic heart disease: an abridged outline

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ABSTRACT

- Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) are diseases of poverty. They occur at world-record rates in Indigenous Australians, yet individual cases are often poorly managed, and most jurisdictions with high rates of these diseases do not have formal control strategies in place.
- New Australian guidelines formulated in 2005 by the National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand for diagnosis and management of ARF and RHD are a valuable resource for clinicians and policymakers.
- Key recommendations of the guidelines include:
  - ➢ New diagnostic criteria for ARF in high-risk populations, including Indigenous Australians, which include echocardiographic evidence of subclinical valvular disease, and polyarthralgia or aseptic monoarthritis as major manifestations.
  - ➢ Clear guidance about treatment of ARF. Non-steroidal anti-inflammatory drugs should be withheld until the diagnosis is confirmed, and corticosteroids may be an option in severe acute carditis. Most cases of chorea do not require medication, but use of carbamazepine or sodium valproate is recommended if medication is needed.
  - ➢ Clear guidance about dose, dosing frequency and duration of secondary prophylaxis. Benzathine penicillin G is the preferred medication for this purpose.
  - ➢ Establishment of a coordinated control program for all regions of Australia where there are populations with high prevalence of ARF and RHD. Key elements and indicators for evaluation are recommended.
  - ➢ Active screening and legislated notification of ARF and RHD, where possible.
  - ➢ Development of a structured care plan for all patients with a history of ARF or with established RHD, to be recorded in the patient's primary health care record.

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For editorial comment, see page 557

The National Heart Foundation of Australia (NHFA) and the Cardiac Society of Australia and New Zealand (CSANZ) recently launched an evidence-based review and guideline entitled *Diagnosis and management of acute rheumatic fever and rheumatic heart disease in Australia*.[1] The full review and guideline, along with five quick reference guides for professionals, are available at the NHFA website (http://www.heartfoundation.com.au). The process of guideline development is summarised in Box 1 and the grades of recommendation are outlined in Box 2.

Background

Acute rheumatic fever (ARF) is an autoimmune consequence of infection with group A streptococci (GAS). It causes an acute generalised inflammatory response and an illness that selectively affects the heart, joints, brain and skin. People with ARF are often severely unwell and in great pain, and require hospitalisation. Despite the dramatic nature of an acute episode, ARF leaves no lasting damage to the brain, joints or skin.

However, damage to the heart valves, particularly the mitral and aortic valves, may persist after an acute episode has resolved. This involvement of the cardiac valves is known as rheumatic heart disease (RHD). People who have had ARF previously are much more likely to have subsequent episodes, and these recurrences may cause further damage to the cardiac valves. Thus RHD steadily worsens in people who have multiple episodes of ARF.[4]

A recent review of the global burden of GAS-related disease estimated that there are at least 15.6 million people with RHD, another 1.9 million with a history of ARF but no carditis (still requiring preventive treatment), 470 000 new cases of ARF each year, and over 230 000 deaths due to RHD annually.[5] Almost all cases and deaths occur in developing countries. Because of its high prevalence in developing countries, RHD is the most common form of paediatric heart disease in the world. In many countries it is the most common cause of cardiac mortality in children and adults aged less than 40 years.

Currently, the highest documented rates of ARF and RHD in the world are in Indigenous Australians. A recent summary of the data on the disease burden of ARF and RHD in Australia concluded that these diseases are almost exclusively restricted to Aboriginal and Torres Strait Islander people living in regional and remote areas of central and northern Australia.[6] The annual incidence of ARF in Aboriginal children aged 5–14 years in the Northern Territory ranged from 250 to 350 per 100 000. Enhanced surveillance in northern Queensland between 1999 and 2004 found an annual incidence of ARF in Aboriginal children aged 5–14 years of 162 per 100 000.[7]

In the NT in 2002, the prevalence of RHD was 13–17 cases per 1000 Aboriginal people of all ages, compared with less than two

1 Guideline development process

1. Expert writing group prepared an initial draft.
2. Thirty-seven selected individuals reviewed the draft chapters, and their suggestions were incorporated into a second draft.
3. The revised draft was widely circulated to 17 stakeholder organisations for comment, and stakeholders were also invited to a 1-day workshop in November 2004.
4. At the workshop, the stakeholders reviewed the draft and reached a consensus on areas of disagreement.
5. A third draft was prepared and redistributed to stakeholder organisations for further comment.
6. The final draft was then prepared and endorsed by 10 stakeholder organisations as well as the National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. ◆

* Other members of the Writing Group were Warren Walsh, Clive Hadfield, Diana Lennon, Lynette Purton and Gavin Wheaton.

2 Grades of recommendation and levels of evidence*

Grade	Level of evidence
A	Rich body of high-quality RCT data
B	Limited body of RCT data or high-quality non-RCT data
C	Limited evidence
D	No evidence available — panel consensus judgement

RCT = randomised controlled trial. * Levels of evidence and grades of recommendation are adapted from National Health and Medical Research Council levels of evidence for clinical interventions[2] and US National Institutes of Health clinical guidelines.[3] ◆

cases per 1000 non-Indigenous people living in the same region.[6] Indigenous people are up to eight times more likely than non-Indigenous Australians to be hospitalised for ARF and RHD, and nearly 20 times as likely to die. Forty-five per cent of Indigenous people receiving heart valve surgery for RHD are aged less than 25 years, compared with only 4% of non-Indigenous Australians.

Several factors contribute to inadequate diagnosis and management of ARF and RHD in Australia:

- Although strategies for preventing RHD are proven, simple, cheap and cost-effective, they are not adequately implemented — indeed, they are sometimes not implemented at all in the populations at highest risk of the disease;
- As ARF is rare in most metropolitan centres where health professionals train and practise, the majority of clinicians will have seen very few, if any, cases of ARF;
- There is great variability in the management of ARF and RHD. Lack of up-to-date training and experience occasionally results in inappropriate management of these diseases; and
- Population groups who experience the highest rates of ARF and RHD have limited access to health care services.

The guidelines do not cover primordial or primary prevention in detail. This is because the key to primordial prevention is reducing exposure to GAS, which requires dramatic improvements in housing, hygiene infrastructure and access to health care for Indigenous Australians. Beyond these reforms, there are no systematic approaches that are proven to reduce the incidence of ARF. It should be noted that penicillin treatment of streptococcal pharyngitis (primary prophylaxis) has been proven to reduce the occurrence of subsequent episodes of ARF,[8] and this should continue to be emphasised to health staff. However, systematic screening and treatment of sore throats — for example, in school-based programs — has not been proven to be cost-effective.[9]

3 2005 Australian guidelines for the diagnosis of acute rheumatic fever (ARF)*

	High-risk groups†	All other groups
Initial episode of ARF	2 major *OR* 1 major and 2 minor manifestations, *PLUS* evidence of a preceding group A streptococcal infection‡	As for high-risk groups
Recurrent attack of ARF in a patient with known past ARF or RHD	2 major *OR* 1 major and 2 minor *OR* 3 minor manifestations, *PLUS* evidence of a preceding group A streptococcal infection‡	As for high-risk groups
Major manifestations	Carditis (including subclinical evidence of rheumatic valvular disease on echocardiogram) Polyarthritis or aseptic monoarthritis or polyarthralgia§ Chorea¶ Erythema marginatum** Subcutaneous nodules	Carditis (excluding subclinical evidence of rheumatic valvular disease on echocardiogram) Polyarthritis§ Chorea¶ Erythema marginatum** Subcutaneous nodules
Minor manifestations	Fever†† ESR ⩾ 30 mm/h or CRP level ⩾ 30 mg/L Prolonged PR interval on ECG‡‡	Fever†† ESR ⩾ 30 mm/h or CRP level ⩾ 30 mg/L Prolonged PR interval on ECG‡‡ Polyarthralgia or aseptic monoarthritis§

CRP = C-reactive protein. ECG = electrocardiogram. ESR = erythrocyte sedimentation rate. RHD = rheumatic heart disease.

* All categories assume that other more likely diagnoses have been excluded. Patients who do not fulfil the criteria listed here, but for whom the clinician remains suspicious that the diagnosis may be ARF, should be offered a single dose of benzathine penicillin G at secondary prophylaxis doses and reviewed in 1 month with a repeat echocardiogram to detect the appearance of new lesions. If there is evidence of rheumatic valvular disease, clinically or on echocardiogram, the diagnosis is confirmed, and long-term secondary prophylaxis can be continued. See text for details about specific manifestations.

† High-risk groups are those living in communities with high rates of ARF (incidence > 30 per 100 000 per year in 5–14-year-olds) or RHD (all-age prevalence > 2 per 1000). Indigenous Australians living in rural or remote settings are known to be at high risk. Data are not available for other populations, but Indigenous Australians living in urban settings, Māori and Pacific Islander people, and, potentially, immigrants from developing countries may also be at high risk.

‡ Elevated or rising level of antistreptolysin O or other streptococcal antibody; positive throat swab culture; or positive rapid antigen test for group A streptococci.

§ A definite history of arthritis is sufficient to satisfy this manifestation. Other causes of arthritis or arthralgia should be excluded, particularly in the case of monoarthritis (eg, septic arthritis due to disseminated gonococcal infection); infective or reactive arthritis (eg, due to Ross River virus, Barmah Forest virus, influenza virus, rubella virus, *Mycoplasma*, cytomegalovirus, Epstein–Barr virus, parvovirus, hepatitis or *Yersinia*); autoimmune arthropathy (eg, associated with juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis or sarcoidosis). If polyarthritis is present as a major manifestation, polyarthralgia or aseptic monoarthritis cannot be considered an additional minor manifestation in the same person.

¶ Rheumatic (Sydenham's) chorea does not require other manifestations or evidence of preceding group A streptococcal infection, provided other causes of chorea are excluded.

** Erythema marginatum is a distinctive rash. Care should be taken not to label other rashes, particularly non-specific viral exanthems, as erythema marginatum.

†† Oral, tympanic or rectal temperature ⩾ 38°C on admission or documented during the current illness.

‡‡ If carditis is present as a major manifestation, prolonged PR interval cannot be considered an additional minor manifestation in the same person. ◆

4 Priorities in managing acute rheumatic fever (ARF)

Admission to hospital

Confirmation of diagnosis

- Observation before anti-inflammatory treatment (paracetamol [first-line] or codeine for fever or joint pain)
- Investigations (see text under "Diagnosis of acute rheumatic fever")

Treatment

All cases

- Single dose intramuscular benzathine penicillin G (preferable) or oral penicillin V for 10 days (intravenous penicillin not needed; oral erythromycin may be used if patient allergic to penicillin)

Arthritis and fever

- Paracetamol (first-line) or codeine until diagnosis confirmed
- Aspirin (first-line) or naproxen once diagnosis confirmed, if arthritis or severe arthralgia present
- Mild arthralgia and fever may respond to paracetamol alone
- Influenza vaccine for children receiving aspirin during the influenza season (autumn/winter)

Chorea

- No treatment for most cases
- Carbamazepine or valproic acid if treatment is necessary (see text under "Treatment of acute rheumatic fever")

Carditis/heart failure

- Bed-rest
- Urgent echocardiogram
- Anti-heart failure medication
  - Diuretics/fluid restriction for mild to moderate heart failure
  - ACE inhibitors for more severe heart failure, particularly if aortic regurgitation present
  - Glucocorticoids optional for severe carditis (consider treating for possible opportunistic infections)
  - Digoxin if atrial fibrillation present
- Valve surgery for life-threatening acute carditis (rare)

Long-term preventive measures

- Give first dose of secondary prophylaxis
- Notify case for recording in ARF/RHD register, if available
- Contact local health staff to ensure follow-up
- Provide culturally appropriate education to patient and family
- Arrange dental review and ongoing dental care to reduce risk of endocarditis

ACE = angiotensin-converting enzyme. RHD = rheumatic heart disease. ◆

Diagnosis of acute rheumatic fever

The Jones criteria for diagnosing ARF divide its clinical features into major and minor manifestations.[10] There is some concern that these criteria may not be sensitive enough in high-incidence populations, where the consequences of under-diagnosis may be greater than those of over-diagnosis. An expert group convened by the World Health Organization in 2001 provided additional guidelines on how the Jones criteria should be applied in primary and recurrent episodes.[4] Because the Jones and WHO criteria appear too restrictive for diagnosing ARF in Australian Indigenous populations, the NHFA and CSANZ have agreed on new criteria for use in high- and low-risk populations in Australia (Box 3). In high-risk populations, echocardiographic evidence of subclinical valvular disease and polyarthralgia or aseptic monoarthritis can be considered major manifestations of ARF. All patients with possible or confirmed ARF should undergo echocardiography to help in making the diagnosis and to determine the level of cardiac involvement (grade C recommendation).

Other investigations recommended in suspected ARF cases include biochemical testing (for levels of inflammatory markers), blood culture (if the patient is febrile), an electrocardiogram (to detect a prolonged PR interval), a chest x-ray (if there is evidence of carditis), a throat swab (for culture of GAS), and streptococcal serology (both antistreptolysin O and anti-DNase B titres). In seeking alternative diagnoses, investigations may include repeat blood cultures for possible endocarditis; joint fluid culture for possible septic arthritis; levels of copper, ceruloplasmin, antinuclear antibody and drugs for other causes of abnormal movements; and serology and autoimmune markers for arboviral, autoimmune or reactive arthritis.

Two recently described putative syndromes associated with infection with GAS are post-streptococcal reactive arthritis and paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections. However, because of difficulties in differentiating them from ARF, these conditions should rarely, if ever, be diagnosed in high-risk populations such as Indigenous Australians (grade C recommendation).

Treatment of acute rheumatic fever

The priorities in managing ARF are listed in Box 4. With very few exceptions, all patients with definite or possible ARF should be admitted to hospital (grade D recommendation). The arthritis of ARF is exquisitely responsive to treatment with non-steroidal anti-inflammatory drugs (NSAIDs). Indeed, this can be a useful diagnostic feature, as arthritis continuing unabated more than 3 days after starting NSAID therapy is unlikely to be due to ARF. Equally, withholding NSAIDs in patients with monoarthralgia or mono-arthritis to observe the development of polyarthritis can help confirm the diagnosis of ARF. Therefore, salicylates or NSAIDs should be withheld until the diagnosis is confirmed. Joint pain can be treated with paracetamol (grade D recommendation). Most cases of chorea can be managed without medication. Where necessary — because of severe movement disorder or distress — treatment with carbamazepine or valproic acid is recommended (grade B recommendation). Treatment with corticosteroids has not been proven to alter the likelihood of developing RHD or its degree of severity.[11] However, this statement is not based on high-quality evidence, and there are no published trials of the possible shorter-term benefits of corticosteroids in severe carditis, the condition for which they are usually reserved. Therefore, the use of these medications is optional, but if used, they should be reserved for severe carditis only (grade D recommendation).

Secondary prevention

Regular administration of antibiotics to prevent infection with GAS and subsequent possible recurrent ARF is recommended for all people with a history of ARF or RHD (grade A recommendation). This strategy has been proven in randomised controlled trials to

5 Recommended antibiotic regimens and their duration for secondary prevention of acute rheumatic fever (ARF) and rheumatic heart disease (RHD)

Antibiotic	Dose	Route	Frequency	Duration
First-line treatment				
Benzathine penicillin G (BPG)	1 200 000 U (body weight ⩾ 20 kg) 600 000 U (body weight < 20 kg)	Deep intramuscular injection	4-weekly, or 3-weekly for selected groups*	All people with ARF or RHD: • Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer)
Second-line treatment (if intramuscular routine not possible or refused)†				Status after initial period has elapsed:‡ • No RHD or mild RHD: discontinue at that time • Moderate RHD: continue until age 35 years • Severe RHD: continue until age 40 years or longer§
Phenoxymethylpenicillin (penicillin V)	250 mg	Oral	Twice daily	
Treatment in cases of documented penicillin allergy				
Erythromycin	250 mg	Oral	Twice daily	

* 3-weekly BPG injections may be considered for patients with moderate to severe carditis or a history of valve surgery who demonstrate good adherence to less frequent injections and for those who have confirmed breakthrough ARF despite full adherence to 4-weekly BPG treatment. Monthly BPG injections are an acceptable alternative only if it is considered that the practicalities of monthly dosing will substantially improve adherence.

† If oral regimens are prescribed, adherence should be carefully monitored.

‡ Decisions to cease secondary prophylaxis should be based on clinical and echocardiographic assessment. See full guideline for definitions of RHD severity.

§ Risk of recurrence is extremely low in people aged > 40 years. In some cases (eg, if a patient wants to reduce even a minimal risk of recurrence), prophylaxis may be continued beyond age 40 years, or even for life. ◆

prevent streptococcal pharyngitis and recurrent ARF and to reduce the severity of and mortality from RHD. Intramuscular benzathine penicillin G (BPG) is superior to oral penicillin.[12] Dosing regimens are shown in Box 5. A lower dose for children weighing < 20 kg has been shown in New Zealand to maintain low ARF recurrence rates, although the number of children receiving this regimen was small.[13] BPG is most effectively given as a deep intramuscular injection into the upper outer quadrant of the buttock or the anterolateral thigh.

Three-weekly delivery of BPG has been shown to be more effective than 4-weekly delivery in some studies.[12,14-16] Although Australian Indigenous people are at higher risk of developing ARF than other ethnic groups in Australia, the benefits of 3-weekly BPG injections for Indigenous Australians in remote areas are offset by the difficulty of achieving good treatment adherence, even to the standard 4-weekly regimen. Moreover, prospective data from New Zealand have shown that there are few, if any, recurrences of ARF among people who adhere to a strict 4-weekly BPG regimen.[13] Thus the use of 4-weekly BPG is currently the treatment of choice, except in patients with moderate to severe carditis or a history of valve surgery who show good adherence to less frequent injections, and those who have confirmed breakthrough ARF despite full adherence to a 4-weekly BPG regimen, in whom 3-weekly administration is recommended (grade D recommendation). Monthly rather than 4-weekly administration of BPG is an acceptable alternative only if it is considered that the practicalities of monthly dosing will substantially improve adherence (grade D recommendation).

Oral penicillin is less effective than BPG in preventing group A streptococcal infections and subsequent recurrences of ARF. Moreover, in comparison with intramuscular BPG, twice-daily oral regimens are likely to result in poorer rates of adherence over long time periods and less predictable serum penicillin concentrations. Oral penicillin should be reserved for patients who refuse intramuscular BPG (grade B recommendation). If a patient is offered oral penicillin, the consequences of missed doses must be emphasised, and adherence needs to be carefully monitored (grade D recommendation).

Before commencing penicillin treatment, patients should be carefully questioned about known allergies to penicillin or other β-lactam antibiotics. If there is substantive evidence of immediate and severe allergic reaction to penicillin, a non-β-lactam antimicrobial (eg, erythromycin) should be used instead (grade D recommendation). If patients state they are allergic to penicillin but there is no unequivocal evidence of a previous allergic reaction, they should be investigated for penicillin allergy, preferably in consultation with an allergist. Penicillin desensitisation is not applicable to these patients, as it would have to be repeated before each dose of BPG.

As there is no evidence of teratogenicity associated with it, penicillin prophylaxis for prevention of recurrent ARF should continue for the duration of pregnancy (grade D recommendation). Erythromycin is also considered safe for use in pregnancy, although controlled trials have not been conducted. Intramuscular bleeding from BPG injections, used in conjunction with anticoagulation therapy in Australia, is rare. Thus, BPG injections should be continued for patients receiving an anticoagulant, unless there is evidence of uncontrolled bleeding or the international normalised ratio is above the defined therapeutic range (grade D recommendation).

The appropriate duration of secondary prophylaxis is determined by age, time since the last episode of ARF, and potential harm from recurrent ARF (Box 6) (grade D recommendation).

Poor adherence to treatment is rarely due to injection refusal, the pain of injections, or a lack of knowledge or understanding of ARF/RHD in remote Indigenous communities.[17] The major determining factors are the availability and acceptability of health services and the presence of active reminder and recall systems. Adherence is substantially better in health centres where there is active follow-up if BPG doses are missed and where a dedicated staff member administers the BPG (AB, unpublished data).

Infective endocarditis is a dangerous complication of RHD and a common adverse event after prosthetic valve replacement in Indigenous Australians.[18] People with established RHD or prosthetic valves should receive antibiotic prophylaxis before undergoing

6 Recommended routine review and management plan for acute rheumatic fever (ARF) and rheumatic heart disease (RHD)

Classification	Criteria*	Review and management plan	Frequency†
Low risk	ARF with no evidence of RHD *OR* Trivial to mild valvular disease	• Secondary prophylaxis (BPG) • Doctor review • Echocardiography	• 4-weekly • Yearly • Children 2-yearly;‡ adults 2–3-yearly‡
Medium risk	Any moderate valve lesion in the absence of symptoms and with normal left ventricular function *OR* Mechanical prosthetic valves	• Secondary prophylaxis (BPG) • Doctor review • Influenza vaccination • ECG (optional) • Cardiologist/physician/paediatrician review • Echocardiography • Dental review • Polysaccharide pneumococcal vaccination (Pneumovax 23 [Merck Sharp & Dohme]) • Endocarditis prophylaxis	• 4-weekly • 6-monthly • Yearly • Yearly • Yearly • Yearly • Yearly • 5-yearly (maximum 3 doses) • As required
High risk§	Severe valvular disease *OR* Moderate to severe valvular lesion with symptoms *OR* Tissue prosthetic valves and valve repairs	• Secondary prophylaxis (BPG) • Doctor review • Cardiologist/physician/paediatrician review • Influenza vaccination • Echocardiography • Dental review • Polysaccharide pneumococcal vaccination (Pneumovax 23 [Merck Sharp & Dohme]) • Endocarditis prophylaxis • Warfarin + aspirin	• 3–4-weekly • 3–6-monthly • 3–6-monthly • Yearly • 3–6-monthly • Within 3 months and yearly thereafter • 5-yearly (maximum 3 doses) • As required • As prescribed
Additional considerations	Following valve surgery	• Medical assessment • ECG • Chest radiograph • Echocardiography • Full blood count • Levels of urea, creatinine, electrolytes • INR if indicated	• 3–4 weeks after discharge
	Missed doses of BPG	• Patient should be contacted if he or she has not presented within 3 days of due injection	
	Patient travelling to another community when injection due	• Consider bringing forward date of injection to 2–3 weeks, or make arrangements with other service providers in advance	

BPG = benzathine penicillin G. ECG = electrocardiogram. INR = international normalised ratio.

* Serial echocardiographic assessments are required for long-term management of RHD. If cultural differences or difficulties with communication hinder standard clinical measures of heart failure (eg, New York Heart Association criteria), serial echocardiography becomes an essential tool for determining the progress of cardiac damage and optimal timing of surgery. Thus, risk stratification should be based on clinical and echocardiographic findings.

† Frequency of review should be determined according to individual needs and local capacity. Most critically, review should become more frequent in the event of symptom onset, symptomatic deterioration, or a change in clinical findings.

‡ In patients with no evidence of valvular disease on echocardiography, no documented ARF recurrences, good adherence to secondary prophylaxis, and no cardiac murmurs on examination at follow-up appointments, echocardiography may be needed less frequently.

§ Any patient with severe valvular disease or moderate to severe valvular disease with symptoms should be referred for cardiological and surgical assessment as soon as possible. ◆

procedures expected to produce bacteraemia. Individuals with a history of ARF but no valvular damage do not require antibiotic prophylaxis. Those already receiving penicillin for secondary prophylaxis should be offered a different antibiotic for prophylaxis of endocarditis. Recommendations are outlined in the full guideline.

Rheumatic heart disease control programs

A coordinated control program is the most effective approach for improving BPG adherence and clinical follow-up of people with RHD.[4,13,19] The other aims of control programs are to identify and register new cases of ARF and RHD; to provide education and training for health care providers; to provide education and health promotion for individuals, families and the community; to promote primary prevention of ARF; and to monitor patient outcomes and improve program strategies.

Maintaining registers of people with RHD or a history of ARF is a key element of RHD control at individual, community and national levels. Register-based programs improve case detection, increase adherence to secondary prophylaxis, reduce recurrences of ARF, and reduce hospitalisations for ARF and RHD. Registers also provide a mechanism for monitoring patient movements,

orientating staff to ongoing care requirements (eg, BPG injections, clinic appointments and echocardiograms), and identifying individuals with poor adherence to long-term therapy for targeted educational activities and other interventions. Registers can also provide data for monitoring the success of programs and changes in disease epidemiology. It is recommended that all regions of Australia in which there are substantial numbers of people with ARF or RHD establish a coordinated control program (see full guideline for recommended elements of programs) (grade C recommendation).

A dedicated coordinator is critical to the success of the program. This person should have skills in data management, basic epidemiology and clinical medicine, or ready access to clinical expertise when individual case management issues arise. Ideally, active surveillance of ARF and RHD should be used to augment passive surveillance. Active surveillance could include mechanisms allowing access to hospital separation data, echocardiography reports, specialist review correspondence, primary health care clinic information, and notifiable disease databases. Where possible, these processes should be automated (eg, with regular downloads of information about patients admitted to hospital with a diagnosis of ARF or RHD). Active screening and legislated notification of ARF and RHD should be considered where possible (grade D recommendation). Proposed indicators for evaluating programs are outlined in the full guideline.

Management of rheumatic heart disease

A structured care plan should be developed and recorded in the primary health care record of all people with a history of ARF or with established RHD (grade D recommendation). The recommended care plan schedules listed in Box 6 may be tailored to the needs of the individual. Medical and surgical management of rheumatic valvular lesions is reviewed in the full guideline, and will be summarised in a separate publication.

Competing interests

None identified.

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