New Zealand
Guidelines for Rheumatic Fever

1. Diagnosis, Management and Secondary Prevention

Evidence-based, best practice Guidelines on:
1. Diagnosis, Management and Secondary Prevention
2. Sore Throat Management
3. Proposed Rheumatic Fever Primary Prevention Programme
Endorsed by:
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Evidence-based, best practice
New Zealand Guidelines for Rheumatic Fever

1. DIAGNOSIS, MANAGEMENT AND SECONDARY PREVENTION

He korokoro ora he manawa ora,
Mo tatou katoa

(A healthy throat, a healthy heart for us all)

JUNE 2006
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1. Scope and Purpose of Guideline

This guideline has been developed by The National Heart Foundation of New Zealand and the Cardiac Society of Australia and New Zealand. This guideline will be complemented by further guidelines on appropriate sore throat management and primary prevention of acute rheumatic fever (ARF).

The objectives of this guideline are:

- to identify and present the evidence for best practice in ARF diagnosis
- to identify the standard of care that should be available to all people in New Zealand
- to identify areas where current management strategies may not be in line with available evidence
- to ensure that high-risk populations receive the same standard of care as that available to other New Zealanders.

2. About the Guideline

This guideline was developed by a writing group comprised of experts in rheumatic fever. Selected individuals with experience in ARF and relevant stakeholders were also involved. These included a range of general and specialist clinicians, allied health professionals, Maori and Pacific professionals, and lay representative groups.

This guideline has been produced for New Zealand and is endorsed by New Zealand organisations.

The chairs of the guideline writing committee were involved in the development of a similar document for the Australian population, with the understanding that the Australian guidelines would be adapted for the New Zealand setting. We are grateful for the contribution of our Australian colleagues.

The development process is described in Appendix A.

Disclaimer

This document has been produced by The National Heart Foundation of New Zealand and the Cardiac Society of Australia and New Zealand for health professionals. The statements and recommendations it contains are, unless labelled as “expert opinion”, based on independent review of the available evidence. Interpretation of this document by those without appropriate health training is not recommended, other than at the request of, or in consultation with, a relevant health professional.

In addition, the recommendations in this guideline are not intended to replace clinical judgment of each individual case. Treatment should take into account comorbidities, drug tolerance, lifestyle, living circumstances, cultural sensibilities and wishes. When prescribing medication, clinicians should observe usual contra-indications, be mindful of potential adverse drug interactions and allergies, monitor responses and ensure regular review.
Outline of grading methodology used

The review includes levels of evidence and accompanying grades of recommendation (Table 1).

Table 1. Levels of Evidence for Clinical Interventions and Grades of Recommendation

<table>
<thead>
<tr>
<th>LEVEL OF EVIDENCE</th>
<th>STUDY DESIGN</th>
<th>GRADE OF RECOMMENDATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Evidence obtained from a systematic review of all relevant randomised controlled trials (RCT)</td>
<td>A</td>
</tr>
<tr>
<td>II</td>
<td>Evidence obtained from at least one properly designed randomised controlled trial</td>
<td>B</td>
</tr>
<tr>
<td>III-I</td>
<td>Evidence obtained from well-designed pseudo-randomised controlled trials (alternate allocation or some other method)</td>
<td>B</td>
</tr>
<tr>
<td>III-2</td>
<td>Evidence obtained from comparative studies with concurrent controls and allocation not randomised (cohort studies), case-control studies, or interrupted time series with a control group</td>
<td>B</td>
</tr>
<tr>
<td>III-3</td>
<td>Evidence obtained from comparative studies with historical control, 2 or more single-arm studies, or interrupted time series with a parallel control group</td>
<td>C</td>
</tr>
<tr>
<td>IV</td>
<td>Evidence obtained from case series, either post-test or pre-test and post-test</td>
<td>C</td>
</tr>
<tr>
<td></td>
<td>Insufficient evidence available – expert opinion or panel consensus judgment</td>
<td>D/I</td>
</tr>
</tbody>
</table>

Note: The levels of evidence and grades of recommendations are adapted from the National Heart Foundation of Australia Rheumatic Fever guidelines. (Details can be found at www.nhf.com.au)

Endorsing organisations

- The Cardiac Society of Australia and New Zealand
- The National Heart Foundation of New Zealand, along with:
  - Te Hotu Manawa Maori
  - Pacific Islands Heartbeat
  - Paediatric Society of New Zealand
  - The Rheumatic Fever Trust.

Organisations consulted

- Australasian Society for Infectious Diseases
- Australasian Faculty of Public Health Medicine
- National Heart Foundation of Australia
- New Zealand Nurses Organisation
- New Zealand Ministry of Health
- Pasifika Medical Association of New Zealand
- Royal Australasian College of Physicians
- Te Ohu Rata o Aotearoa - Maori Medical Practitioners Association.
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Declaration

No conflicts of interest were involved in the development of this guideline. Dr Polly Atatoa-Carr who coordinated the writing of this guideline was funded by The National Heart Foundation of New Zealand and the Australasian Faculty of Public Health Medicine.
3. Introduction

Key points
- Acute rheumatic fever, an auto-immune response to group A streptococcus infection of the upper respiratory tract, may result in damage to the mitral and/or aortic valves and therefore rheumatic heart disease. Recurrences are likely in the absence of preventative measures and may cause further cardiac valve damage.
- Although acute rheumatic fever is rare in industrialised countries, it is a significant cause of disease among Maori and Pacific children in New Zealand. The incidence of rheumatic heart disease is also high among these populations, with significant rates of procedures and death among young adults.
- Appropriate treatment of sore throats in high risk populations will eliminate group A streptococcus in most cases, and prevent individual cases of acute rheumatic fever.
- Prevention of recurrences, and therefore rheumatic heart disease prevention, with intramuscular penicillin is both effective and highly cost-effective.

Acute rheumatic fever (ARF) is an auto-immune consequence of infection with the bacterium group A streptococcus (GAS). It causes an acute generalised inflammatory response and an illness that affects only certain parts of the body, mainly the heart, joints, brain and skin. Individuals with ARF are often severely unwell, in great pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF leaves no lasting damage to the brain, joints or skin. However, the damage to the heart, or more specifically the mitral and/or aortic valves, may remain once the acute episode has resolved. This is known as rheumatic heart disease (RHD). People who have had ARF previously are much more likely than the wider community to have subsequent episodes. These recurrences of ARF may cause further cardiac valve damage. Hence RHD steadily worsens in people who have multiple episodes of ARF.

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.

Pathogenesis
ARF has been shown to develop in approximately one to three percent of those in an epidemic situation of untreated exudative pharyngitis and/or a culture positive for GAS. Despite the high incidence in some ethnic groups (such as Maori and Pacific people in New Zealand), a genetic predisposition to ARF remains unproven. Some strains of GAS have been repeatedly identified as causative in ARF (and therefore labelled “rheumatogenic”) and other rheumatogenic strains continue to appear. The role of skin infections remains uncertain.

Following GAS infection, there is a latent period averaging three weeks before the symptoms of ARF begin. By the time the symptoms develop, the infecting strain of GAS has usually been eradicated by the host immune response.

Epidemiology
The burden of ARF in industrialised countries declined dramatically during the 20th Century, due mainly to improvements in living standards (and hence reduced transmission of GAS) and better availability of medical care. In most affluent populations ARF is now rare. RHD is also rare in younger people in industrialised countries, although it is still seen in some elderly patients, a legacy of ARF half a century earlier.

By contrast, ARF and RHD remain common in many developing countries. A recent review of the global burden of GAS-related disease estimated that there is a minimum of 15.6 million people with RHD, another 1.9 million with a history of ARF but no carditis who still require preventive treatment, 470,000 new cases of ARF each year and over 230,000 deaths due to RHD annually. Almost all cases and deaths occur in developing countries. These figures are all likely to be underestimates of the true burden of the disease.
There is substantial regional variation in the burden of ARF and RHD. The highest documented rates in the world have been found in Māori and Pacific people in New Zealand, Aboriginal Australians and those in Pacific Island nations.\textsuperscript{9,10,11} The prevalence of RHD is also high in Sub-Saharan Africa, Latin America, the Indian subcontinent, the Middle East and Northern Africa.\textsuperscript{8}

New Zealand has had sustained high rates of ARF and RHD for many decades with RHD being a significant cause of premature death in this country.\textsuperscript{9,12,13,14} A number of surveys of ARF and RHD incidence have been conducted since the early 1900s in New Zealand. In the 1920s, surveys of school records in New Zealand determined an approximate annual total population incidence of ARF of 65 per 100,000.\textsuperscript{8} From 1956 to 1973, the Wairoa College Study determined that the decline in incidence of ARF seen in other developed countries was not evident in New Zealand and those pockets of the country which experienced isolation and socio-economic deprivation had significantly higher rates of both ARF and RHD.\textsuperscript{8,9}

From 1995 to 2000, around 100 cases of ARF were notified annually in New Zealand, with an incidence of 13.8 per 100,000 population in 5 to 14 year olds.\textsuperscript{15} From 1993 to 1999, the Auckland Register recorded an incidence of 21.9 per 100,000 population in 5 to 14 year olds. Auckland accounts for 60% of the active cases on New Zealand registers.\textsuperscript{16,17}

ARF is predominantly a disease of children aged between 5 to 14 years, with a peak at around eight years. It is rare to diagnose ARF under the age of three (before full maturation of the immune system).\textsuperscript{18,19} As RHD represents the cumulative heart damage of previous ARF episodes, the prevalence of RHD peaks in the third and fourth decades of life.\textsuperscript{20,21} Therefore, although ARF is a disease with its roots in childhood, its effects are felt throughout adulthood, especially in the young adult years when people might otherwise be at their most productive.

The disparity of ethnicity in rheumatic fever populations has been described in many world centers where population groups experiencing low socio-economic status and living in overcrowded situations present with a high incidence of ARF.\textsuperscript{19} In New Zealand, Māori and Pacific peoples have the highest burden of both ARF and RHD. Despite the significant issues regarding the accuracy of ethnicity data in past morbidity and mortality statistics, the rates of ARF in Māori have always been reported as significantly greater than those seen in non-Māori. For example, from 1949 to 1953 the reported incidence of ARF in Māori children (rates of greater than 1000 per 100,000) was 11 times that of the non-Māori population.\textsuperscript{8} The age-specific annual notification rates for ARF between 1990 to 1995 for children aged 10 to 14 years was 77.7 per 100,000 for Pacific children, 30.4 per 100,000 for Māori children and 1 per 100,000 for European children.\textsuperscript{19} Auckland also displays this pattern: the annual incidence of ARF in 5 to 14 year old Māori children from 1993 to 1999 was 41.2 per 100,000 population, Pacific children 83.7 per 100,000 population/year and the rest of the population 1.4 per 100,000 population/year.\textsuperscript{15} Depending on the year analysed, the Pacific hospitalisation rates are at least nine times that of Europeans/others. The Māori hospitalisation rate is just over five times that of Europeans/others.\textsuperscript{22}

As well as higher rates of initial ARF incidence, Māori and Pacific people also have the highest rates of ARF recurrence. From 1973 to 1982 (prior to the introduction of systematic prophylaxis delivery) recurrence rates in Māori were 40% compared to 22% in non-Māori.\textsuperscript{23} A review of cases in the Auckland rheumatic fever register from 1993 to 1999 found that although the total recurrence rates had dropped significantly from the 1980s (22% to 5.5%), all of the recurrences found were in Māori and Pacific people.\textsuperscript{16,17} It is therefore not surprising that Māori and Pacific people have much higher rates of carditis, RHD and consequent heart failure, as the risk of these complications increases with each attack of ARF.

It has not been proven that Māori and Pacific people have increased genetic susceptibility to rheumatic fever. It is more likely that the over-representation of these sectors of the population reflects a combination of overcrowded conditions, socio-economic deprivation, an increased incidence of upper respiratory infections with GAS, and different treatment options or opportunities for appropriate and effective health care.\textsuperscript{15,19,22}
**Cost to New Zealand**

There are significant personal, community and national costs associated with ARF and RHD. These result from repeated and prolonged hospitalisation, the resources required for medical prophylaxis and treatment, surgical intervention, negative physical and psychological experience, disruption of the lives of cases and their families and often premature death. In 1991, it was estimated that the total cost of ARF and RHD to the Auckland health service alone was $3.6 million, with chronic RHD accounting for 71% of the costs. Costs involved were the direct costs of GP and outpatient visits, prescription charges, travel, radiology and the costs of informal care given by household members. In addition to these direct costs, there are a number of indirect costs of ARF and RHD, which are often difficult to measure. These include not only the loss of quantity of life (it has been estimated that five to ten young people die each year as a direct result of ARF or RHD), but also the loss of quality of life. This occurs through time away from education and occupation, impacts on physical development and family relationships, psychological effects and the loss of ability for children and young adults to realise their full potential.

**Population projections**

Currently Maori and Pacific people in New Zealand make up a sizeable percentage of the childhood population. In 2001, approximately 37% of Maori and 40% of Pacific people in New Zealand were under the age of 15 (compared to 23% European). The median age of Europeans was 36.8 years, while for the Maori and Pacific ethnic groups the comparable figures were 21.9 and 21.0 years respectively. It is reasonable to predict that the New Zealand population in the future will represent high growth and a sustained youthful age structure in the Maori and Pacific populations with many (particularly children) living in poor socio-economic circumstance. All these features have significant implications for ARF incidence, prevalence and prevention.

**Prevention of ARF and RHD**

**Primary prevention**

In the future, a cost-effective vaccine for group A streptococci may be the ideal solution for the primary prevention of ARF. Scientific problems have so far prevented the development of such a vaccine, and currently prevention of an initial attack of ARF requires the prompt and accurate diagnosis and adequate antibiotic treatment of GAS throat infections. ARF can be prevented if the preceding throat infection is treated in a timely and effective way. Recommended treatment of streptococcal throat infection is intramuscular (IM) benzathine penicillin or a ten-day course of oral phenoxy methyl penicillin, both of which eradicate the streptococci from the pharynx. The oral treatment is often used because it is safe, inexpensive and less painful.

**Secondary prevention**

Over the last 30 years one of the major successes in ARF management has been the marked decline in recurrent (and often disabling) attacks of rheumatic fever, due to the availability of effective antibiotics for secondary prophylaxis. Secondary prevention of ARF is defined as the continuous administration of antibiotics (usually parenteral benzathine penicillin every 28 days) to cases with previous ARF or well-documented RHD. The aim of secondary prevention is to stop recolonisation or reinfection of the throat with group A streptococci and thereby preventing recurrence of ARF. The risk of ARF after the first attack of group A streptococci is approximately 0.3-3%, but with subsequent infection this risk rises to 25-75%. In addition, those who suffer carditis during their initial attack are significantly more likely to develop further carditis with subsequent streptococcal throat infections. The systematic use of regular antibiotic prophylaxis in known ARF cases has been shown to reduce the incidence of recurrent rheumatic fever, reduce the need for hospitalisation and surgery, decrease the rapidity and severity of RHD and improve quality of life. Furthermore, national prevention programmes based on secondary prevention have the potential for considerable cost savings, and have been found to be a cost-effective method of reducing mortality and morbidity from ARF internationally and in New Zealand.
Diagnosis and Management
4. Diagnosis of Acute Rheumatic Fever (ARF)

Importance of accurate diagnosis

It is important that an accurate diagnosis of ARF is made as:

- over-diagnosis will result in the individual receiving benzathine penicillin G (BPG) injections unnecessarily every four weeks for a minimum of ten years
- under-diagnosis of ARF may lead to the individual suffering a further attack of ARF, cardiac damage and premature death.

The diagnosis of ARF relies on health professionals being aware of the diagnostic features, particularly when presentation is delayed or atypical. In Auckland for example, between 1993 and 1999, four patients diagnosed with septic arthritis by general medicine and orthopaedic physicians, subsequently developed acute rheumatic fever.16,17

Currently, there is no laboratory test diagnostic for ARF, so diagnosis remains a clinical decision. The pre-test probability for diagnosis of ARF varies according to location and ethnicity. For example, in a region with high incidence of ARF (such as the Northern half of the North Island), a person with fever and arthritis is more likely to have ARF than one in a low incidence region (such as the South Island). Māori and Pacific people are also more likely than non-Māori and Pacific people to have ARF.

Current approaches to diagnosis

The Jones criteria for the diagnosis of ARF were introduced in 1944.36 The criteria divide the clinical features of ARF into major and minor manifestations, based on their prevalence and specificity. Major manifestations are those that make the diagnosis more likely, whereas minor manifestations are considered to be suggestive, but insufficient on their own, for a diagnosis of ARF. The exception to this is in the diagnosis of recurrent ARF.

The Jones criteria have been periodically modified and updated. The 1992 update is currently the most widely used and quoted version.37

There are important circumstances where ARF can be diagnosed without strictly adhering to the Jones criteria and these include:

- chorea as the only manifestation of ARF
- indolent carditis (carditis of insidious onset and slow progression) as the only manifestation of ARF.37

Both these types of patients may have insufficient supporting historical, clinical or laboratory findings to fulfil the Jones criteria.

The 1992 Jones criteria are intended only for the initial attack of ARF. Further discussion of the Jones criteria can be found in Appendix B.

Each change to the Jones criteria was made to improve specificity at the expense of sensitivity, largely in response to the falling incidence of ARF in America. As a result, the criteria may not be sensitive enough to pick up disease in high incidence populations, such as Māori and Pacific people. In such populations, the consequences of under-diagnosis are likely to be greater than those of over-diagnosis.

All cases of suspected ARF should be judged against the most recent version of the Jones criteria, but the criteria need not be rigidly adhered to when ARF is the most likely diagnosis.

An expert group convened by the World Health Organisation (WHO) has recently provided additional guidelines as to how the Jones criteria should be applied in primary and recurrent episodes.38

The main modification made to the Jones 1992 criteria for these New Zealand guidelines is the acceptance of echocardiographic evidence of carditis as a major manifestation. In addition there is greater emphasis that monoarthritis may be a presenting feature if there is a history of non-steroidal anti-inflammatory drug (NSAID) use that is likely to have aborted classical ARF migratory polyarthritis.
Categories of definite, probable and possible ARF can be determined by the application of the New Zealand criteria to each case (Table 2).

### Table 2. New Zealand Guidelines for the Diagnosis of ARF

<table>
<thead>
<tr>
<th>DIAGNOSTIC REQUIREMENTS</th>
<th>CATEGORY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial episode of ARF</td>
<td></td>
</tr>
<tr>
<td>2 major or 1 major and 2 minor manifestations plus evidence of a preceding GAS infection*</td>
<td>Definite ARF</td>
</tr>
<tr>
<td>Initial episode of ARF</td>
<td></td>
</tr>
<tr>
<td>1 major and 2 minor with the inclusion of evidence of a preceding GAS infection* as a minor manifestation (Jones, 1956)</td>
<td>Probable ARF</td>
</tr>
<tr>
<td>Initial episode of ARF</td>
<td></td>
</tr>
<tr>
<td>Strong clinical suspicion of ARF, but insufficient signs and symptoms to fulfill diagnosis of definite or probable ARF</td>
<td>Possible ARF</td>
</tr>
<tr>
<td>Recurrent attack of ARF in a case with known past ARF or RHD</td>
<td></td>
</tr>
<tr>
<td>2 major or 1 major and 2 minor or several** minor plus evidence of a preceding GAS infection* (Jones, 1992)</td>
<td></td>
</tr>
<tr>
<td>Major manifestations modified*** from Jones 1992 (see Table 3 for key points in identifying major manifestations)</td>
<td></td>
</tr>
<tr>
<td>Carditis (including evidence of subclinical rheumatic valve disease on echocardiogram*)</td>
<td></td>
</tr>
<tr>
<td>Polyarthritis (or aspecific monoarthritis with history of NSAID use)</td>
<td></td>
</tr>
<tr>
<td>Chorea (can be stand-alone for ARF diagnosis)</td>
<td></td>
</tr>
<tr>
<td>Erythema marginatum</td>
<td></td>
</tr>
<tr>
<td>Subcutaneous nodules</td>
<td></td>
</tr>
<tr>
<td>Minor manifestations (see Table 4 for key points in identifying minor manifestations)</td>
<td></td>
</tr>
<tr>
<td>Fever</td>
<td></td>
</tr>
<tr>
<td>Raised ESR or CRP</td>
<td></td>
</tr>
<tr>
<td>Polyarthralgia§</td>
<td></td>
</tr>
<tr>
<td>Prolonged P-R interval on ECG.</td>
<td></td>
</tr>
</tbody>
</table>

All categories assume that other more likely diagnoses have been excluded. Please see additional tables for details about specific manifestations. CRP = C-reactive protein; ECG = electrocardiogram; ESR = erythrocyte sedimentation rate; GAS = group A streptococcus; RHD = rheumatic heart disease

* Elevated or rising antistreptolysin O or other streptococcal antibody (Table 5), is sufficient for a diagnosis of definite ARF. A positive throat culture or rapid antigen test for GAS alone is less secure as 50% of those with a positive throat culture will be carriers only. Therefore, a positive culture alone demotes a case to probable or possible ARF

** Most cases of recurrence fulfill the Jones criteria. However in some cases (such as new carditis on previous RHD) it may not be clear. Therefore in order to avoid under-diagnosis, a presumptive diagnosis of rheumatic recurrence may be made where there are several minor manifestations and evidence of a preceding GAS infection in a person with a reliable history of previous ARF or established RHD. In addition, WHO (2004) recommendations state that where there is established RHD, a recurrent attack can be diagnosed by the presence of two minor manifestations plus evidence of a preceding group A streptococcal infection

*** Acceptance of echocardiographic evidence of carditis as a major criterion is a modification to the Jones (1992) update

§ When carditis is present as a major manifestation (clinical and/or echocardiographic), a prolonged P-R interval cannot be considered an additional minor manifestation in the same person.

§§ Other causes of arthritis/arthralgia should be carefully excluded, particularly in the case of monoarthritis e.g. septic arthritis (including disseminated gonococcal infection), infective or reactive arthritis and auto-immune arthropathy (e.g. juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis and sarcoidosis. Note that if polyarthritis is present as a major manifestation, polyarthralgia cannot be considered an additional minor manifestation in the same person.

Special consideration should be given to high-risk population groups such as Māori and Pacific people, and those residing in poor socio-economic circumstances. In these cases, it may be important to err on the side of diagnosis and prophylaxis.
Patients who do not fulfi l these criteria, but in whom the clinician remains suspicious that the diagnosis may be ARF, should be maintained on oral penicillin and reviewed in two to four weeks with a repeat echocardiogram to detect the appearance of new lesions.40,41 If there is evidence of rheumatic valve disease clinically or on echocardiogram, the diagnosis is confi rmed and long-term secondary prophylaxis can be commenced. If there is no evidence of carditis and no alternative diagnosis has been found then ARF may be the diagnosis by exclusion. Those with epidemiological risk factors (Māori, Pacifi c and low socio-economic status) should be commenced on secondary prophylaxis with due consideration of an alternative diagnosis (such as rheumatological) and the need for ongoing review.

Clinical features of acute rheumatic fever - major manifestations

The major manifestations of ARF and features for their diagnosis are presented in Table 3.

Table 3. Major Manifestations of ARF

<table>
<thead>
<tr>
<th>MAJOR MANIFESTATION</th>
<th>POINTS FOR DIAGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arthritis*</td>
<td>• Most common presenting symptom of ARF (occurring in up to 75% of fi rst attacks)</td>
</tr>
<tr>
<td></td>
<td>• Classified as swelling of the joint in the presence of two or more of the following: limitation of movement, hotness of the joint and pain in the joint and/or tenderness. Typically, the arthritis of ARF is extremely painful</td>
</tr>
<tr>
<td></td>
<td>• Large joints are usually affected, especially knees and ankles</td>
</tr>
<tr>
<td></td>
<td>• Polyarthritis is usually asymmetrical and migratory (one joint becoming inflamed as another subsides) but can be additive (multiple joints progressively becoming inflamed without warning)</td>
</tr>
<tr>
<td></td>
<td>• Highly responsive to salicylate and NSAID therapy - usually responds within 3 days</td>
</tr>
<tr>
<td></td>
<td>• Monoarthritis may be a presenting feature if there is a history of NSAID use early in the course of the illness (prematurely aborting the manifestation of polyarthritis). ** This diagnosis is best made by a physician experienced in ARF</td>
</tr>
<tr>
<td></td>
<td>• The diagnosis of arthritis of the hip is accepted by history of pain precluding weight bearing and/or limitation of movement on examination</td>
</tr>
<tr>
<td></td>
<td>• In order to satisfy polyarthritis as a manifestation, at least one joint should have been observed in a clinical setting accompanied by a defi nite history of arthritis in other joints (Grade D)</td>
</tr>
<tr>
<td>Carditis</td>
<td>• Valvulitis usually presents clinically as an apical holosystolic murmur with or without a mid-diastolic fi ow murmur (Carey-Coombs murmur) or an early diastolic murmur at the base of the heart (aortic regurgitation)</td>
</tr>
<tr>
<td></td>
<td>• Although pericarditis and myocarditis may occur, cardiac infl ammation in ARF almost always affects the valves, especially the mitral and aortic valves**30,34</td>
</tr>
<tr>
<td></td>
<td>• Early disease leads to valvular regurgitation, whereas prolonged or recurrent disease may lead to increased valvular regurgitation or stenotic lesions31</td>
</tr>
<tr>
<td></td>
<td>• The rheumatic aetiology can usually be confi rmed by a typical appearance on echocardiography (see Tables 6 and 7)</td>
</tr>
<tr>
<td></td>
<td>• In New Zealand, echocardiographic evidence of subclinical carditis can also be accepted as a major manifestation</td>
</tr>
<tr>
<td></td>
<td>• Congestive heart failure in ARF results from valvular dysfunction secondary to valvulitis and is not due to primary myocarditis30</td>
</tr>
<tr>
<td></td>
<td>• The natural history of valve regurgitation is a 25-50% improvement by one year30</td>
</tr>
<tr>
<td></td>
<td>• If pericarditis is present, the friction rub may obscure valvular murmurs.</td>
</tr>
</tbody>
</table>
**Sydenham’s chorea**
- Consists of jerky, uncoordinated movements, especially affecting the hands, feet, tongue and face. The movements disappear during sleep. They may affect one side only (hemichorea).
- Useful signs include:
  - the “milkmaid’s grip” (rhythmic squeezing when the patient grasps the examiner’s fingers)
  - “spooning” (flexion of the wrists and extension of the fingers when the hands are extended)
  - the “pronator sign” (turning outwards of the arms and palms when held above the head)
  - inability to maintain protrusion of the tongue
- This manifestation affects females predominantly, particularly in adolescence.
- Chorea may occur after a prolonged latent period following GAS infection, therefore no additional manifestations (including raised antibody titres) are required in order to make a diagnosis of ARF.
- Chorea has a strong association with carditis, hence echocardiography is essential for assessment of all patients with chorea, regardless of the presence of cardiac murmurs (Level IV, Grade C). A finding of subclinical carditis by echo will further support the diagnosis of chorea as a manifestation of ARF (Grade D). Even in the absence of echocardiographic evidence of carditis, patients with chorea should be considered at risk of subsequent cardiac damage. Therefore, they should all receive secondary prophylaxis, and be carefully followed up for subsequent development of RHD.
- Chorea is the ARF manifestation most likely to recur and is often associated with pregnancy or oral contraceptive use. The vast majority of cases resolve within 6 months (usually within 6 weeks) although rare cases lasting as long as 3 years have been documented.

**Subcutaneous nodules**
- Rare (less than 2% of cases) but highly specific manifestation of ARF.
- They are 0.5-2.0 cm in diameter, round, firm, freely mobile and painless nodules that occur in crops of up to 12 over the elbows, wrists, knees, ankles, Achilles tendon, occiput and posterior spinal processes of vertebrae.
- Tend to appear 1-2 weeks after the onset of other symptoms, last only 1-2 weeks (rarely more than 1 month).
- Strongly associated with carditis.
- Subcutaneous nodules are rarely seen as the sole major criterion in ARF and should be accompanied by additional major criteria in order to make the diagnosis.

**Erythema marginatum**
- Rare as well as difficult to detect (especially in dark-skinned people).
- Occurs as circular patterns of bright pink macules or papules that blanch under pressure and spread outwards in a circular or serpiginous pattern on the trunk and proximal extremities (almost never on face). The rash may be more apparent after showering.
- Not itchy or painful and not affected by anti-inflammatory medication.
- May recur for weeks or months, despite resolution of the other features of ARF.
- Erythema marginatum is rarely seen as the sole major criterion in ARF and should be accompanied by additional major criteria in order to make the diagnosis.

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* ARF should always be considered in the differential diagnosis of patients presenting with arthritis in high-risk populations. In the hospital setting, physicians and surgeons should collaborate when the diagnosis of arthritis is unclear. Patients with sterile joint aspirates in the absence of previous antibiotic exposure should never be treated speculatively for septic arthritis without further investigation, particularly in areas with high ARF/RHD prevalence.

** Note that in New Zealand, NSAIDs are now readily available over the counter and have therefore often been used prior to presentation.

*** During recent outbreaks of ARF in the USA, up to 71% of patients with chorea had carditis. Even though clinically evident carditis increases the risk of later development of RHD, prior to cardiac echocardiography approximately 25% of patients with “pure” chorea also eventually developed RHD. This is explained by the finding that over 50% of patients with chorea, but without cardiac murmurs, have echocardiographic evidence of mitral regurgitation.
Clinical features of acute rheumatic fever - minor manifestations

The minor manifestations of ARF and features for their diagnosis are presented in Table 4.

Table 4. Minor Manifestations of ARF

<table>
<thead>
<tr>
<th>MINOR MANIFESTATION</th>
<th>POINTS FOR DIAGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arthralgia</td>
<td>• May suggest ARF if the arthralgia occurs in the same pattern as rheumatic polyarthritis (migratory, asymmetrical and affecting large joints)</td>
</tr>
<tr>
<td></td>
<td>• If polyarthritis is present as a major manifestation, polyarthralgia cannot be considered an additional minor manifestation in the same person</td>
</tr>
<tr>
<td></td>
<td>• Alternative diagnoses (as suggested in Table 8) should be considered in a patient with arthralgia that is not typical of ARF</td>
</tr>
<tr>
<td>Fever</td>
<td>• Most manifestations of ARF are accompanied by fever (with the exception of chorea)</td>
</tr>
<tr>
<td></td>
<td>• In New Zealand, an oral, tympanic or rectal temperature greater than or equal to 38°C on admission, or documented during the current illness, should be considered as fever (Level IV, Grade C)</td>
</tr>
<tr>
<td></td>
<td>• Fever, like arthritis and arthralgia, is usually quickly responsive to salicylate/NSAID therapy</td>
</tr>
<tr>
<td>Elevated acute phase reactants</td>
<td>• In New Zealand, a serum CRP level of ≥30mg/L or ESR of ≥50mm/h meets this diagnostic criterion (Grade D)</td>
</tr>
<tr>
<td></td>
<td>• The ESR in ARF is typically &gt;90mm/hr, usually remains elevated for &gt;4 weeks, and may remain elevated for 3-6 months despite a much shorter duration of symptoms</td>
</tr>
<tr>
<td></td>
<td>• The serum CRP concentration rises more rapidly than the ESR and also falls more rapidly with resolution of the attack</td>
</tr>
<tr>
<td>Prolonged P-R interval</td>
<td>• An electrocardiogram (ECG) should be performed in all cases of suspected ARF (Level IV, Grade C)</td>
</tr>
<tr>
<td></td>
<td>• The P-R interval increases normally with age therefore needs to be age-adjusted. The following upper limits of normal are used in New Zealand:*</td>
</tr>
<tr>
<td></td>
<td>• Age 3-12 years: 0.16 seconds</td>
</tr>
<tr>
<td></td>
<td>• Age 12-16 years: 0.18 seconds</td>
</tr>
<tr>
<td></td>
<td>• Age 17+ years: 0.20 seconds</td>
</tr>
<tr>
<td></td>
<td>• A prolonged P-R interval is occasionally a normal variant, but one that resolves over the ensuing days to weeks may be a useful diagnostic feature of ARF in cases where the clinical features are not definitive.** In these cases, a repeat ECG after 1-2 months may be useful</td>
</tr>
<tr>
<td></td>
<td>• Extreme first degree block sometimes leads to a junctional rhythm, usually with a heart rate similar to the sinus rate</td>
</tr>
<tr>
<td></td>
<td>• Second degree, and even complete heart block, can occur and, if associated with a slow ventricular rate, may give the false impression that carditis is not significant</td>
</tr>
<tr>
<td></td>
<td>• In the absence of clinical or echocardiographic carditis, a second or third degree block accompanied by other manifestations of ARF is highly supportive of the diagnosis (Grade D)</td>
</tr>
<tr>
<td></td>
<td>• When carditis is present as a major manifestation (clinical and/or echocardiographic), prolonged P-R interval cannot be considered an additional minor manifestation in the same person.</td>
</tr>
</tbody>
</table>

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* Adapted from Park M K. p42.
** In a recent resurgence of ARF in the USA, 32% of patients had abnormal AV conduction, usually a prolonged P-R interval. A small proportion had more severe conduction abnormalities, which were sometimes found by auscultation or echocardiography in the absence of evidence of valvulitis.
Evidence of a preceding group A streptococcal infection

GAS are isolated from throat swabs in less than ten percent of ARF cases in New Zealand and less than five percent of cases in Aboriginal Australians. This latter figure may be a result of later presentation of ARF, as 28% of Aboriginal Australians have been found to present as chorea compared to six percent of ARF cases in Auckland (1993–1999). A positive culture without supportive antibody elevation may be carriage in up to 50% of cases. Streptococcal antibody titres are therefore crucial in confirming the diagnosis. The most commonly used tests are the plasma antistreptolysin O (ASO) and the antideoxyribonuclease B (anti-DNase B) titres. The serum ASO titre reaches a maximum at about three to six weeks after infection and the serum anti-DNase B titre can take up to six to eight weeks to reach a maximum. The rate of decline of these antibodies varies enormously, with the ASO titre starting to fall six to eight weeks and the anti-DNase B titre three months after infection. In the absence of reinfection, the ASO titre usually approaches pre-infection levels after six to 12 months, whereas the anti-DNase B titre tends to remain elevated for longer. The reference range for these antibody titres varies with age and geographical location. In a population with a high rate of streptococcal infections, many children will have high background streptococcal titres. The upper limit of normal approach attempts to determine a raised titre over and above this background, and therefore select out those children who have had a recent streptococcal infection. In New Zealand, an ASO titre of greater than or equal to 480 and/or an anti-DNase B titre of greater than or equal to 680 is accepted as significant (Grade D) Table 5.

Table 5. Upper Limits of Normal for Serum Streptococcal Antibody Titres Used in New Zealand for ARF Diagnosis

<table>
<thead>
<tr>
<th>ANTIBODY TEST</th>
<th>TITRE (Iu/ML)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASO (anti-streptolysin O)</td>
<td>≥480</td>
</tr>
<tr>
<td>Anti-DNase B</td>
<td>≥680</td>
</tr>
</tbody>
</table>

Established from residual sera from children (under 15 years) hospitalised in Auckland in 1982. Lower levels may be acceptable in the very young or those over the age of 15 years. A two-tube (two-fold) rise or fall in antibody titres after 10-14 days would also be diagnostic. Note that evidence of a preceding GAS infection is not necessary for the diagnosis of chorea as ARF.

All cases of suspected ARF (chorea is an exception) should have elevated serum streptococcal serology demonstrated. If the initial titre is below the upper limit of normal, testing should be repeated 10 to 14 days later (Grade D).

Other less common clinical features

These include epistaxis, abdominal pain, rheumatic pneumonia (pulmonary infiltrates in patients with acute carditis), mild elevations of plasma transaminase levels and microscopic haematuria, pyuria or proteinuria. None is specific for ARF but epistaxis and abdominal pain occur commonly.

Echocardiography

Prior to the introduction of echocardiography, the diagnosis of rheumatic carditis relied on clinical evidence of valvulitis or pericarditis, supported by radiographic evidence of cardiomegaly. Today, all patients with suspected or definite ARF should undergo echocardiography, if possible, to identify evidence of carditis (Grade C).

In New Zealand, echocardiography facilities are readily available in the larger centers for populations at high-risk of ARF. The use of echocardiography as a major criterion for ARF diagnosis requires expert interpretation adhering to echocardiographic diagnostic standards. These standards concur with recent WHO echocardiographic criteria for ARF and are summarised in Table 6 (Level IV). These criteria can readily distinguish a small colour jet of physiological regurgitation in a normal child from pathological regurgitation in a child with RHD.
Table 6. Minimal Echocardiographic Criteria to Allow a Diagnosis of Pathological Valvular Regurgitation

### AORTIC REGURGITATION

- **Colour:**
  Substantial colour jet seen in 2 planes extending greater than or equal to 1 cm beyond the valve leaflets
- **Continuous wave or pulsed Doppler:**
  Holodiastolic with well-defined high velocity spectral envelope

### MITRAL REGURGITATION

- **Colour:**
  Substantial colour jet seen in 2 planes extending greater than or equal to 2 cm beyond the valve leaflets
- **Continuous wave or pulsed Doppler:**
  Holosystolic with well-defined high velocity spectral envelope

If the aetiology of aortic or mitral regurgitation on Doppler echocardiography is not clear, the following features support a diagnosis of rheumatic valve damage:

- Both mitral and aortic valves have pathological regurgitation
- The mitral regurgitant jet is directed posteriorly, as anterior mitral valve prolapse is more common than posterior valve prolapse
- Multiple jets of mitral regurgitation
- The presence of morphological or anatomical changes consistent with RHD (see text), but excluding slight thickening of valve leaflets:
  - Definite thickening of mitral valve leaflets, indicative of chronic RHD*
  - Elbow or dog leg deformity** of anterior mitral valve leaflet.

**Source:** Adapted with permission from Wilson N J & Neutze J M. These criteria further evolved as part of the development of the Australian guidelines on rheumatic fever diagnosis and the WHO working groups on echocardiography.

* Echocardiography allows the operator to comment on the appearance of valves that are affected by rheumatic inflammation. The degree of thickening gives some insight into the duration of valvulitis, no significant thickening occurs in the first weeks of acute rheumatic carditis (Level IV)

** Only after several months is immobility of the subchordal apparatus and posterior leaflet observed. Several other findings have also been reported, including acute nodules, seen as a beaded appearance of the mitral valve leaflets. Although none of these morphological features is unique to ARF, the experienced echocardiographic operator can use their presence as supportive evidence of a rheumatic aetiology of valvulitis.

In New Zealand, ARF carditis is classified mild, moderate or severe (Table 7) and these categories are used to guide the duration of secondary prophylaxis (see Section 7 and Table 13).
Table 7. Severity of ARF Carditis

**MILD CARDITIS**
- Mild mitral or aortic regurgitation clinically and/or on echo (fulfilling the minimal echo standards in Table 6) with no clinical evidence of heart failure and no evidence of cardiac chamber enlargement on CXR, ECG or echocardiography

**MODERATE CARDITIS**
- Any valve lesion of moderate severity clinically (e.g. mild or moderate cardiomegaly), or
- Any echocardiographic evidence of cardiac chamber enlargement or
- Any moderate severity valve lesion on echo
  - Mitral regurgitation is considered moderate if there is a broad high-intensity proximal jet filling half the left atrium or a lesser volume high-intensity jet producing prominent blunting of pulmonary venous inflow
  - Aortic regurgitation is considered moderate if the diameter of the regurgitant jet is 15% to 30% of the diameter of the left ventricular outflow tract with flow reversal in upper descending aorta

**SEVERE CARDITIS**
- Any impending or previous cardiac surgery for RHD, or
- Any severe valve lesion clinically (significant cardiomegaly expected, and/or heart failure), or
- Any severe valve lesion on echo:
  - Abnormal regurgitant colour and Doppler flow patterns in pulmonary veins are a prerequisite for severe mitral regurgitation
  - Doppler reversal in lower descending aorta is required for severe aortic regurgitation

* Valvular regurgitation is usually relatively mild in the absence of pre-existing disease; in first episodes of ARF, severe mitral and aortic regurgitation occurred in less than 10% of patients in New Zealand

** When there is both mitral and aortic regurgitation, one must be moderate by echo criteria in order for the carditis to be classified of moderate severity.

Tricuspid and pulmonary regurgitation graded mild or greater may be seen in people with normal hearts who have fever, volume overload or pulmonary hypertension. For this reason a diagnosis of carditis should not be based on right-side regurgitation alone. Although pulmonary and tricuspid regurgitation are often seen in association with left-sided lesions in ARF, pressure and volume overload must be excluded before attributing even moderate tricuspid regurgitation to valvulitis. If both left and right-sided lesions coexist in ARF carditis, then the predominant influence for diagnosis is the severity of the left-sided lesion.

If valvulitis is not found at presentation, it may appear within two weeks, or occasionally within one month but no longer. Thus an equivocal initial echocardiograph should be followed up in two to four weeks if the findings would alter the diagnosis of ARF.

Usually it is not possible to confidently distinguish between acute carditis and pre-existing rheumatic valve disease by echocardiography. In a patient with known previous RHD, the diagnosis of acute carditis during a recurrence of ARF relies on accurate documentation of the cardiac findings before the recurrence, so that new clinical or echocardiographic features can be confirmed. But, in a patient with no prior history of ARF or RHD, it makes little difference whether echocardiographic changes are new or old.

Further details on the use of echo in ARF can be found in Appendix C.
Differential diagnosis

Many of the clinical features of ARF are non-specific, so a wide range of differential diagnoses should be considered as shown in Table 8.\textsuperscript{32,67}

Table 8. Differential Diagnoses of Common Major Manifestations of ARF

<table>
<thead>
<tr>
<th>POLYARTHRITIS AND FEVER</th>
<th>CARDITIS</th>
<th>CHOREA</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Other infections(^*) (including gonococcal)</td>
<td>• Innocent murmur</td>
<td>• Systemic lupus erythematosus</td>
</tr>
<tr>
<td>• Connective tissue and other auto-immune disease(^**)</td>
<td>• Mitral valve prolapse</td>
<td>• Drug ingestion (extrapyramidal syndrome)(^*)</td>
</tr>
<tr>
<td>• Reactive arthropathy</td>
<td>• Congenital heart disease</td>
<td>• Wilson’s disease (usually adult onset)</td>
</tr>
<tr>
<td>• Sickle cell anaemia</td>
<td>• Infective endocarditis</td>
<td>• Tic disorder(^#)</td>
</tr>
<tr>
<td>• Infective endocarditis</td>
<td>• Hypertrophic cardiomyopathy</td>
<td>• Congenital, e.g. hyperbilirubinaemia</td>
</tr>
<tr>
<td>• Leukaemia or lymphoma</td>
<td>• Myocarditis — viral or idiopathic</td>
<td>• Choreoathetoid cerebral palsy</td>
</tr>
<tr>
<td>• Gout and pseudogout</td>
<td>• Pericarditis — viral or idiopathic</td>
<td>• Encephalitis</td>
</tr>
<tr>
<td>• Henoch-Schönlein purpura</td>
<td></td>
<td>• Familial chorea (including Huntington’s)</td>
</tr>
<tr>
<td>• Post-streptococcal reactive arthritis(^***)</td>
<td></td>
<td>• Intracranial tumour</td>
</tr>
<tr>
<td>• Other, e.g. HIV/AIDS, leukaemia</td>
<td></td>
<td>• Hormonal(^\§)</td>
</tr>
</tbody>
</table>

Source: Adapted from Lennon D. 2004,\textsuperscript{32} and Carapetis J et al. 2005.\textsuperscript{67}

\(^*\) Includes bacterial arthritis (e.g. Staphylococcus aureus, Neisseria gonorrhoea), influenza b, cytomegalovirus, Epstein-Barr Virus, mycoplasma, rubella (also post-vaccination), hepatitis B, parvovirus, Yersinia spp and other gastrointestinal pathogens

\(^**\) Includes rheumatoid arthritis, juvenile chronic arthritis, inflammatory bowel disease, systemic lupus erythematosus, systemic vasculitis, sarcoidosis and others

\(^***\) Some patients present with arthritis not typical of ARF, but with evidence of recent streptococcal infection and are said to have post-streptococcal reactive arthritis. In these cases the arthritis may affect joints that are not commonly affected in ARF (such as the small joints of the hand), and is less responsive to anti-inflammatory treatment. These patients are said not to be at risk of carditis, and therefore do not require secondary prophylaxis. However, some patients diagnosed with post-streptococcal reactive arthritis have developed later episodes of ARF, indicating that the initial diagnosis should have been atypical ARF (Level IV)\textsuperscript{68,69}

It is recommended that the diagnosis of post-streptococcal reactive arthritis should rarely, if ever, be made in high-risk populations, and with caution in low-risk populations (Grade C). Patients so diagnosed should receive secondary prophylaxis for at least 5 years (Grade D). Echocardiography (see algorithm 2) should be used to confirm the absence of valvular damage in all of these cases before discontinuing secondary prophylaxis (Grade D)

\(^#\) Drugs and toxins include anticonvulsants, antidepressants, lithium, scopolamine, calcium channel blockers, methylphenidate, theophylline and antihistamines

\(^\§\) Some cases of chorea are mild or atypical and may be confused with motor tics or the involuntary jerks of Tourette’s syndrome. There may therefore be confusion between Sydenham’s chorea and these conditions. The term PANDAS (Pediatric Auto-immune Neuropsychiatric Disorder Associated with Streptococcal infection) refers to a subgroup of children with tic or obsessive-compulsive disorders (OCD), whose symptoms may develop or worsen following GAS infection.

Five criteria have been used to define the PANDAS subgroup:\textsuperscript{70,71}

- The presence of a Tic disorder and/or OCD
- Pre-pubertal age of onset (usually between 3 and 12 years of age)
- Abrupt symptom onset and/or episodic course of symptom severity
- Temporal association between symptom exacerbations and streptococcal infection (approx 7-14 days)
- Presence of neurologic abnormalities during periods of symptom exacerbation (typically adventitious movements or motoric hyperactivity)
Investigations

The recommended investigations in ARF are listed in Table 9. Other investigations may be appropriate depending on the clinical picture and potential differential diagnoses.

Table 9. Investigations in Suspected ARF

<table>
<thead>
<tr>
<th>RECOMMENDED FOR ALL CASES</th>
</tr>
</thead>
<tbody>
<tr>
<td>• White blood cell count</td>
</tr>
<tr>
<td>• Erythrocyte sedimentation rate (repeat weekly once diagnosis confirmed)</td>
</tr>
<tr>
<td>• C-reactive protein</td>
</tr>
<tr>
<td>• Blood cultures if febrile</td>
</tr>
<tr>
<td>• Electrocardiogram (repeat as necessary if conduction abnormality more than first degree)</td>
</tr>
<tr>
<td>• Chest x-ray if clinical or echocardiographic evidence of carditis</td>
</tr>
<tr>
<td>• Echocardiogram (repeat as necessary in 2-4 weeks if equivocal or if serious carditis)</td>
</tr>
<tr>
<td>• Throat swab (preferably before giving antibiotics) — culture for group A streptococcus</td>
</tr>
<tr>
<td>• Anti-streptococcal serology: both anti-streptolysin O and anti-DNase B titres, if available (repeat 10-14 days later if 1st test not confirmatory)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>TESTS FOR ALTERNATIVE DIAGNOSES, DEPENDING ON CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Serology and auto-immune markers for auto-immune or reactive arthritis (including ANA - Anti Nuclear Antibody)</td>
</tr>
<tr>
<td>• Repeated blood cultures if possible endocarditis or septic arthritis</td>
</tr>
<tr>
<td>• Joint aspirate (microscopy and culture) for possible septic arthritis*</td>
</tr>
<tr>
<td>• Joint X-ray</td>
</tr>
<tr>
<td>• Copper, caeruloplasmin, anti-nuclear antibody, drug screen, and consider CT/MRI head for choreiform movements.**</td>
</tr>
</tbody>
</table>

* Typically, the synovial fluid in joints affected by ARF contains 10,000 to 100,000 white blood cells/mm³ (predominantly neutrophils). The protein concentration is approximately 4g/dL, glucose levels are normal, gram stain negative and a good mucin clot is present.

** The chorea of ARF can be readily diagnosed on the basis of history, physical examination and laboratory evaluation. Neuroimaging is seldom necessary and should be reserved for cases who have an atypical presentation such as hemichorea.
5. Management of ARF

The major priority in the first few days after presentation in ARF is confirmation of the diagnosis. Except in the case of heart failure management, none of the treatments offered to cases with ARF have been proven to alter the outcome of the acute episode or the amount of damage to heart valves.74,75 Thus, there is no urgency to begin definitive treatment. The priorities in managing ARF are outlined in Table 10.

Table 10. Priorities in Managing Acute Rheumatic Fever

<table>
<thead>
<tr>
<th>ADMISSION TO HOSPITAL*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ideally, all those with suspected ARF (first episode or recurrence) should be hospitalised as soon as possible after onset of symptoms (Grade D). This ensures that all investigations are performed and, if necessary, observations completed for a period prior to commencing treatment to confirm the diagnosis. Hospitalisation also provides an ideal opportunity for education</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>CONFIRMATION OF THE DIAGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Observation prior to anti-inflammatory treatment (paracetamol [1st line] for fever or joint pain)</td>
</tr>
<tr>
<td>Investigations (as per Table 9)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>All cases</td>
</tr>
</tbody>
</table>

**Antibiotics***

- Oral penicillin V (250mg twice daily) should be commenced in all cases while the diagnosis is being established. To reliably eradicate GAS, oral penicillin should be given for the full 10 days
- Oral erythromycin used in cases with reliably documented penicillin allergy.*** (10 days of erythromycin ethylsuccinate (EES) 40mg/kg per day in 2-4 divided doses, maximum 1g/day in children or 400mg twice daily in adolescents and adults).76 EES is currently the only fully subsidised oral erythromycin in New Zealand
- Intravenous antibiotics are not indicated. Roxithromycin is not recommended because of the limited available evidence that it is not as effective as erythromycin in eradicating GAS from the upper respiratory tract.77
- The first dose of intramuscular benzathine penicillin G (BPG 1,200,000 U or 600,000 U if less than 20kg) should also be given in hospital in association with education about the importance of secondary prophylaxis. Once the first dose of BPG is given, the oral penicillin is stopped

<table>
<thead>
<tr>
<th>Arthritis/arthralgia</th>
</tr>
</thead>
</table>

**Salicylates/NSAIDS**

- The arthritis of ARF has been shown in controlled trials to respond dramatically to salicylates and has also been noted to respond to other NSAID therapy.78,79,80 often within hours and almost always within 3 days (Level I)
- Salicylates are recommended as first line treatment because of the extensive experience with their use in ARF.78,79,80 They should be commenced in cases with arthritis or severe arthralgia as soon as the diagnosis of ARF has been confirmed (Grade B), but they should be withheld if the diagnosis is not certain. In such cases, paracetamol or codeine should be used instead for pain relief
- Aspirin should be started at a dose of 60-100mg/kg/day (4-8g/day in adults) in 4-5 divided doses. If there is an incomplete response within 2 weeks, the dose may be increased to 125mg/kg/day, but with these higher doses careful observation for features of salicylate toxicity is advised. If facilities are available, blood levels may be monitored every few days, and the dose increased until serum levels of 220-330mg/100dl are reached. Toxic effects (tinnitus, headache, hyperpnoea) are likely above 200mg/100dl, but often resolve after a few days
- Most cases require 10 days or less of aspirin therapy and therefore blood level monitoring is seldom necessary. Many need aspirin for only 1-2 weeks, although some need it for up to 6 weeks. In such cases, the dose can often be reduced to 60-70mg/kg/day
TREATMENT CONTINUED

after the initial 1-2 weeks.\textsuperscript{52} As the dose is reduced, or within 3 weeks of discontinuing aspirin, joint symptoms may recur. This does not indicate recurrence, and can be treated with another brief course of high-dose aspirin. Most ARF episodes subside within 6 weeks, and 90% resolve within 12 weeks. Approximately 5% of cases require 6 months or more of salicylate therapy\textsuperscript{52}

- There is also the risk of Reye’s syndrome in children receiving salicylates who develop certain viral infections, particularly influenza. It is recommended that children receiving aspirin during the influenza season (autumn/winter) also receive an influenza vaccine (Grade D)

- Naproxen has been used (10-20mg/kg/day) successfully in those with ARF, including one small randomised trial, and has been advocated as a safer alternative to aspirin (Level III-I).\textsuperscript{83,84} It has the advantage of only twice-daily dosing, less hepatotoxicity, and it is also available in an elixir for young children. The experience with this medication is limited, so the recommendation currently is to restrict it to those intolerant to aspirin, or to use it as a step-down treatment once cases are discharged from hospital (Grade D)

Paracetamol

- Mild arthralgia and fever may respond to paracetamol alone

Fever

Low-grade fever does not require specific treatment. Fever will usually respond dramatically to salicylate therapy. Fever alone, or fever with mild arthralgia or arthritis, may not require salicylates, but can instead be treated with paracetamol

Carditis/heart failure

Bed rest

In the pre-penicillin era, prolonged bed rest in those with rheumatic carditis was associated with shorter duration of carditis, fewer relapses and less cardiomegaly.\textsuperscript{86} Ambulation should be gradual and as tolerated in cases with heart failure, or severe acute valve disease, especially during the first 4 weeks, or until the serum CRP level has normalised and the ESR has normalised or dramatically reduced. Those with milder or no carditis should remain in bed only as long as necessary to manage other symptoms, such as joint pain (Grade D).

A guide for activity levels is shown below (Adapted from Lennon D. 2004\textsuperscript{32}) (Grade D).

<table>
<thead>
<tr>
<th>Activity</th>
<th>Arthritis alone</th>
<th>Mild carditis</th>
<th>Moderate carditis</th>
<th>Severe carditis</th>
</tr>
</thead>
<tbody>
<tr>
<td>In hospital</td>
<td>1-2 weeks</td>
<td>2-3 weeks</td>
<td>4-6 weeks</td>
<td>2-4 months</td>
</tr>
<tr>
<td></td>
<td>Mobilise freely as tolerated</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>House arrest (activity and school work at home)</td>
<td>1-2 weeks after discharge</td>
<td>2-3 weeks</td>
<td>4-6 weeks</td>
<td>2-4 months</td>
</tr>
<tr>
<td>School</td>
<td>2 weeks</td>
<td>2-4 weeks</td>
<td>1-3 months</td>
<td>2-3 months</td>
</tr>
<tr>
<td></td>
<td>Gradual return to full activity</td>
<td></td>
<td></td>
<td>Avoid sport and physical education</td>
</tr>
<tr>
<td>Full activity (sport)</td>
<td>After 6 weeks</td>
<td>After 3 months</td>
<td>After 3-6 months</td>
<td>Variable</td>
</tr>
</tbody>
</table>

Urgent echocardiogram:

An urgent echocardiogram and cardiology assessment are recommended for all cases with heart failure.
Anti-failure medication:

- Diuretics/fluid restriction for mild-moderate failure
- ACE inhibitors for more severe failure, particularly if aortic regurgitation present
- Glucocorticoids\(^\text{§}\) optional for severe carditis\(^{11}\)
- Digoxin if atrial fibrillation present
- There is little experience with beta-blockers in heart failure due to acute carditis, and their use is not recommended\(^{57}\)

Detailed recommendations for the management of heart failure can be found in a separate Heart Foundation clinical guideline (available at http://www.nzgg.org.nz)

Valve surgery

Surgery is usually deferred until active inflammation has subsided. Rarely, valve leaflet or chordae tendinae rupture leads to severe regurgitation which requires emergency surgery. This can be safely performed by experienced surgeons, although the risk appears to be slightly higher than when surgery is performed after active inflammation has resolved.\(^{87}\) Valve replacement, rather than repair, is usually performed during the acute episode, because of the technical difficulties of repairing friable, inflamed tissue. Nevertheless, very experienced surgeons may achieve good results with repair in this situation.

Chorea

Sydenham's chorea is self-limited. Most cases will resolve within weeks and almost all cases within 6 months,\(^{48}\) although rare cases may last as long as 2-3 years.\(^{44,48}\) Mild or moderate chorea does not require any specific treatment, aside from rest and a calm environment. Over-stimulation or stress can exacerbate the symptoms. Sometimes hospitalisation is useful to reduce the stress that families face in dealing with abnormal movements and emotional lability.

Because chorea is benign and self-limiting, and anti-chorea medications are potentially toxic, treatment should only be considered if the movements interfere substantially with normal activities, place the person at risk of injury or are extremely distressing to the patient, family and friends. Aspirin and glucocorticoid therapy do not have a significant effect on rheumatic chorea\(^{47}\)

Small studies of intravenous immunoglobulin (IVIG) have suggested more rapid recovery from chorea, but have not demonstrated reduced incidence of long-term valve disease in non-chorea ARF.\(^{41,91}\) Until more evidence is available, IVIG is not recommended, except for severe chorea refractory to other treatments (Level II / IV, Grade C)

Carbamazepine\(^{2}\) and valproic acid\(^{7}\) are now preferred to haloperidol, which was previously considered the first-line medical treatment for chorea.\(^{92,93}\) A small, prospective comparison of these 3 agents recently concluded that valproic acid was the most effective\(^{94}\)

Other anti-chorea medications should be avoided because of potential toxicity. Because of the small potential for liver toxicity with valproic acid, it is recommended that carbamazepine be used initially for severe chorea requiring treatment, and that valproic acid be considered for refractory cases (Level III 2, Grade C). A response may not be seen for 1-2 weeks, and successful medication may only reduce, but not eliminate, the symptoms. Medication should be continued for 2-4 weeks after chorea has subsided and then withdrawn. Recurrences of chorea are usually mild and can be managed conservatively but, in severe recurrences, the medication can be re-started if necessary.

**CLINICAL FOLLOW-UP**

- All cases should receive regular review and outpatient follow-up should be initiated prior to discharge
- The frequency and duration of review is dependent on the individual clinical needs and local capacity and should become more frequent in the event of symptom onset, symptomatic deterioration or a change in clinical findings
- Particular care should be taken when cases are transferred from paediatric to adult services. A case can be made for maintaining less severe cases in the paediatric services until discharge at age 21 in order to ensure continuity of follow-up
- Joint cardiology and general paediatric/physician management for cases with severe carditis are recommended
- Further information regarding frequency and nature of routine review can be found in Section 11
COMMENCEMENT OF LONG-TERM PREVENTIVE MEASURES

Secondary prophylaxis

• Obtain consent from caregiver/case for IM penicillin treatment

• First dose of secondary prophylaxis should be delivered in hospital

Notification

• Case should be notified to a local ARF register if available (see Section 10.3). There should be an easy means to do this, via a standard notification form, telephone call or otherwise

• In addition, as ARF is a notifiable disease in New Zealand, each case should be notified to the local public health unit for national infectious disease surveillance

Contact community services to ensure follow-up

• The register coordinator (if available) should notify community health staff about ARF cases in their area. The notifying medical practitioner should also make direct contact with those in the community responsible for prophylaxis delivery in order to ensure that they are aware of the diagnosis, the need for secondary prophylaxis and any other specific follow-up requirements. This may include district nurses, public health nurses, medical officer of health and other public health staff

• A community nurse and/or community health worker for the area where the case resides should also do a ward and/or family visit if possible before discharge

• Where relevant, it is also important for consent to be obtained from the case (or caregiver) for their local Māori or Pacific provider to know about the illness

Education

• At the time of diagnosis, it is essential that the disease process be explained to the patient and their family in a culturally appropriate way, using available educational materials and interactive discussion. Further education, using culturally appropriate educational materials should follow once the case has returned home

• For further information regarding education see Section 10.2

Organise dental check and ongoing dental care

• This is critical in the prevention of endocarditis. As those without rheumatic valve damage may still be at long-term risk of developing RHD, particularly in the event of recurrent episodes of ARF, dental care is essential, regardless of the presence or absence of carditis

• Each case should be notified to the appropriate school dental service or dentist

Contact management

• All symptomatic and asymptomatic household contacts of the index case aged 3 years and older should have a throat swab if the contact was no longer than one month before the onset of ARF in the index case. This should be organised through the appropriate public health unit and all contacts with positive GAS cultures should be offered antibiotic treatment. Streptococcal acquisition rates of 25% or greater have been recorded in family contacts of streptococcal pharyngitis

Opportunistic care

• It is important to note this opportunity to provide information and other services for ARF cases, whom frequently have other challenges to their general wellbeing. This may include promoting a healthy diet, exercise and hygiene, as well as assistance with socioeconomic stressors, and the opportunity for on going support.

Occasionally, when the diagnosis has already been confirmed and the case is not unwell (e.g. mild recurrent chorea in a child with no other symptoms or signs), outpatient management may be appropriate. In such cases health staff must ensure that investigations, treatment, health education, registration (where available) and notification are all completed and prophylaxis commenced

Controlled studies have failed to show that treating ARF with large doses of penicillin affects the outcome of rheumatic valvular lesions 1 year later. Despite this, most authorities recommend a course of penicillin, even if throat cultures are negative, to ensure eradication of streptococci that may persist in the upper respiratory tract (Grade D)

Most people labelled as being allergic to penicillin are not. Because penicillin is the best antibiotic choice for secondary prophylaxis it is recommended that those with stated penicillin allergy be investigated carefully, preferably with the help of an allergist, before being accepted as truly allergic (Grade D) (Section 6)

If the symptoms and signs do not remit substantially within 3 days of commencing anti-inflammatory medications, a diagnosis other than ARF should be considered
The use of glucocorticoids and other anti-inflammatory medications in rheumatic carditis has been studied in two meta-analyses. All of these studies of glucocorticoids were performed more than 40 years ago, and did not use drugs in common use today. These meta-analyses failed to suggest any benefit of glucocorticoids or IVIG over placebo, or of glucocorticoids over salicylates, in reducing the risk of long-term heart disease (Level I). The available evidence suggests that salicylates do not decrease the incidence of residual RHD (Level IV). Therefore, salicylates are not recommended to treat carditis (Grade C). Glucocorticoids may be considered for those with heart failure in whom acute cardiac surgery is not indicated (Grade D). This recommendation is not supported by evidence, but is made because many clinicians believe that glucocorticoids may lead to more rapid resolution of cardiac compromise, and even be life-saving in severe acute carditis. The potential major adverse effects of short courses of glucocorticoids, including gastrointestinal bleeding and worsening of heart failure as a result of fluid retention, should be considered before they are used. If glucocorticoids are used, the drug of choice is oral prednisone or prednisolone (1-2mg/kg/day, to a maximum of 80mg once daily or in divided doses). Intravenous methyl prednisolone may be given in very severe cases. If a week or less of treatment is required, the medication can be ceased when heart failure is controlled, and inflammatory markers are improving. For longer courses (usually no more than 3 weeks is required), the dose may be decreased by 20-25% each week. Treatment should be given in addition to the other anti-failure treatments outlined below. Mild to moderate carditis does not warrant any specific treatment. As glucocorticoids will control joint pain and fever, salicylates can usually be discontinued, or the dose reduced, during glucocorticoid administration. Salicylates may need to be recommenced after glucocorticoids are discontinued to avoid rebound joint symptoms or fever.

Side effects of carbamazepine include CNS adverse reactions (dizziness, headache, ataxia, drowsiness, fatigue and diplopia); gastrointestinal disturbances (nausea and vomiting), as well as allergic skin reactions. Uncommon side effects include abnormal involuntary movements (e.g. tremor, asthenia, dystonia and tics) and nystagmus. Rarely carbamazepine can cause orofacial dyskinesia, oculomotor disturbances, speech disorders (e.g. dysarthria and slurred speech), choreoathetotic disorders, peripheral neuritis, paresthesia, muscle weakness and paretic symptoms.

Side effects of valproic acid include pancreatitis, hepatic toxicity, hyperammonaemia and thrombocytopenia.

Observation and general hospital care
Guidelines for general in-hospital care are provided in Table 11 (Grade D).

Table 11. Guidelines For General In-Hospital Care

<table>
<thead>
<tr>
<th>NURSING RECORDINGS</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Temperature, pulse, RR, BP 4 times daily</td>
</tr>
<tr>
<td>• Sleeping pulse (e.g. 0200 hrs)</td>
</tr>
<tr>
<td>• If pulse &gt;100bpm, record apical HR</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>DIET</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Free fluids (if no heart failure)</td>
</tr>
<tr>
<td>• Normal diet (limit extras)</td>
</tr>
<tr>
<td>• Early dietary advice if overweight and in failure, to avoid further weight gain</td>
</tr>
<tr>
<td>• Weekly weight</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>BED REST AND GENERAL CARE</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Examine daily for the pattern of arthritis and the presence of heart murmur, choreiform movements, skin rash and subcutaneous nodules</td>
</tr>
<tr>
<td>• If clinical carditis present:</td>
</tr>
<tr>
<td>• Document cardiac symptoms and signs</td>
</tr>
<tr>
<td>• Daily weight and fluid balance chart</td>
</tr>
<tr>
<td>• Medications as appropriate (see Table 10 and Appendix D)</td>
</tr>
<tr>
<td>• See general guidelines for bed rest (Table 10)</td>
</tr>
<tr>
<td>• Cardiology opinion</td>
</tr>
<tr>
<td>• Repeat investigations as necessary</td>
</tr>
<tr>
<td>• Provide cultural support (as relevant)</td>
</tr>
<tr>
<td>• Plan care to provide rest periods</td>
</tr>
<tr>
<td>• Provide age-appropriate activities</td>
</tr>
<tr>
<td>• Notify school teacher</td>
</tr>
<tr>
<td>• Involve family in care.</td>
</tr>
</tbody>
</table>

Source: Adapted from Lennon D. 2004. [32]
Note: RR = respiratory rate; BP = blood pressure; HR = heart rate.
Discharge

Timing of discharge
The duration of treatment is dictated by the clinical response and improvement in inflammatory markers (ESR and CRP). Most cases of ARF without severe carditis can be discharged from hospital after approximately two weeks. The length of admission will partly depend on the social and home circumstances. If cases come from remote communities or other settings with limited access to high quality medical care, it is advisable to discuss discharge timing with the person, family and the local primary health care team (particularly Maori or Pacific health providers where possible). In some cases, it may be advisable to prolong the hospital stay until recovery is well advanced.

Advice on discharge
All cases should have a good understanding of the cause of rheumatic fever and the need to have sore throats treated early in other family members. Contact management (as per Table 10) should be discussed.

Cases and their families should understand the reason for secondary prophylaxis and the consequences of missing a BPG injection. The first dose of BPG is usually given in a hospital setting. Arrangements for the first injection post discharge should be made. They should be given clear information about where to go for secondary prophylaxis once discharged, know who to contact with questions concerning their follow-up or secondary prophylaxis, and be given written information on appointments for follow-up with their local medical practitioner, physician/paediatrician and cardiologist (if needed). They should be advised of the appropriate activity level until their next clinic appointment.

Cases and their families should also be reminded of the importance of additional antibiotic prophylaxis for dental and other procedures to protect against endocarditis (Appendix H).

Copies of the discharge summary should go to the following services: community nursing staff responsible for prophylaxis delivery (such as district nurse, public health nurse), rheumatic fever secretary or staff responsible for the register (where applicable), primary care provider and the family.
Secondary Prevention
SECONDARY PREVENTION

Secondary prevention of rheumatic fever is defined as the continuous administration of antibiotics to cases who have had a previous attack of ARF or well-documented RHD. The purpose is to prevent infection of the upper respiratory tract with GAS and the development of recurrent rheumatic fever.13

6. Prophylaxis Regimes

The regular administration of antibiotics to prevent infection with group A streptococcal (GAS) and recurrent ARF is recommended for all people with a history of ARF or RHD. This strategy has been proven in randomised controlled trials to prevent streptococcal pharyngitis and recurrent ARF.

Penicillin

In early studies of ARF prophylaxis using sulphonamides, 1.5% of treated cases developed ARF recurrences, compared to 20% of untreated cases. Subsequently, penicillin was found to be more efficacious than sulphonamides (Level I).35,83 A recent Cochrane meta-analysis101 concluded that the use of penicillin (compared to no therapy) is beneficial in the prevention of recurrent ARF, and that intramuscular benzathine penicillin G (BPG) is superior to oral penicillin in the reduction of both recurrent ARF (87–96% reduction) and streptococcal pharyngitis (71–91% reduction) (Level I) (Appendix E).

Secondary prophylaxis also reduces the severity of RHD. It is associated with regression of heart disease in approximately 50-70% of those with adequate adherence over a decade (Level III 2),56,102,103 and reduces mortality (Level III 2).104

Dose

The internationally accepted standard dose of BPG for the secondary prevention of ARF in adults is 1,200,000 U.3,38,105 The dose for children is less clear. In New Zealand, it is recommended that 1,200,000 U of BPG should be used for secondary prophylaxis for all persons weighing 20kg or more (Level III-2, Grade B), and 600,000 U for those weighing less than 20kg (Grade D).106

Frequency

While BPG is usually administered every four weeks (28 days), serum penicillin levels may be low or undetectable 28 days following a dose of 1,200,000 U.107 Fewer streptococcal infections and ARF recurrences occurred among those receiving three-weekly BPG (Level I).107,108,109 Moreover, the three-weekly regimen resulted in greater resolution of mitral regurgitation in a long-term randomised study in Taiwan (66% vs 46%) (Level II).110 Prospective data from New Zealand however, showed that recurrences were rare among people who were fully adherent to a four-weekly BPG regimen. In Auckland (1993 to 1999), the rate of recurrence in fully adherent individuals on a 28 day regime was 0.07 per 100 patient years. Failure on the prophylaxis programme (i.e. including those who were less than fully adherent) was 1.4 per patient years.36,17 This compares favourably to prophylaxis failure reported in Taiwan of 0.25 (21-day programme) and 1.29 (28-day programme) per 100 patient years.111 Furthermore, a four-weekly regime is preferable to a three-weekly regime because of the resource and compliance implications (Grade D). In New Zealand, three weekly (21-day) BPG is recommended only for those who have confirmed recurrent ARF despite full adherence to four-weekly (28-day) BPG delivery (Grade C).36,17

An alternative strategy is the administration of larger doses of BPG, leading to a higher proportion of people with detectable serum penicillin levels four weeks after injection.112 However, until more data are available, this strategy cannot be recommended.
Table 12. Recommended Antibiotic Regimens for Secondary Prevention of Acute Rheumatic Fever/Rheumatic Heart Disease

<table>
<thead>
<tr>
<th>ANTIBIOTIC</th>
<th>DOSE</th>
<th>ROUTE</th>
<th>FREQUENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>First line</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Benzathine penicillin G (BPG)</td>
<td>1,200,000 U $\geq$ 20kg</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>600,000 U &lt; 20kg</td>
<td></td>
<td>BPG is most effectively given as a deep intramuscular injection*</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>4-weekly (28 days), or 3-weekly for those who have had confirmed recurrent ARF despite full adherence to 4-weekly BPG</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>BPG is most effectively given as a deep intramuscular injection</td>
</tr>
</tbody>
</table>

| Second line (If intramuscular route is not possible or refused)* |                       |         |                                    |
|----------------------------------------------------------------|-----------------------|---------|                                    |
| Phenoxymethylpenicillin (Penicillin V)                          | 250mg                  | Oral    | Twice daily                        |
| Following documented penicillin allergy**                      |                       |         |                                    |
| Erythromycin (EES)                                             | 40mg/kg per day (children) | Oral    | 2-4 divided doses (maximum 1g/day) |
| 400mg (adolescents and adults)                                 | Oral                  |         | Twice daily                        |

* 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. In addition, oral penicillin V incurs a cost to the patient, while IM BPG is free when provided through an ARF prevention programme. Oral penicillin should be reserved for cases who refuse intramuscular BPG (Level II, Grade B). If a patient is offered oral penicillin, the consequences of missed doses must be emphasised and adherence carefully monitored (Grade D).

** 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. A prospective study of 1,790 ARF/RHD patients found similar rates of allergic reactions in those receiving long-term penicillin therapy and those receiving short-term therapy for sexually transmitted diseases (Level III-2). Before commencing penicillin treatment, cases should be carefully questioned about known allergies to penicillin and other beta-lactam antibiotics. When patients state they are allergic to penicillin or when a non-specific reaction has been reported but there is no unequivocal evidence, they should be investigated for 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. Penicillin desensitisation is not applicable to these patients, even with a regime of more frequent injections, as it would have to be repeated before each dose of BPG. A RAST (RadioAllergoSorbent Test) may be used as a screening tool only. Because this is a specific but not very sensitive test, a negative RAST test must be followed up in all cases with penicillin skin testing and/or consideration of a graded challenge if appropriate (Grade D).

New Zealand has been affected by inconsistent supply of benzathine penicillin over recent years. This poses potential risks to those requiring four-weekly prophylaxis. Organisational approaches to secondary prevention should seek to ensure consistent supply at the national, regional and local levels. However, when benzathine penicillin is unavailable, oral penicillin or erythromycin can be given (as per Table 12).

Secondary prophylaxis while breast feeding, during pregnancy and while on oral contraceptives

As there is no evidence of teratogenicity, penicillin prophylaxis should continue for the duration of pregnancy for the prevention of recurrent ARF (Grade D). Erythromycin is also considered safe in pregnancy, although controlled trials have not been conducted.

Penicillin is also generally considered to be safe to use during breast feeding. Concentrations lower than plasma levels are excreted in breast milk. No adverse effects have been reported. Erythromycin is also excreted into breast milk, but there are no reports of adverse effects in infants and it is considered safe to use (Grade D).
Oral contraceptives are still recommended for women of child-bearing age while on BPG prophylaxis. Progesterone-only oral contraceptives do not interact with BPG therapy. An interaction between IM BPG and the combined oral contraceptive is possible, although this interaction is suggested to only be of significance for short courses of antibiotic therapy (less than three weeks). In addition, the risk of interaction with antibiotics is small enough that it may not be identifiable from the one to three percent risk of oral contraceptive failure (Grade C). Caution is advised when considering the use of the combined oral contraceptive pill in women with complicated rheumatic heart disease/valve disease or atrial fibrillation, especially for cases also on warfarin. A levonorgestrel-releasing intra uterine contraceptive device (such as Mirena) would be more suitable (if in a stable relationship) (Grade D).

Secondary prophylaxis in anti-coagulated cases

Intramuscular bleeding from BPG injections, used in conjunction with anticoagulation therapy in New Zealand, is rare. Thus, BPG injections should be continued for those who are anti-coagulated, unless there is evidence of uncontrolled bleeding or the international normalised ratio (INR) is outside the defined therapeutic window (Grade D). Cases discharged from hospital on oral penicillin following valve surgery should recommence BPG as soon as is practical.

7. Duration of Secondary Prophylaxis

The appropriate duration of secondary prophylaxis depends on a number of factors. These include:

- age (ARF recurrence is less common after the age of 25 and uncommon after the age of 30)
- clinical pattern (presence or absence of carditis or RHD and severity of carditis or RHD)
- environment (particularly the likelihood of ongoing exposure to GAS)
- time elapsed since last episode of ARF (ARF recurrences are less common greater than five years since last episode)

Based on these factors, the recommended duration of secondary prophylaxis is outlined in Table 13. The duration of prophylaxis recommended is also outlined in Algorithm 3.

Table 13. New Zealand Recommendations for the Duration of Secondary Prophylaxis

<table>
<thead>
<tr>
<th>CATEGORY*</th>
<th>DURATION OF PROPHYLAXIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>All persons with ARF with no or mild carditis</td>
<td>Minimum of 10 years after most recent episode ARF or until age 21 years (whichever is longer)**</td>
</tr>
<tr>
<td>All persons with ARF with moderate carditis</td>
<td>Minimum of 10 years after most recent episode ARF or until age 30 years*** (whichever is longer)</td>
</tr>
<tr>
<td>All persons with ARF with severe carditis</td>
<td>Minimum of 10 years after most recent episode ARF or until age 30 years (whichever is longer), and then specialist review for consideration of the need for continuation of prophylaxis, probably lifelong.</td>
</tr>
</tbody>
</table>

* Definition of categories:

- Mild carditis:
  - Any valve lesion(s) graded mild clinically, or by echocardiography, with no clinical evidence of heart failure and no evidence of cardiac chamber enlargement on CXR, ECG or echo

- Moderate carditis:
  - Any valve lesion of moderate severity clinically (e.g. mild or moderate cardiomegaly), or
  - Any moderate severity valve lesion on echocardiography, or
  - Any echocardiographic evidence of cardiac chamber enlargement
• Severe carditis:
  • Any severe valve lesion clinically (significant cardiomegaly expected, and/or heart failure), or
  • Any severe valve lesion on echocardiography, or
  • Any impending or previous cardiac surgery for RHD

• Pure aortic stenosis is rarely due to ARF and in such cases an alternative diagnosis should be considered

• When there is both mitral and aortic regurgitation, one of them must be graded moderate by echo standards in order for the carditis to be classified of moderate severity

• Tricuspid and pulmonary regurgitation graded mild or greater may be seen in people with normal hearts who have fever, volume overload or pulmonary hypertension. For this reason a diagnosis of carditis should not be based on right-side regurgitation alone. Although pulmonary and tricuspid regurgitation are often seen in association with left-sided lesions in ARF, pressure and volume overload must be excluded before attributing even moderate tricuspid regurgitation to valvulitis. If both left and right-sided lesions coexist in ARF carditis, then the predominant influence is the severity of the left-sided lesion

** A review of data from the Auckland Acute Rheumatic Fever Register (1993-1999) in New Zealand found that recurrences occurred up to 21 years after completion of prophylaxis programmes. 77% were within the first seven years, and 30% were greater than ten years. The mean overall recurrence interval between last attack and recurrence was 8.6 years. Of the cases that received ten years prophylaxis, there were two ARF recurrences after discharge and an estimated 2,200 patient years of follow-up (0.1/100 patient years). Two "breakthrough" recurrences occurred in this series in cases who were inadvertently discharged early off prophylaxis (aged 16 and 17 years). This data suggest that in the New Zealand environment, maintenance of prophylaxis to 21 years of age in cases with absent or mild heart disease is safe and effective (Level IV, Grade C)

*** Of the Auckland (1993-1999) cases, only five recurrences occurred after the age of 30. Therefore it is reasonable to cease secondary prophylaxis at that age, except when individual circumstances warrant continuing (e.g. when cases wish to reduce even a small chance of a recurrence) (Level IV, Grade C)

• Individuals working or living with children or in a living situation where there is overcrowding or close proximity to others (such as boarding schools, barracks, and hostels) have a higher risk of exposure to GAS and subsequent development of ARF. In these cases, consideration should be given to extending the duration of prophylaxis (Grade D)

• For those presenting at an older age (over the age of 21 years), with no or mild carditis, it is possible to consider discharge from prophylaxis after 5 years (Grade D)

• The duration of prophylaxis presented here refers to ‘definite’ and ‘probable’ cases of ARF (see Section 4). For those with ‘possible’ ARF (where there is strong clinical suspicion, but insufficient signs and symptoms to fulfill the diagnosis), a minimum of 5 years prophylaxis should be considered, with regular review (Grade D)

• For those presenting with RHD for whom no initial episode of ARF can be identified, the decision to commence and cease penicillin prophylaxis should be taken on an individual basis with regard to the age of the case, severity of the disease, possible age of first attack and risk of exposure to GAS.

Before stopping prophylaxis, recipients who are known to have had carditis should be evaluated for symptomatic deterioration and the stability and severity of valve lesions. This should include echocardiographic assessment (Grade D). Where limited echocardiography is available, preference should be given to those with a history of moderate or greater carditis, a history of one or more ARF recurrences or clinical evidence of carditis (e.g. a murmur) (Grade D). The anticipated and actual dates of cessation should be documented in the medical records and on the ARF register where possible, (see Section 10.3). The date of cessation may be reviewed if there is a change in clinical or echocardiographic severity, specialist recommendation, a change in environmental exposure to GAS, or a recurrence of ARF (Grade D).
8. Protocol for Secondary Prophylaxis Delivery

In the New Zealand environment, it is recommended that secondary prophylaxis is delivered by community nursing staff at schools, in the workplace or at home (Table 14).

In each area this delivery should be supported by the presence of a rheumatic fever register (see Section 10.3), and it is also recommended that in each area specific medical staff sign three-monthly designated authorisation for the nurses to deliver BPG. The generation of these prescriptions will also be assisted by a register system.

Table 14. Suggested Protocol for the Delivery of Secondary Prophylaxis by Community Nurses

<table>
<thead>
<tr>
<th>PREPARATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Identify client (full name and date of birth)*</td>
</tr>
<tr>
<td>• Confirm that consent** has been given for BPG delivery at school (if appropriate)</td>
</tr>
<tr>
<td>• Check allergy status, if symptoms of allergy reported from previous injection then withhold injection, document and report to GP and specialist</td>
</tr>
<tr>
<td>• Note appropriate dose of adrenaline required for current age if necessary for an anaphylactic reaction (see Appendix F)</td>
</tr>
<tr>
<td>• Check the prescription: date, frequency and dose</td>
</tr>
<tr>
<td>• Check weight for children on 0.6 megaunits (mU) of bicillin to ensure dose for next injection is appropriate (i.e. remain at &lt;20kg). Record weight in progress notes. If dose should change, document and inform the local prescriber and register coordinator to ensure the dose is changed for the next delivery</td>
</tr>
<tr>
<td>• Give full explanation to client</td>
</tr>
<tr>
<td>• Position client lying or as preferred</td>
</tr>
<tr>
<td>• Wash hands</td>
</tr>
<tr>
<td>• Prepare BPG (bicillin cartridge and Tubex is the current system). If 0.6 mU dose is required dispose of half the syringe contents prior to administration. Warm in hands</td>
</tr>
<tr>
<td>• Alcohol swab injection site, allow to dry</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>DELIVERY***</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Apply pressure to injection site for 10 seconds and consider other measures to reduce pain (Table 15)</td>
</tr>
<tr>
<td>• Administer bicillin slowly into ventrogluteal, dorsogluteal area of buttock or vastus lateralis or thigh (or as per local area policy)</td>
</tr>
<tr>
<td>• Dispose of the used syringe in a sharps container after removing Tubex</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>OBSERVATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Observe client for a minimum of 10 minutes after administration of bicillin for any signs and symptoms of an allergic reaction. Local policy may suggest a 20 minute observation period</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>EVALUATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Complete record of administration</td>
</tr>
<tr>
<td>• Review education needs/knowledge</td>
</tr>
<tr>
<td>• Confirm next appointment and any other follow-up needs</td>
</tr>
<tr>
<td>• Consider opportunity to review broader health issues such as mobility and activity levels, nutritional status and dietary habits, dental hygiene and support.</td>
</tr>
</tbody>
</table>

* If under 16: confirm identification with another responsible person (i.e. caregiver, school receptionist)

** Consent for the duration of BPG delivery will have been obtained in hospital prior to the initiation of prophylaxis (Table 10). A separate consent needs to be obtained from the caregiver/parent only for delivery at school

*** If the client is not available, and the full syringe has been maintained in cold chain, it can be returned to the medicine fridge. If the full syringe has not been maintained within the cold chain, then it needs to be discarded.
9. Anaphylaxis

Anaphylaxis is an uncommon reaction following IM or IV antibiotics, immunisation or other medicines. Anaphylaxis to benzathine penicillin is rare. In a prospective international study after 32,430 injections during 2,736 patient years of observation, 57 (3.2%) of the 1,790 patients had an allergic reaction and four (0.2% or 1.2 per 10 000 injections) had anaphylaxis. The long-term benefits of prophylaxis therefore far outweigh the potential risk of a serious allergic reaction. The response to an anaphylactic reaction to a BPG dose and the management of anaphylaxis can be found in Appendix F.

10. Improving Adherence to Secondary Prophylaxis

The persistence of recurrent ARF in some areas of New Zealand highlights the continued failure of secondary prevention.

Failure to prevent recurrent ARF in a study from the Gisborne area, was thought to be due to a range of factors including a lack of recognition of the efficacy of parenteral BPG compared to oral regimens, inadequate adherence, unreliable data collection and the lack of long-term continuity of care. Improved adherence to prophylaxis is seen with active follow-up of cases when BPG doses are missed, the identification of local dedicated staff members responsible for delivery of secondary prophylaxis, developing a personal rapport with each case and coordinating routine care. Effective communication between health staff and families is important. In New Zealand, it is particularly important to support and utilise the expertise, experience, community knowledge, culture and language skills of Māori and Pacific health workers in order to assist with adherence to secondary prophylaxis.

Three methods for improving compliance will be discussed further in this guideline:

- reducing the pain of the BPG injection
- education
- the use of rheumatic fever registers.

Reducing the pain of BPG injections

The pain of BPG injections is usually not a critical factor in determining adherence to secondary prophylaxis. Nonetheless, techniques that safely reduce injection pain (Table 15) should be promoted.

Table 15. Measures That May Reduce the Pain of Benzathine Penicillin G Injections

<table>
<thead>
<tr>
<th>Technique</th>
<th>Note</th>
</tr>
</thead>
<tbody>
<tr>
<td>Use a 23-gauge needle</td>
<td>*</td>
</tr>
<tr>
<td>Apply pressure with thumb for 10 seconds before inserting needle</td>
<td>**</td>
</tr>
<tr>
<td>Warm syringe to room temperature before using</td>
<td>***</td>
</tr>
<tr>
<td>Allow alcohol from swab to dry before inserting needle</td>
<td>***</td>
</tr>
<tr>
<td>Use of ethylchloride spray prior to injection</td>
<td>#</td>
</tr>
<tr>
<td>Deliver injection very slowly (preferably over at least 2–3 mins)</td>
<td>***</td>
</tr>
<tr>
<td>Distraction techniques during injection (e.g. with conversation)</td>
<td></td>
</tr>
<tr>
<td>Good rapport with the case, assisted by having a designated nurse for each case, is a significant aid to injection comfort, compliance, and understanding</td>
<td>***</td>
</tr>
</tbody>
</table>

* A smaller gauge needle and increasing the volume of injection to 3.5ml improved acceptability in Taiwan.
** Direct application of pressure to the injection site has been shown to decrease pain of intra-muscular injections.
*** As these measures are logical and benign they are recommended, despite lack of evidence (Grade D)
# Although merely a topical agent, some cases have reported reduced pain and bruising following the appropriate use of ethylchloride spray (Grade D)

The addition of 0.5–1.0 ml of 1% lignocaine is used elsewhere. It significantly reduces pain immediately and in the first 24 hours after injection, while not significantly affecting serum penicillin concentrations. Procaine penicillin added to BPG also reduces pain and local reactions. The combination is effective for the treatment of streptococcal pharyngitis, but the formulations tested to date have not sustained adequate serum penicillin levels for long enough for secondary prophylaxis. However, with the pre-loaded (“Tubex”) system syringes currently used in New Zealand it is not recommended or possible for community staff to add lignocaine or procaine penicillin (Grade D).
Education

Health education is critical at all levels. Lack of parental awareness of the causes and consequences of ARF/RHD was a key contributor to poor adherence among children on long-term prophylaxis in Egypt. In a number of regions in India, comprehensive health education has improved community awareness of sore throat, ARF and RHD and assisted case identification. Comprehensive health education and promotion was also a key component in the successful control of RHD in the French Caribbean. Improved health staff awareness of the diagnosis and management of ARF and RHD is necessary in order to improve case findings, encourage compliance with prophylaxis and to improve the quality and delivery of health education delivered to cases and their families.

Education provided to the case and their family should cover:

- the cause and complications of ARF
- the reason for secondary prophylaxis and the signs and symptoms of recurrence
- the prevention of endocarditis and the differences between this and secondary prophylaxis of ARF
- sore throat management
- the importance of medical and dental follow-up
- how to contact the relevant people or agencies should they require further information or assistance.

The National Heart Foundation of New Zealand produces a booklet called “What is Rheumatic Fever?” to assist in education provision to cases and their families. This is available to order from www.heartfoundation.org.nz

ARF Registers

Registers of people with RHD or a history of ARF are a key element in ARF recurrence and RHD control at an individual, community and national level.

In 1978, the WHO promoted the use of disease registers as part of community programmes to help coordinate prevention of ARF recurrences and of RHD. The use of these registers has been proven in both developing and developed countries to enhance the impact of secondary prevention strategies for ARF and to effectively reduce morbidity and mortality.

Register-based RHD control programmes have been successful in New Zealand. By the early 1980s, ARF registers had been implemented in Waikato, Northland, Auckland, Gisborne and Rotorua. Despite similarities, each programme developed independently of any national framework, and each was shown to be effective at reducing admissions for ARF recurrences. In New Zealand, ARF became a notifiable condition under the national surveillance and management framework in 1986.

In 2001, a survey describing register-based ARF prevention programmes in New Zealand was conducted. Two types of registers were described: management and surveillance. Register-based ‘management’ programmes use a register to coordinate community-based prophylaxis provided predominantly by district nursing services, collate information on prophylaxis delivery and encourage parental prophylaxis. Management programmes also use their registers to perform a varying range of other functions including informing health care workers (such as dentists and GPs) of those who are receiving prophylaxis, generating or prompting penicillin prescriptions and accumulating data for evaluation. Six register-based management programmes were operating in New Zealand in 2001 (predominantly through public health units in collaboration with clinicians). These were based in Northland, Auckland (district nurses in association with paediatricians), Rotorua (established by an association of GP’s), Gisborne, Hawkes Bay and Lower Hutt. Collectively, these programmes covered nine health districts containing 51.1% of the population and 81.9% of ARF notifications between 1995 and 2000. A further three ‘surveillance’ programmes, without clinician input, were described in Whakatane, Wanganui and Palmerston North. These programmes maintained a record of cases receiving prophylaxis, but did not have a role in coordinating the provision of prophylaxis.

In total, these register systems covered 94% of notified ARF cases, and they were considered largely responsible for reducing ARF recurrence from 22% (of all ARF episodes) between 1972 and 1981 to only 6% between 1982 and 1992.

The Auckland Acute Rheumatic Fever Register, established in 1982, is a population-based register covering 60% of New Zealand ARF registrations. The register is used both as a surveillance register and a tool to generate dental referrals and delegated authority prescriptions to aid penicillin delivery by the district nursing service. Those who miss their prophylaxis are actively sought for three to six months before being inactivated on the register. Community nurses from other areas can also refer confirmed cases to the register for ongoing prophylaxis. A recent study evaluated the effectiveness of the Auckland ARF Register and of 28 day penicillin prophylaxis by auditing recurrences notified to the register in this time period for those with mild or absent heart disease without active follow-up after at least ten years. In this study, an overall programme failure rate of 1.4 per 100 patient years was determined with a penicillin failure rate of 0.07 per 100 patient years. Earlier audits of the same register from 1972 to 1981 (1.5 per 100 patient years) and 1982 to 1992 (0.6 per 100 patient years) reached similar conclusions. These rates of programme failure are highly acceptable when compared to other published data (0.0-2.8 per 100 patient years).
It is recommended that all regions of New Zealand with substantial populations with ARF or RHD establish a coordinated ARF register (preferably computerised) which provides individual and community reports, recall lists, reports on ARF/RHD epidemiology and monitors the effectiveness of the local prevention programme (Grade C).

The main aims of ARF registers are summarised in Table 16.

Table 16. Primary Aims of ARF Register Systems

- Increase uptake of and adherence to secondary prophylaxis
- Reduce recurrences of ARF and decease hospitalisations from ARF/RHD (Level III)
- Improve case detection
- Record prophylaxis delivery
- Employ recall and reminder systems for ARF cases, identify individuals with poor adherence to long-term therapy for targeted educational activities and other interventions
- Monitor the movement of ARF cases (who are typically highly mobile), while conforming to privacy legislation and patient confidentiality
- Improve the coordination of ongoing care requirements and follow-up
- Identify and register new cases of ARF and RHD
- Use data to improve programme strategies and determine changes in disease epidemiology
- Fulfil legal requirements of disease notification
- Improve awareness amongst health professionals
- Centralised registers can also support the provision of prophylaxis for those who move between communities.

The register can then be used as the basis for a coordinated control programme. This is the most effective approach to improving BPG adherence and clinical follow-up of people with RHD, including specialist review and echocardiography (Level III-3). Elements of such a programme are listed in Table 17 (Grade C).

Table 17. Recommended Elements of a Register-Based Control Programme

- A local (preferably computerised) ARF register, established within existing health care networks or public health units, with all the properties and data as described in Tables 16 and 18
- Commitment from regional and local services, particularly to ensure long-term funding
- Activities guided by locally relevant, evidence-based guidelines
- A coordinator for each register programme
- A commitment to partnerships between clinicians and public health services in order to support the needs of people with ARF/RHD and the community
- An ability to assess and monitor the burden of disease
- Provision of education for health practitioners, the community, those with rheumatic fever or rheumatic heart disease and their families
- Provision or support for the provision of health education within the local community, community health service and for community health workers
- A follow-up system (such as dedicated ARF/RHD clinics) that ensures that ongoing care is delivered, particularly to those at highest risk
- A mechanism for monitoring delivery of secondary prophylaxis and ongoing care, programme reporting and independent evaluation
- Some areas may also be able to have an effective advisory committee that may include cardiologists, paediatricians, general practitioners, physicians, epidemiologists, nurses, public health practitioners and relevant community representative.

* A dedicated coordinator with data entry support is critical to the success of the programme. 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. To ensure that the programme continues to function well despite staffing changes, activities must be integrated into the established health system.
Key data elements of ARF/RHD registers

A possible dataset for ARF/RHD registers is outlined in Table 18.

Table 18. Dataset for Acute Rheumatic Fever Register*

<table>
<thead>
<tr>
<th>DOMAIN</th>
<th>DATA ELEMENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographics</td>
<td>National Hospital Index and name(s)</td>
</tr>
<tr>
<td></td>
<td>Date of birth</td>
</tr>
<tr>
<td></td>
<td>Gender</td>
</tr>
<tr>
<td></td>
<td>Address and phone numbers (including cellphone for text message contacting), alternate address</td>
</tr>
<tr>
<td></td>
<td>Details of parents/caregivers</td>
</tr>
<tr>
<td></td>
<td>Ethnicity</td>
</tr>
<tr>
<td></td>
<td>GP details</td>
</tr>
<tr>
<td></td>
<td>School at diagnosis (where relevant) **</td>
</tr>
<tr>
<td>Initial ARF diagnosis</td>
<td>Date and place of diagnosis and date of admission to hospital</td>
</tr>
<tr>
<td></td>
<td>Definite, probable or possible diagnosis</td>
</tr>
<tr>
<td></td>
<td>Medications taken prior to presentation/admission</td>
</tr>
<tr>
<td></td>
<td>Major criteria:</td>
</tr>
<tr>
<td></td>
<td>• Presence (and severity) of carditis</td>
</tr>
<tr>
<td></td>
<td>• Presence (and site) of arthritis</td>
</tr>
<tr>
<td></td>
<td>• Presence of chorea</td>
</tr>
<tr>
<td></td>
<td>• Presence of erythema marginatum and/or subcutaneous nodules</td>
</tr>
<tr>
<td></td>
<td>Minor criteria:</td>
</tr>
<tr>
<td></td>
<td>• Fever</td>
</tr>
<tr>
<td></td>
<td>• Acute phase reactants</td>
</tr>
<tr>
<td></td>
<td>• P-R interval</td>
</tr>
<tr>
<td></td>
<td>Evidence of a preceding GAS infection:</td>
</tr>
<tr>
<td></td>
<td>• History of sore throat</td>
</tr>
<tr>
<td></td>
<td>• Throat swab</td>
</tr>
<tr>
<td></td>
<td>• Titres</td>
</tr>
<tr>
<td>ARF recurrences***</td>
<td>Onset date</td>
</tr>
<tr>
<td></td>
<td>Presence and severity of carditis</td>
</tr>
<tr>
<td></td>
<td>Other symptoms and signs at each recurrence</td>
</tr>
<tr>
<td></td>
<td>Prophylaxis status at time of recurrence</td>
</tr>
<tr>
<td>DOMAIN</td>
<td>DATA ELEMENTS</td>
</tr>
<tr>
<td>-----------------------------</td>
<td>-------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>RHD diagnosis</td>
<td>Onset date/date of diagnosis</td>
</tr>
<tr>
<td></td>
<td>Documented history of ARF</td>
</tr>
<tr>
<td></td>
<td>Valvular dysfunction and disease severity at time of diagnosis</td>
</tr>
<tr>
<td></td>
<td>Surgery</td>
</tr>
<tr>
<td></td>
<td>Dentist</td>
</tr>
<tr>
<td>Secondary prophylaxis</td>
<td>Antibiotic used</td>
</tr>
<tr>
<td></td>
<td>Dose and frequency</td>
</tr>
<tr>
<td></td>
<td>Date commenced on prophylaxis</td>
</tr>
<tr>
<td></td>
<td>Date of last dose</td>
</tr>
<tr>
<td></td>
<td>Date of next expected dose</td>
</tr>
<tr>
<td></td>
<td>Designated authority</td>
</tr>
<tr>
<td></td>
<td>Expected date of cessation</td>
</tr>
<tr>
<td></td>
<td>Annual adherence data</td>
</tr>
<tr>
<td>Follow-up/recall</td>
<td>Date and place of last review</td>
</tr>
<tr>
<td></td>
<td>Date and place of next scheduled review by each provider (cardiologist,</td>
</tr>
<tr>
<td></td>
<td>paediatrician, physician, surgeon, echocardiography)</td>
</tr>
<tr>
<td></td>
<td>Recall system for missed BPG</td>
</tr>
<tr>
<td></td>
<td>Recall system for missed appointment</td>
</tr>
<tr>
<td>Mortality</td>
<td>Date and cause of death according to agreed criteria (e.g., due to RHD, not</td>
</tr>
<tr>
<td></td>
<td>due to RHD).</td>
</tr>
</tbody>
</table>

---

* This dataset is an amalgamation of systems currently in use in New Zealand. Some of the functions may be fulfilled elsewhere. Other information such as details of surgical procedures and medical management may also be included.

** To facilitate the set-up of school-based primary prevention programmes.

*** It is recommended that each ARF recurrence notified to the register is thoroughly investigated to determine if any changes in the system of prophylaxis delivery need to be made to prevent such recurrences from occurring in the future.
Outreach and out-of-town

The non-compliant and the non-presenting groups continue to be a major challenge to secondary prophylaxis. Transient living patterns or shifting without notifying staff of a forwarding address can create follow-up difficulties. In Auckland (1993 to 1999), 13 people suffered 14 recurrences because penicillin had been discontinued prematurely.\[16,17\]

As the populations at the highest risk of ARF are Māori and Pacific, the involvement of Māori and Pacific health workers, with their skills in outreach and their community knowledge, is important.

In addition, the presence of local ARF registers in New Zealand allows for inter-register referral (often nurse to nurse) of diagnosed ARF cases. This ensures continuity of care and prophylaxis when cases transfer to a new area.

Non-compliance

If a case is non-compliant, it is recommended that a number of methods of contact, over a number of months are attempted. Every effort should be made to utilise community contacts in the area, and a period “on hold” with continued attempts to contact, should be used prior to considering discharge. In Auckland early discharge off prophylaxis due to persistent non-compliance, is rare.

A protocol for the management of non-compliant cases can be found in Appendix G.
11. Routine Review and Structured Care Planning

A structured care plan should be developed and recorded in the notes of all persons with a history of ARF, or with established RHD. Table 19 lists the recommended review frequency (Grade D). This schedule may be tailored to the needs of the individual and may also differ depending on local resources. Auckland, for example (with 60% of New Zealand ARF cases), may not be able to see cases as frequently as is possible in other areas with a smaller case load.

Table 19. Recommended Routine Review and Management Plan for ARF and RHD*

<table>
<thead>
<tr>
<th>CLASSIFICATION</th>
<th>CRITERIA</th>
<th>REVIEW AND MANAGEMENT PLAN</th>
<th>FREQUENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low risk</td>
<td>ARF with no evidence of RHD or Trivial to mild valvular disease</td>
<td>Secondary prophylaxis (BPG)</td>
<td>4-weekly**</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Doctor review</td>
<td>3-5 yearly, or more frequently depending on local resource</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Echocardiography</td>
<td>Children on clinical change Adults on discharge</td>
</tr>
<tr>
<td>Medium or high risk#</td>
<td>Any moderate or severe valve lesion or Mechanical prosthetic valves or Tissue prosthetic valves and valve repairs</td>
<td>Secondary prophylaxis (BPG)</td>
<td>4-weekly**</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Influenza vaccination</td>
<td>Yearly</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cardiologist/physician/paediatrician review with echocardiography</td>
<td>6-24 monthly***</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dental review§</td>
<td>6 monthly</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Polysaccharide pneumococcal vaccination (Pneumovax 23)</td>
<td>5-yearly (max 3 doses)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Endocarditis prophylaxis</td>
<td>As required</td>
</tr>
<tr>
<td>Additional considerations</td>
<td>Following valve surgery</td>
<td>Medical assessment ECG Chest radiograph Echocardiography Full blood count Urea, creatinine, electrolytes INR if indicated</td>
<td>3-4 weeks post-discharge</td>
</tr>
</tbody>
</table>

* Review frequency 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 New Zealand, 4-weekly BPG is recommended unless confirmed recurrent ARF has occurred despite full adherence to prophylaxis. In this case, 3-weekly BPG is recommended (Grade D).

*** Close supervision until stable

# Anyone with severe valvular disease or moderate to severe valvular disease with symptoms should be referred for cardiological and surgical assessment as soon as is possible (Grade D).

§ Routine dental care is critically important in cases with a history of ARF and/or RHD. All patients should receive education about oral hygiene, and should be referred promptly for dental assessment and treatment when required. This is especially important prior to valvular surgery, when all oral/dental pathology should be investigated and treated accordingly (Grade D).
12. Prevention of Infective Endocarditis

Infective endocarditis is a dangerous complication of RHD. Although the effectiveness of additional antibiotic prophylaxis prior to dental or surgical procedures has not been proven, its use is supported by animal models of endocarditis and empirical observations, such as the reduction of bacteraemia.

Therefore, persons with established RHD or prosthetic valves should receive antibiotic prophylaxis prior to 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 for the procedures that require endocarditis prophylaxis and the appropriate antibiotics are currently being updated. These can be found on The National Heart Foundation of New Zealand website (http://www.nhf.org.nz). Some of these recommendations are also outlined on a wallet card to be carried by cases (Appendix H).

13. Case Finding Surveillance and Screening

Surveillance

Passive surveillance of ARF usually depends on case identification from health care providers. In New Zealand, ARF and recurrent ARF are notifiable conditions. Historically however, reliance on notification has under-estimated the burden of disease due to inaccuracies and incompleteness. In under-resourced settings, the deficiencies of passive surveillance are exacerbated by high turnover of hospital and primary care staff and lack of awareness of ARF by many health care providers.

Ideally, active surveillance should be used to augment passive surveillance (Grade D). This entails establishing mechanisms to identify new cases of ARF and to update information about existing cases. This could include:

- mechanisms allowing access to hospital coding data
- echocardiography reports
- specialist review correspondence
- primary health care information.

Where possible, these processes should be automated (including regular downloads of information regarding cases admitted to hospital with a diagnosis of ARF). This would have to be compliant with the Health Information Privacy Code 1994.

RHD is not a notifiable condition, and is unlikely to be in the near future. It is important to note however that relying only on ARF notification would not identify a number of Maori and Pacific people with RHD. Furthermore, there is great potential for RHD notification to improve outcomes for people with RHD because, unlike for most notifiable diseases, there is a simple, cheap and proven intervention — secondary prophylaxis.
Screening for rheumatic heart disease

New Zealand criteria for assessing screening programmes are as follows (Table 20):

Table 20. Recommended Elements of a Screening Programme in New Zealand

<table>
<thead>
<tr>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>• The condition is a suitable candidate for screening. The condition should be an important health problem from both an individual and a community perspective. The epidemiology and natural history of the condition, including development from latent to declared disease, should be adequately understood and there should be a detectable risk factor or disease marker and a latent period or pre-symptomatic stage</td>
</tr>
<tr>
<td>• There is a suitable test: safe, simple, reliable, accurate, sensitive, and specific</td>
</tr>
<tr>
<td>• There is an effective and accessible treatment or intervention for the condition identified through early detection. There should be evidence that early treatment leads to better outcomes than late treatment</td>
</tr>
<tr>
<td>• There is high quality evidence, ideally from randomised controlled trials, that a screening programme is effective in reducing mortality or morbidity</td>
</tr>
<tr>
<td>• The potential benefit from the screening programme should outweigh the potential physical and psychological harm (caused by the test, diagnostic procedures and treatment)</td>
</tr>
<tr>
<td>• The health care system will be capable of supporting all necessary elements of the screening pathway, including diagnosis, follow-up and programme evaluation</td>
</tr>
<tr>
<td>• There is consideration of social and ethical issues. There should be evidence that the complete screening programme (identification and invitation, test, diagnostic procedures and treatment/ intervention) is clinically, socially and ethically understood and acceptable to health professionals and the wider public</td>
</tr>
<tr>
<td>• There is consideration of cost-benefit issues</td>
</tr>
<tr>
<td>• When considering and evaluating a prospective screening programme, it is important to consider the direct benefit to participants and any public good benefits that may result</td>
</tr>
<tr>
<td>• Screening programmes need to specifically consider and respond to Māori, if they are to ensure participation by Māori, which is crucial to reducing inequalities in morbidity and mortality in New Zealand.</td>
</tr>
</tbody>
</table>

Source: National Advisory Committee on Health and Disability (2003) 154

In the Māori and Pacific populations in New Zealand, RHD fulfils some of these properties:

- RHD is an important health problem in these populations, with significant mortality, morbidity, social and economic burden. The natural history of RHD is well understood (thanks to classic studies of the 20th Century), with a latent or early symptomatic stage.
- Good adherence to secondary prophylaxis prevents the development or worsening of RHD and leads to disease resolution in many cases.
- Milder valve lesions, which are often asymptomatic and thus the most common lesions that will be detected with screening, are more likely to resolve than more severe lesions in those who adhere to secondary prophylaxis.
- Auscultation and echocardiography could provide appropriate testing tools that are highly sensitive and specific for the disease, as well as being acceptable to the person screened.

The WHO recommends school-based screening for RHD as a tool for estimating the disease burden, and also for identifying cases in areas with a high prevalence of RHD. The ideal method of RHD screening however is not known. The WHO Global Programme on RHD undertook auscultatory screening of over one million children. In some regions, this was augmented by echo to confirm the diagnosis of RHD. The sensitivity of cardiac auscultation is highly dependent on the skill of the operator, and the specificity of auscultation for rheumatic carditis is low. Therefore, the addition of echocardiography to confirm the diagnosis has been proposed.

In New Zealand, a national comprehensive RHD screening programme would not be cost-effective. Any screening for RHD here would have to target high-risk populations in order to improve the pre-test probability. It is possible that auscultatory school-based screening (such as at school-entry, or at age 11 coordinated with the immunisation programme) could discover undetected RHD in these populations. Where echocardiography was not available to review all children with murmurs, a highly experienced auscultator could select all children with non-innocent murmurs for echocardiography (Grade D).
Low school attendance for children of high-risk groups in some areas may influence the effectiveness of such a programme. A pilot programme to estimate the prevalence of undetected RHD in a specified population may be required (Grade D).

**Suggested indicators for evaluation**

Control programmes for ARF/RHD should be evaluated on criteria for routine care and key epidemiological objectives. These include measurement of individual and community adherence to secondary prophylaxis, indicators of satisfactory care specified in best-practice guidelines and rates of disease occurrence, recurrence and mortality.

Further consideration should be given to:

- assessing the delivery of specialist cardiology services
- availability and accessibility of echocardiography
- referral practices and structures
- transportation for cases
- support structures and appropriate follow-up processes.

As has been highlighted throughout the developing world, the availability of and support for routine health care is essential to controlling ARF/RHD. Indicators used to evaluate ARF/RHD control programmes should be relevant, structured, measurable, routinely available and affordable. In particular, they should not overburden health care providers and should lead to improved clinical results. A list of suggested indicators is provided in Table 21 (Grade D).

**Table 21. Proposed Indicators for Evaluating ARF/RHD Control Programmes**

<table>
<thead>
<tr>
<th><strong>Secondary prophylaxis</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>The proportion of scheduled BPG injections delivered in the previous 12 months</td>
</tr>
<tr>
<td>Individual, community and regional figures, expressed as:</td>
</tr>
<tr>
<td>Median percentage of doses delivered</td>
</tr>
<tr>
<td>Proportion who receive 80% or less of scheduled doses</td>
</tr>
<tr>
<td>Proportion who receive 50% or less of scheduled doses</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Medical review</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Proportion of registered individuals who are more than 3 months overdue for specialist or other medical review, as defined by local guidelines</td>
</tr>
<tr>
<td>Proportion of individuals who have echocardiography performed within 3 months of scheduled timing</td>
</tr>
<tr>
<td>Median time elapsed between recommendation and performance of valvular surgery</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Epidemiology</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Yearly (or other appropriate time frame) age-specific incidence rates of ARF</td>
</tr>
<tr>
<td>Proportion of ARF episodes in the register classified as recurrences</td>
</tr>
<tr>
<td>Rates of ARF recurrence per 100 patient-years</td>
</tr>
<tr>
<td>Number of deaths and age-standardised rates of mortality due to ARF/RHD in the previous 12 months (or other appropriate time frame)</td>
</tr>
<tr>
<td>Yearly age-specific and overall point prevalence of RHD</td>
</tr>
<tr>
<td>Proportion of ARF cases notified to and recorded by public health authorities in the previous 12 months (or other appropriate time frame)</td>
</tr>
<tr>
<td>Proportion of newly registered individuals with an initial diagnosis being established of RHD (rather than ARF).</td>
</tr>
</tbody>
</table>
There are a number of driving forces that will assist the implementation of this guideline. There is national practitioner demand for standardisation of the diagnosis of ARF in order to minimise over- and under-diagnosis and ensure that the high-risk populations receive appropriate care. There is also demand for effective and cost-effective management and avoidance of ARF recurrence and subsequent disabling RHD. Restraining forces that have the potential to hinder the implementation of this guideline include: reduced access of cases to diagnostic tests and specialist services, limited resources available, reluctance of practitioners to change current practice, incomplete understanding of ARF amongst primary and secondary care professionals and inconsistent access to certain treatments including BPG.

**Suggested implementation strategies include:**

**Streamlined processes for the diagnosis, management and prevention of ARF**
- Consistent New Zealand standards for ARF diagnosis
- Consistent standards for streptococcal serology methodology, reporting and assay between laboratories.

**Provision of streamlined specialist services**
- Where possible, regions have the opportunity for regular specialist rheumatic fever clinics (potentially involving both paediatric and medical input, in close association with available cardiology services). These should coordinate with rheumatic fever registers, the community services involved in BPG delivery and with primary care providers (particularly Maori and Pacific). This has the potential for reducing cases that are lost to follow-up and to secondary prophylaxis and therefore reduce rheumatic fever recurrence, hospitalisations and RHD.

**Ensure funding for training**
- Maintain echocardiography standards for ARF and training of echo technicians in all main centers of ARF prevalence.

**Education**
- Professional education targeting both primary and secondary care providers, doctors, nurses, dentists, pharmacy, medical and nursing students
- Increased understanding in primary care of early management, and the need for hospitalisation in ARF.

**Community awareness and health promotion**
- Raise awareness, especially in families and communities at high risk, of the “sore throats do matter” message and of the signs and symptoms of ARF.

**Ensure regular supply of benzathine**
- The supply of BPG has been inconsistent, with occasional periods where no BPG was available. These guidelines provide the opportunity to discuss with PHARMAC the means of ensuring an uninterrupted supply of BPG, including the possibility of having an alternative supplier.
- Discussions with hospitals pharmacies on storing back-up supplies of BPG could ensure a contingency plan should supplies run out.

**Resource and support for a local ARF register in each area**
- Professional leadership
- Adequate administrative support.

**Case follow-up**
- Because a number of ARF cases (particularly in Auckland) involve Pacific people, there is opportunity for greater links to be forged between New Zealand and the Pacific Islands. Key contacts on each Pacific Island need to be identified. They should be able to access information on New Zealand registers and provide reciprocal information to the New Zealand registers. This will improve the continuity of prophylaxis therapy and care for cases that travel between these countries.
• In addition, there should be continued support for the outreach capacity of primary care providers in order to reduce the number of cases that are non-compliant or do not present for prophylaxis.

**Dissemination of guidelines**

It is hoped that this guideline will be used widely. The following are suggestions for dissemination of this guideline:

- The National Heart Foundation of New Zealand through printed resources, including this guideline and web-based information
- The Cardiac Society of Australia and New Zealand (CSANZ), specifically the launch of the guidelines at the CSANZ meeting in Auckland in May, 2006
- Dissemination by members of the writing group, reviewers and contributors and the endorsing organisations
- Production and distribution of an additional resource consisting of the algorithms from this and future guidelines
- Published articles
- Health promotion initiatives and discussion of this guideline in regions of relatively high prevalence of ARF.
15. Algorithms

Algorithm 1: Guide for the diagnosis of ARF

Note: this algorithm is to help in the decision making process for the diagnosis of ARF with a clinical presentation of a major criteria. The investigations and observations are not intended to be in chronological order, and the clustering of symptoms and signs presented are commonly more complex.

**Chorea** (Can be stand-alone for ARF diagnosis)

- Clinical carditis
  - Yes
    - Definite ARF
    - If no other diagnosis definite ARF
  - No
    - Echo
      - carditis
        - Yes
          - Definite ARF
        - No
          - Explore alt. diagnosis and repeat echo
      - No carditis

**Arthritis** (Polyarthritis or mono with NSAID)

- Echo
  - ARF carditis
    - No carditis
      - Definite ARF
      - Explore alt. serology for arthritis and do strep. serology
        - +ve GAS and no alt. dx.
          - Definite ARF
        - -ve GAS and no alt. dx.
          - Minor criteria present
            - Definite ARF
            - Unlikely ARF. Consider follow-up
  - No carditis

**Carditis** (Audible murmur and/or heart failure)

- Echo
  - ARF carditis
    - No carditis
      - Definite ARF
      - Consider further echo (algorithm 2)
      - Other major
        - Yes
          - Definite ARF
        - No
          - Strep. serology
            - +ve
              - Minor criteria present
                - RHD
            - -ve
              - Probable ARF

Possible ARF if no other diagnosis and high-risk group
Repeat echo at 2-4 weeks, repeat serology, consider prophylaxis
Note also that cases can fulfill the Jones criteria but not have ARF. Discussion with an experienced clinician, with reference to the full guideline is recommended.

* Echo here refers to fulfilling the echocardiographic criteria of ARF. See guidelines for further details.

Abbreviations:
alt. = alternative
ARF = acute rheumatic fever
dx = diagnosis
Echo = echocardiogram

GAS = group A streptococcus
mono = monoarthritis
NSAID = non-steroidal anti-inflammatory drug
RHD = rheumatic heart disease
Strep. = streptococcus
Algorithm 2: Guide for the use of echocardiography in ARF

Any person with suspected ARF and a cardiac murmur, or any case of chorea, should have an **echocardiogram** shortly after admission to hospital.

- **Equivocal**
  - Normal
  - Repeat at 2-4 weeks
- **Abnormal**
  - Tables 6 & 7

Pursue alternative diagnoses
- Tables 8 & 9

**Second echocardiogram** at 2-4 weeks if no alternative diagnosis. A second echo is usually unnecessary with a presentation of chorea.

- **Normal**
- **Abnormal**
  - Tables 6 & 7

**Second echocardiogram** at 4-6 weeks if:
- Signs progress
- Medication commenced
- Recommended by cardiologist

- **Normal**
- **Abnormal**
  - Tables 6 & 7
Algorithm 3: Guide for the duration of secondary prophylaxis

New Zealand standard recommendations are for four-weekly (28-day) IM BPG prophylaxis. A 21-day schedule of prophylaxis is recommended only for those cases with ARF recurrence while compliant with the four-weekly schedule. Refer to the text of the guideline for further details.

![Algorithm 3](image)

* See Table 2 for definitions of possible, probable and definite ARF

** It is recommended that cases with established valvular disease have regular dental care and follow the guidelines for endocarditis prophylaxis

*** Individuals working or living with children, or in a living situation where there is overcrowding or close proximity to others (such as boarding schools, barracks, and hostels) have a higher risk of exposure to GAS and subsequent development of ARF.
16. References


34. Strasser T. Cost-effective control of rheumatic fever in the community. Health Policy. 1985; 5: 159-164.


88. Lessof MH, Bywaters EG. The duration of chorea. BMJ. 1956; 1520-1523.


158. Committee of Rheumatic Fever and Bacterial Endocarditis of the American Heart Association. Jones criteria (revised) for guidance in the diagnosis of rheumatic fever. Circulation. 1984; 69; 204A-08A.


Appendix A: Guideline development process

- Relevant literature regarding ARF was identified primarily using computerised Medline, CINAHL, ProQuest and other databases. Publications were limited to those in the English language. Articles found through this methodology were then searched for relevant information and further articles identified through bibliographic references. A substantial physical library of ARF references held at the School of Population Health was also reviewed for key articles. In addition to journal article searches, regular review and searches were made of internet sites such as the World Health Organisation, New Zealand Ministry of Health, New Zealand Environment Scientific Research (ESR) and the New Zealand Department of Statistics.

- In 2005, a steering group which arose out of the New Zealand members of the writing group for the Australian guidelines met and agreed to develop the New Zealand version of guidelines for the diagnosis, management and prevention of ARF.

- A writing group comprising experts in the area reviewed the Australian draft and reached consensus on areas of disagreement.

- Selected individuals re-wrote the Australian guidelines for the New Zealand context, and according to the outline recommended by the New Zealand Guidelines Group (NZGG).

- Members of the writing group with experience in ARF/RHD diagnosis, management, and prevention then reviewed each chapter and their suggestions were incorporated into a second draft.

- The revised draft was widely distributed to a range of stakeholders, who were then invited to comment.

- The stakeholders reviewed the draft and reached consensus on areas of disagreement.

- The comments were then incorporated into a final draft, which was endorsed by the stakeholders.
Appendix B: Jones criteria for the diagnosis of ARF

There is no single symptom, sign, or laboratory test that is diagnostic for ARF. The Jones criteria were introduced in 1944. Major manifestations (least likely to lead to an incorrect diagnosis) at that time included carditis, joint symptoms, subcutaneous nodules and chorea. Historical evidence of ARF or RHD was also a major manifestation. Minor manifestations (suggestive, but not sufficient for the diagnosis) included clinical signs such as fever, erythema marginatum, and abdominal pain and laboratory markers of inflammation such as ESR and leukocytosis. Since a previous history of ARF was considered a major criterion, cases only needed minor manifestations in order to fulfill the diagnosis (one major and two minor).21,28

In order to improve specificity, in 1956 arthritis replaced joint symptoms as a major manifestation, and erythema marginatum was reconsidered as a major manifestation. A preceding ARF or RHD was reclassified as a minor manifestation, and other minor manifestations of arthralgia, and evidence of a preceding GAS infection were added.29 In subsequent revisions in 1965 and 1984, evidence of a GAS infection was considered essential.28,158,159

<table>
<thead>
<tr>
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<th></th>
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<th></th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Carditis</td>
<td></td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
<tr>
<td>Long PR†</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Arthritis</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Arthralgia</td>
<td></td>
<td></td>
<td>✓</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Subcutaneous nodules</td>
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<td></td>
<td></td>
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<tr>
<td>Chorea</td>
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<td></td>
</tr>
<tr>
<td>Erythema marginatum</td>
<td></td>
<td></td>
<td>✓</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-existing RF/RHD</td>
<td></td>
<td></td>
<td>✓</td>
<td></td>
<td></td>
<td>✓</td>
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<tr>
<td>Fever</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>WBC, ESR, CRP a</td>
<td></td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>Epistaxis, abdominal pain, anemia, pulmonary findings</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Recent streptococcal infection</td>
<td></td>
<td></td>
<td>✓</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>


† PR = PR interval in the electrocardiogram; WBC = leukocytosis; ESR = erythrocyte sedimentation rate; CRP = C-reactive protein.
The current Jones Criteria (1992)\textsuperscript{27} are designed to establish the diagnosis of the initial attack of ARF and a previous history of ARF or RHD is excluded from the list of minor manifestations. The sensitivity of ARF arthritis to NSAIDs and salicylates, and therefore the potential for the use of these medications to aid in diagnosis, is described. In addition, the 1992 criteria define three circumstances in which the diagnosis of ARF can be made without strictly adhering to the Jones criteria.

These are:
- chorea occurring as the only manifestation of ARF
- indolent carditis occurring as the only manifestation of ARF
- a presumptive diagnosis of rheumatic fever recurrence may be made when a single major or several minor manifestations are present in a patient with a reliable history of ARF or established RHD, provided there is evidence of a recent GAS infection.\textsuperscript{27}

The Jones criteria Working Group met again in 2000 to review the adequacy of existing guidelines for the diagnosis of the initial attack of ARF. The consensus opinion at this time was that no new version of the criteria was justified. It was reiterated that the epidemiological setting where diagnosis is being made is important, and that strict adherence to the Jones criteria in areas of high prevalence may result in under-diagnosis.\textsuperscript{160} This group determined that echocardiography is useful for confirming clinical findings, assessing severity of valvular disease, chamber size and ventricular function, and noting the presence and size of pericardial effusions. Echocardiography was also noted to be useful for the management of ARF, and to exclude ARF as a cause of murmur. However, the use of echocardiography in the diagnosis of ARF was determined by this working group to be too controversial to classify as a major or minor criterion. Controversy arose because of "normal" valvular regurgitation (which increases with age), regurgitation with febrile illnesses unrelated to ARF, and the uncertainty over the long-term prognostic significance of echocardiography.\textsuperscript{160}
Appendix C: Use of echocardiography in ARF

Echocardiography is now recommended for all suspected cases of ARF. The uses of echocardiography in ARF are presented in Table 22.

Table 22. Uses of Echocardiography in ARF

<table>
<thead>
<tr>
<th>DIAGNOSIS</th>
<th>PERICARDITIS</th>
<th>MYOCARDITIS AND CONGESTIVE HEART FAILURE</th>
<th>VALVULITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Echocardiographic evidence of subclinical carditis is sufficient as a major manifestation of ARF</td>
<td>Confirming the presence of a pericardial effusion</td>
<td>Defining left ventricular function</td>
<td>Visualisation of anatomy of the valves, especially in mitral regurgitation. This is paramount in surgical decision-making</td>
</tr>
<tr>
<td></td>
<td>Revealing inaudible or subclinical valvular regurgitation in presence of a friction rub</td>
<td>Confirming the severity of valvulitis (valvulitis is usually present in ARF with heart failure)</td>
<td>Defining the severity of mitral, aortic and/or tricuspid regurgitation</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Defining the severity of mixed valve disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Identifying subclinical evidence of rheumatic valve damage.</td>
</tr>
</tbody>
</table>

The anatomy and physiology of ARF as shown by echocardiography M-mode and 2-dimensional echocardiography (2DE) are used in evaluating chamber size and ventricular function. More complex formulae based on 2DE can also be used to calculate left ventricular function (e.g. single plane ellipse and Simpson’s methods of discs). 2DE allows visualisation of the functional anatomy of acute mitral regurgitation. The degree of annular dilatation is easily shown by relating annular size to body surface area. Mitral valve prolapse is a frequent finding with greater degrees of mitral regurgitation. Chordal elongation and sometimes chordal rupture may occur in the presence of significant valve prolapse.

Valvular regurgitation can be accurately graded with pulsed and colour Doppler echocardiography as nil, physiological, mild, moderate and severe for both rheumatic and non-rheumatic valve disease. Colour Doppler echocardiography shows the direction of the regurgitant jet, which is directed posteriorly with anterior mitral valve leaflet prolapse, and anteriorly with the less common posterior leaflet prolapse.

Echocardiography and physiological valvular regurgitation

Trivial valvular regurgitation is commonly detected on echocardiography as a normal finding. It can now be readily distinguished from pathological regurgitation. First, valve closure is associated with physiological displacement of a small amount of blood, the closing volume, which is detectable by colour flow Doppler imaging. Second, true regurgitant jets, albeit trivial in nature, may be observed in normal individuals of all ages. These leaks extend beyond the valve coaptation point, but usually by only 1cm or less. They may have a high velocity component, generally for only part of systole or diastole.
Trivial right-sided regurgitation is very common, but trivial aortic regurgitation is uncommon, occurring in 0-1% of normal subjects, except in one study where closing volumes were included. The characteristic Doppler echocardiographic feature of trivial mitral regurgitation in normal subjects is an aliasing flow pattern in early systole, with a velocity usually <1m/s. One study reported holosystolic flow signals, but they were recorded only at the valve leaflets, and had a poorly defined spectral envelope. Sometimes a brief high velocity component may be detected.

Subclinical evidence of rheumatic valve damage

In those with suspected ARF and a murmur, reliance on clinical findings alone may result in misclassification of carditis. Some cases have been shown on echocardiography to have a physiological or flow murmur, or even congenital heart disease. The likelihood of misclassification has increased in recent years, as physicians’ auscultatory skills have become less proficient. There is convincing evidence that subclinical or silent rheumatic valve damage detected by echocardiography is part of the spectrum of rheumatic carditis and should not be ignored. This has been confirmed by investigators in many regions around the world with high rates of rheumatic fever, including New Zealand, Australia, USA, Qatar, Brazil, Turkey, Chile, Tahiti, Nepal, Portugal, Egypt and India. A single report from India describing 28 patients with polyarthritis or chorea failed to detect any subclinical carditis. In experienced hands, subclinical rheumatic valve damage can usually be differentiated on echocardiography from physiological regurgitation. A World Health Organisation expert committee concurred that subclinical rheumatic valve damage exists. However, because the clinical significance of this finding is not yet known, they decided against recommending its inclusion in the Jones criteria. In the opinion of the authors of this review, echocardiographic diagnosis of subclinical valve damage can help experienced clinicians in making the diagnosis of ARF, or in confirming the presence of carditis in cases of ARF without an obviously pathological heart murmur. Therefore, it is recommended that echocardiographically suggested valve damage (subclinical or otherwise), diagnosed by a clinician with experience in echocardiography of patients with ARF/RHD, be included as a major manifestation (Table 3) (Level IV, Grade C).

A World Health Organisation expert committee concurred that subclinical rheumatic valve damage exists. However, because the clinical significance of this finding is not yet known, they decided against recommending its inclusion in the Jones criteria. In the opinion of the authors of this review, echocardiographic diagnosis of subclinical valve damage can help experienced clinicians in making the diagnosis of ARF, or in confirming the presence of carditis in cases of ARF without an obviously pathological heart murmur. Therefore, it is recommended that echocardiographically suggested valve damage (subclinical or otherwise), diagnosed by a clinician with experience in echocardiography of patients with ARF/RHD, be included as a major manifestation (Table 3) (Level IV, Grade C).

Subclinical valve damage influences the diagnosis of ARF in relatively few individuals. Most cases have either migratory polyarthritis, or clinically overt carditis that can be confirmed by echocardiography. However, there are some cases in which the finding may help to confirm the diagnosis, and to reinforce in the minds of cases and their families the importance of adherence to a secondary prophylactic regimen (Table 23).

Table 23. Diagnostic and Clinical Utility of Subclinical Rheumatic Valve Damage in ARF
### Appendix D: Medications used in ARF

#### Table 24. Medications Used in ARF

<table>
<thead>
<tr>
<th>MEDICATION</th>
<th>INDICATION</th>
<th>REGIMEN</th>
<th>DURATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benzathine penicillin G IM or Penicillin V PO</td>
<td>Treat streptococcal infection</td>
<td>900mg (1,200,000 U) &gt;20kg 450mg (600,000 U) ≤20kg</td>
<td>Single dose</td>
</tr>
<tr>
<td>Erythromycin ethyl succinate PO</td>
<td>Penicillin V PO (250mg bd 10 days)</td>
<td>40mg/kg per day in 2-4 divided doses maximum 1g/day (children) 400mg bd (adolescents and adults)</td>
<td>10 days</td>
</tr>
<tr>
<td>Paracetamol PO</td>
<td>Arthritis or arthralgia - mild or until diagnosis confirmed</td>
<td>60mg/kg/day (max 4g) given in 4-6 doses/day. May increase to 90mg/kg/day if needed, under medical supervision</td>
<td>Until symptoms relieved or NSAID started</td>
</tr>
<tr>
<td>Codeine PO</td>
<td>Arthritis or arthralgia until diagnosis confirmed</td>
<td>0.5-1.0mg/kg/dose (adults 15-60mg/ dose) 4-6h</td>
<td>10 days</td>
</tr>
<tr>
<td>Aspirin PO</td>
<td>Arthritis or severe arthralgia (when ARF diagnosis confirmed)</td>
<td>80-100mg/kg/day (4-8 g/d in adults) given in 4-5 doses/day Reduce to 60-70mg/kg/day when symptoms improve Consider ceasing in the presence of acute viral illness, and consider influenza vaccine if administered during autumn/winter</td>
<td>Until joint symptoms relieved</td>
</tr>
<tr>
<td>Naproxen PO</td>
<td>Arthritis (if aspirin-intolerant)</td>
<td>10-20mg/kg/day (max 1250mg) given bd</td>
<td>As for aspirin</td>
</tr>
<tr>
<td>Prednisone or Prednisolone PO</td>
<td>Severe carditis, heart failure, pericarditis with effusion</td>
<td>1-2mg/kg/day (max 80mg). If used &gt;1 week, taper by 20-25% per week.</td>
<td>Usually 1 to 3 weeks</td>
</tr>
<tr>
<td>Frusemide PO/IV (can also be given IM)</td>
<td>Heart failure</td>
<td>Children: 1-2mg/kg stat, then 0.5-1mg/kg/dose 6-24 hrly (max 6mg/kg/dose) Adults: 20-40mg/dose 12-24 hrly up to 250-500mg/day</td>
<td>Until failure controlled and carditis improved</td>
</tr>
<tr>
<td>MEDICATION</td>
<td>INDICATION</td>
<td>REGIMEN</td>
<td>DURATION</td>
</tr>
<tr>
<td>------------------</td>
<td>-----------------------------</td>
<td>-------------------------------------------------------------------------</td>
<td>------------------------------------</td>
</tr>
<tr>
<td>Spironolactone PO</td>
<td>Heart failure</td>
<td>1-3mg/kg/day (max 100-200mg/day) in 1-3 doses. Round dose to multiple of 6.25mg (quarter of a tab)</td>
<td>As for frusemide</td>
</tr>
</tbody>
</table>
| Enalapril PO     | Heart failure               | Children: 0.1mg/kg/day in 1-2 doses increased gradually over 2 wks to max of 1mg/kg/day in 1-2 doses  
|                  |                             | Adults Initial: 2.5mg daily Maintenance: 10-20mg daily (max 40mg)       | As for frusemide                   |
| Lisinopril PO    | Heart failure               | Children: 0.1-0.2mg/kg once daily up to 1mg/kg/dose                     | As for frusemide                   |
|                  |                             | Adults: 2.5-20mg once daily (max 40mg/day)                               |                                    |
| Digoxin PO/IV    | Heart failure/atrial fibrillation | Children: 15mcg/kg stat and then 5mcg/kg after 6 hrs, then 3-5 mcg/kg/dose (max 125 mcg) 12-hourly.  
|                  |                             | Adults: 125-250 mcg daily                                               | Seek advice from specialist        |
| Carbamazepine PO | Severe chorea               | 7-20mg/kg/day (7-10mg/kg/day usually sufficient) given tds.             | Until chorea controlled for several weeks, then trial off medication |
| Valproic acid PO | Severe chorea (may affect salicylate metabolism) | Usually 15-20mg/kg/day (can increase to 30mg/kg/day) given tds          | As for carbamazepine.              |
Appendix E: Comparison of intramuscular penicillin and oral penicillin for secondary prevention

A search was conducted by Manyemba and Mayosi (2002). The search strategy included the Controlled Trials Register (Cochrane Library Issue 2, 2001), Medline (January 1996 to July 2000), Embase (January 1985 to July 2000), reference lists of articles and consultation with experts.

Randomised and quasi-randomised studies comparing: (i) oral with intramuscular penicillin; and (ii) two-weekly or three-weekly with four-weekly intramuscular penicillin in patients with previous ARF. Two reviewers independently assessed the trial quality and extracted the data of six included studies (1,707 patients).

Four trials (1,098 patients) compared IM with oral penicillin and all showed that IM penicillin was more effective than oral in reducing recurrence of ARF and streptococcal throat infections.

One trial compared two-weekly with four-weekly IM penicillin. Penicillin given every two weeks was better at reducing ARF recurrence (relative risk (RR) 0.52, 95% confidence interval (CI) 0.33-0.83) and streptococcal throat infections (RR 0.60, 95% CI 0.42-0.85).

One trial (249 patients) showed that three-weekly IM penicillin injections were more effective than four-weekly IM penicillin at reducing streptococcal throat infections (RR 0.67, 95% CI 0.48-0.92).

The conclusions made therefore were that IM penicillin seemed to be more effective than oral penicillin in preventing ARF recurrence and streptococcal throat infections. Two-weekly or three-weekly injections appeared to be more effective than four-weekly injections. However, the evidence was based on poor-quality trials and the use of outdated formulations of oral penicillin.
Appendix F: Anaphylaxis recognition and management

The signs and symptoms of an anaphylactic reaction include: rapid weak pulse, wheeze, tightness in chest, pruritis, urticaria, giddiness or headache, flushing and/or periorbital oedema.

Response procedure:
- do not leave the patient alone
- call for assistance
- lie patient in recovery position (may be better sitting up if severe respiratory distress)
- ensure airway is clear, apply oxygen if available
- give adrenaline (Table 25)
- ring 111 for ambulance
- check vital signs, note colour, tone and perfusion
- if signs of further deterioration, repeat adrenaline after 10 minutes
- up to 3 doses of adrenaline can be given.

Adrenaline dosage:

Table 25. Recommended Dose of Adrenaline in Anaphylaxis

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Adrenaline Dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>12 YEARS OF AGE AND OVER</td>
<td>0.5ml of 1:1000 adrenaline, deep IM injection</td>
</tr>
<tr>
<td>UNDER 12 YEARS OF AGE</td>
<td>Approximately 0.01ml/kg of 1:1000 adrenaline, deep IM injection</td>
</tr>
<tr>
<td>Age 0-3 years:</td>
<td>0.1ml</td>
</tr>
<tr>
<td>Age 4-6 years:</td>
<td>0.2ml</td>
</tr>
<tr>
<td>Age 6-8 years:</td>
<td>0.3ml</td>
</tr>
<tr>
<td>Age 9-12 years:</td>
<td>0.4ml</td>
</tr>
</tbody>
</table>

* Up to 3 doses of adrenaline can be given
Appendix G: Protocol for follow-up of non-compliant cases

Case is non-compliant with injections on 3-4 concurrent occasions. All attempts at contact are clearly documented in the patient's file. **These attempts should include the use of multiple modalities for contact including telephone calls, visits, texting and the use of the local knowledge of community health workers.**

Discuss with primary nurse and refer to community health worker, public health nurse, or other community staff as fitting in the area for follow up. Note also opportunity to involve staff from Māori or Pacific primary health providers, if appropriate.

Community health worker (or other community staff responsible) follows up with case (and family) to determine reason for non-compliance. Where necessary and appropriate, provides ongoing support, education, and arranges appointments for review at outpatient clinic.

If compliance is no longer a problem, continue routine secondary prophylaxis.

If non-compliance continues, letter of planning to discharge is copied to the case, case file, and GP after discussion with primary nurse and community health worker.

Case file goes “on hold” for up to six months (local area policy may suggest regular attempts at contact while case is on hold).

At the end of the holding period, the primary nurse and community health worker review the case and if considered appropriate a discharge letter is to be sent to the case, with a copy to the patient file, GP, and rheumatic fever register (if available).
Appendix H: Wallet card for infective endocarditis prevention

**Information For Patient/Parents/Guardians**

Has a heart disorder and therefore needs antibiotic protection to be given before some of the procedures that dentists and doctors may need to do.

**YOU MUST SHOW THIS CARD TO ANY DENTIST/DENTAL THERAPIST OR DOCTOR BEFORE TREATMENT IS STARTED.**

**General Advice**

1. Regular teeth cleaning and avoiding sugary foods and drinks will reduce the need for dental surgery.
2. Regular dental check ups will help keep teeth healthy.

**HOSPITAL CHECK UPS DO NOT REPLACE VISITS TO YOUR LOCAL DENTIST/DENTAL THERAPIST.**

3. Using a mouth guard for contact sports will help protect teeth.
4. Antibiotics are not needed for natural loss of baby teeth.

**Information for Doctor/Dentist/Dental Therapist**

This patient is at risk of bacterial endocarditis and requires prophylaxis as detailed below. Antibiotic prophylaxis is necessary for all procedures involving manipulation/bleeding of the gingival tissues and any instrumentation through the apex of the tooth.

**Dental/Oral/Respiratory Tract/Oesophageal Procedures**

Patients who have not received Penicillin or Cephalosporin in the last two weeks and are not on long term Penicillin:

1. **Children Under 10 years**
   - Amoxycillin 250 mg in 5 ml, oral suspension 50 mg/kg (max 2 g) one hour prior to procedure then, 25 mg/kg (max 1 g) six hours later.

2. **Patients with Penicillin allergy or treated with Penicillin or Cephalosporin within the last two weeks, or on long term Penicillin prophylaxis Adults and Children Over 10 years**
   - Clarithromycin tab 500 mg orally one hour prior to procedure. A single dose only is required.

**Genitourinary and Gastrointestinal (excluding oesophageal) procedures**

For standard risk patients who have not received Penicillin/Cephalosporin in the last two weeks and are not on long term Penicillin, Amoxycillin as per previous dosages. For all other patients including high risk, discuss with Paediatrician/Physician/Cardiologist.

**INFEKTIVE ENDOCARDITIS PROPHYLAXIS**

- **Name:**
- **NHI:**
- **Diagnosis:**
- **GP:**
- **Hospital Doctor:**

**Standard Risk**

- **Children Under 10 years**
  - Amoxycillin 250 mg in 5 ml, oral suspension 50 mg/kg (max 2 g) one hour prior to procedure then, 25 mg/kg (max 1 g) six hours later.

- **Children Under 10 years**
  - Clarithromycin 125 mg/5 ml oral liquid 15 mg/kg (max 500 mg) one hour prior to procedure. A single dose only required.

- **Patients with Penicillin allergy or treated with Penicillin or Cephalosporin within the last two weeks, or on long term Penicillin prophylaxis Adults and Children Over 10 years**
  - Clarithromycin tab 500 mg orally one hour prior to procedure. A single dose only is required.

- **Dental/Oral/Respiratory Tract/Oesophageal Procedures**
  - Adults and Children Over 10 years
    - Clarithromycin tab 500 mg orally one hour prior to procedure.

- **Children Under 10 years**
  - Amoxycillin 250 mg in 5 ml, oral suspension 50 mg/kg (max 2 g) one hour prior to procedure then, 25 mg/kg (max 1 g) six hours later.

- **Children Under 10 years**
  - Clarithromycin 125 mg/5 ml oral liquid 15 mg/kg (max 500 mg) one hour prior to procedure. A single dose only required.

- **Patients with Penicillin allergy or treated with Penicillin or Cephalosporin within the last two weeks, or on long term Penicillin prophylaxis Adults and Children Over 10 years**
  - Clarithromycin tab 500 mg orally one hour prior to procedure. A single dose only is required.

- **Dental/Oral/Respiratory Tract/Oesophageal Procedures**
  - Adults and Children Over 10 years
    - Clarithromycin tab 500 mg orally one hour prior to procedure.

**High Risk**

- **Children Under 10 years**
  - Amoxycillin 250 mg in 5 ml, oral suspension 50 mg/kg (max 2 g) one hour prior to procedure then, 25 mg/kg (max 1 g) six hours later.

- **Patients with Penicillin allergy or treated with Penicillin or Cephalosporin within the last two weeks, or on long term Penicillin prophylaxis Adults and Children Over 10 years**
  - Clarithromycin tab 500 mg orally one hour prior to procedure. A single dose only is required.

- **Dental/Oral/Respiratory Tract/Oesophageal Procedures**
  - Adults and Children Over 10 years
    - Clarithromycin tab 500 mg orally one hour prior to procedure.

- **Children Under 10 years**
  - Amoxycillin 250 mg in 5 ml, oral suspension 50 mg/kg (max 2 g) one hour prior to procedure then, 25 mg/kg (max 1 g) six hours later.

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  - Clarithromycin tab 500 mg orally one hour prior to procedure. A single dose only is required.

- **Dental/Oral/Respiratory Tract/Oesophageal Procedures**
  - Adults and Children Over 10 years
    - Clarithromycin tab 500 mg orally one hour prior to procedure.
18. Glossary

2DE .................................................. 2-dimensional echocardiography
alt. .................................................. alternative
ANA .................................................. anti nuclear antibody
anti-DNase B ................................. antideoxyribonuclease B
ARF .................................................. acute rheumatic fever
ASO .................................................. antistreptolysin O
BP ..................................................... blood pressure
BPG .................................................. benzathine penicillin G
CRP .................................................. C-reactive protein
CSANZ .......................... Cardiac Society of Australia and New Zealand
ECG .................................................. electrocardiogram
Echo .............................................. echocardiography
ESR .................................................. erythrocyte sedimentation rate
GAS .................................................. group A streptococcus
HR ................................................... heart rate
IM ..................................................... intramuscular
INR .................................................. international normalised ratio
IV ..................................................... intravenous
mU .................................................. megaunits
NHF .............................................. The National Heart Foundation of New Zealand
NHI .................................................. National Hospital Index
NSAID ........................................... non-steroidal anti-inflammatory drug
PANDAS ...................................... paediatric auto-immune neuropsychiatric disorders associated with streptococcal infections
PO .................................................. per oral
RAST ............................................. RadioAllergoSorbent Test
RHD .................................................. rheumatic heart disease
ULN .................................................. upper limits of normal
WHO ............................................. World Health Organisation
19. Notes
Cardiovascular disease is the leading cause of death in New Zealand, accounting for 40 percent of all deaths annually (approx. 10,500 people).

Since its inception in 1968, the Heart Foundation has played a major role in reducing the high incidence of death from cardiovascular disease, including:

• Funding vital heart-related medical and scientific research in New Zealand
• Working with at-risk groups through intervention programmes
• Supporting and implementing cardiac rehabilitation programmes
• Working with food industry groups to promote healthier foods
• Providing education programmes promoting healthy eating and physical activity
• Providing heart health resources to health professionals and the general public
• Working with Pacific people through Pacific Islands Heartbeat (PIHB).

Without the generosity of New Zealanders’ donations and legacies, the Heart Foundation could not achieve many of these goals. Any help you can give is greatly appreciated.

For more information on heart health and/or supporting the Heart Foundation, visit our website www.heartfoundation.org.nz or please contact:

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Tel: 0064 9 571 9191
Fax: 0064 9 571 9190
Email: info@nhf.org.nz

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