THE NEW ZEALAND MEDICAL JOURNAL
Journal of the New Zealand Medical Association

Rheumatic fever diagnosis, management, and secondary prevention: a New Zealand guideline

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Abstract

Aim The National Heart Foundation of New Zealand, and the Cardiac Society of Australia and New Zealand (CSANZ) recently launched an evidence-based review and guideline entitled *New Zealand Guideline for Rheumatic Fever Diagnosis, Management, and Secondary Prevention*. This paper is a brief summary.

Method This Guideline was developed by a writing group comprising experts in the area. Relevant literature was identified and reviewed, and the Australian guideline for rheumatic fever and rheumatic heart disease was reviewed and adapted for the New Zealand context. A peer review and stakeholder consultation process followed the development of the draft document.

Results The final draft of the New Zealand guideline was endorsed by Te Hotu Manawa Māori, Pacific Islands Heart Beat, The Paediatric Society of New Zealand, and the Rheumatic Fever Trust of New Zealand—plus approved by a number of organisations including the Royal Australasian College of Physicians, the Australasian Society for Infectious Disease, the Pasifika Medical Association, and Te Ohu Rata o Aotearoa. Two subsequent New Zealand guidelines for rheumatic fever: *Sore Throat Management* and *Primary Prevention* are also in production. The complete guideline, and associated summary algorithms, can be downloaded from www.nhf.org.nz

Conclusion A New Zealand Guideline for Rheumatic Fever Diagnosis, Management and Secondary Prevention of Acute Rheumatic Fever should result in improved consistency in the approach to this disease, and reduced mortality and morbidity from acute rheumatic fever and rheumatic heart disease.

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial pharyngeal infection with group A streptococcus (GAS) which can lead to lasting damage to heart valves—mainly mitral and aortic valvulitis. This is known as rheumatic heart disease (RHD) and is an important cause of premature mortality. Almost all cases of RHD and associated deaths are preventable.[3]

In most affluent populations, ARF is now rare. By contrast, 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.[4–6] Almost all cases and deaths occur in developing countries and RHD is the most frequent form of heart disease in children worldwide.[7] RHD is a significant cause of premature death in New Zealand.[8–10]

In the 1920s, surveys of school records in New Zealand determined an approximate annual school population incidence of ARF of 65 per 100,000.[4] 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 socioeconomic deprivation had significantly higher rates of both ARF and RHD.[11] 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.[10]

In New Zealand, the rates of ARF in Māori and Pacific people (mostly of Samoan, Tongan, Niuean, or Cook Islands origin) have always been reported as significantly greater than those seen in non-Māori (i.e. European, Asian). 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.[4] 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.[10]

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 penicillin prophylaxis delivery) recurrence rates in Māori were 40% compared to 22% in non-Māori.[12] Data from the Auckland Rheumatic Fever Register shows that although the recurrence rates dropped significantly from 22% in the 1980s to 5.5% by 1999, all recurrences were in Māori and Pacific people.[13] 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.

There is no evidence to support Māori and Pacific people having an increased genetic susceptibility to rheumatic fever. It is more likely that their increased burden reflects social, political, and economic influences that result in overcrowded conditions, socioeconomic deprivation, an increased incidence of upper respiratory infections with GAS, and different options or opportunities for appropriate and effective health care.[6,14,15]

There are considerable personal, community, and national costs associated with this burden of ARF and RHD. These result from direct medical costs, time away from education and occupation, negative physical and psychological experience, disruption of the lives of patients and their families, loss of the ability for children and young adults to realise their full potential, and often from premature death.[8,16,17]

Diagnosis of acute rheumatic fever

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 4 weeks for a minimum of 10 years; and
- Under-diagnosis of ARF may lead to the individual suffering a further attack of ARF, probable cardiac damage, and possible premature death.

Jones criteria

The Jones criteria for the diagnosis of ARF were introduced in 1944.[18] 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 Jones criteria have been periodically modified and updated. The 1992 update is currently the most widely used and quoted version, and is intended only for the initial attack of ARF.[19] Each change of the Jones criteria is made to improve specificity at the expense of sensitivity, largely in response to the falling incidence of ARF in the USA. 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.

Important circumstances where ARF can be diagnosed without fulfilling the Jones criteria are noted. These include:

- Chorea as the only manifestation of ARF; and
- Indolent carditis (carditis of insidious onset and slow progression) as the only manifestation of ARF.[19]

In both these situations, cases may have insufficient supporting historical, clinical, or laboratory findings to fulfil the Jones criteria.

New Zealand Criteria

Initial episode of ARF—The main modification made to the Jones 1992 criteria for the New Zealand situation is the acceptance of echocardiographic evidence of carditis as a major manifestation.[20] In addition, there is a greater emphasis that monoarthritis may be a presenting feature if there is a history of non-steroidal anti-inflammatory (NSAID) use that is likely to have aborted classical ARF migratory polyarthritis (as alluded to in the Jones Criteria of 1992).

Categories of definite, probable, and possible ARF can be determined by the application of the New Zealand criteria to each case (Table 1). See the full guideline text for definitions of the major and minor manifestations of ARF [1] and Algorithm 1: Guide for diagnosis of ARF.[1]

Recurrent ARF—Most episodes of recurrent ARF fulfil the Jones criteria for ARF. The New Zealand guideline adopts the World Health Organization (2004) recommendations: where there is established RHD or a reliable history of ARF a recurrent attack can be diagnosed by the presence of several minor manifestations plus evidence of a preceding GAS infection[21] (Table 1).

Table 1. New Zealand guidelines for the diagnosis of acute rheumatic fever

Variables	Diagnostic requirements	Category
Initial episode of ARF	2 major **or** 1 major and 2 minor manifestations **plus** evidence of a preceding GAS infection (see text)	Definite ARF
Initial episode of ARF	1 major and 2 minor with the inclusion of evidence of a preceding GAS infection as a minor manifestation (Jones 1956)[22]	Probable ARF
Initial episode of ARF	Strong clinical suspicion of ARF, but insufficient signs and symptoms to fulfil diagnosis of definite or probable ARF	Possible ARF
Recurrent attack of ARF in a case with known past ARF or RHD	2 major **or** 1 major and 2 minor **or** several minor **plus** evidence of a preceding GAS infection (Jones 1992)[19]	
Major manifestations modified from Jones 1992[19]	Carditis (including evidence of subclinical rheumatic valve disease on echocardiogram)* Polyarthritis** or aseptic monoarthritis with history of NSAID use Chorea (can be stand-alone for ARF diagnosis) Erythema marginatum Subcutaneous nodules	
Minor manifestations	Fever Raised ESR or CRP Polyarthralgia** Prolonged P-R interval on ECG	

*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; **Other causes of arthritis/arthralgia should be carefully excluded, particularly in the case of mono-arthritis. If polyarthritis is present as a major manifestation, polyarthralgia or aseptic mono-arthritis cannot be considered an additional minor manifestation in the same person. All categories assume that other more likely diagnoses have been excluded; CRP=C-reactive protein; ECG=electrocardiogram; ESR=erythrocyte sedimentation rate; GAS=group A streptococcus; RHD=rheumatic heart disease.

Patients who do not fulfil these criteria, but in whom the clinician remains suspicious that the diagnosis may be ARF, should be maintained on oral penicillin and reviewed in 2 to 4 weeks with a repeat echocardiogram to detect the appearance of new lesions.[23,24]

If there is evidence of rheumatic valve disease clinically or on echocardiogram, the diagnosis is confirmed and long-term secondary prophylaxis can be commenced. If there is no evidence of carditis in a case of migratory polyarthritis, and no alternative diagnosis has been found then ARF is the diagnosis of exclusion. The putative new syndromes of paediatric auto-immune neuropsychiatric disorder associated with streptococcal infection (PANDAS)[2,5,26] and post-streptococcal reactive arthritis[27,28] should be diagnosed with extreme caution in New Zealand, particularly in Māori and Pacific populations.

Special consideration should be given to high-risk population groups such as Māori and Pacific people, and those residing in poor socioeconomic circumstances who may not be readily available for review. In these cases, it may be important to err on the side of diagnosis and treatment, and ensure ongoing review.

Echocardiography—Prior to the introduction of echocardiography, the diagnosis of rheumatic carditis relied on clinical evidence of valvulitis or pericarditis, supported in some situations by radiographic evidence of cardiomegaly. Today, all patients with suspected or definite ARF should undergo echocardiography to identify evidence of carditis. The role of echocardiography in the New Zealand setting is critical to the diagnosis of ARF. The use of echocardiography as a major criterion for ARF diagnosis[20,23,24] requires expert interpretation and adhering to echocardiographic diagnostic standards. In New Zealand, ARF carditis is classified mild, moderate, or severe (Table 2); these categories are used to guide the duration of secondary prophylaxis.

Table 2. Severity of ARF carditis

Mild carditis*
Mild mitral or aortic regurgitation clinically and/or on echo, 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 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. [24] 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.[24]
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. [24] • Reversal in lower descending aorta is required for severe aortic regurgitation. [24]

*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[24]; **When there is both mitral and aortic regurgitation, one must be moderate by echo criteria 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.

Evidence of a preceding group A streptococcal (GAS) infection—Streptococcal antibody titres are crucial in confirming a diagnosis of ARF. In study conditions in the New Zealand sore throat school clinic study, 74% (14/19) of cases of RF in children enrolled in the programme had a documented sore throat.[29] The most commonly used tests are the plasma antistreptolysin O (ASO) and the antideoxyribonuclease B (anti-DNase B) titres. The reference range for these antibody titres varies with age and background rate of streptococcal infections.[30]

In New Zealand, an ASO titre of greater than or equal to 480 units and/or an anti DNase B titre of greater than or equal to 680 units is accepted as significant. A positive throat culture or rapid antigen test for GAS alone demotes a case to probable or possible ARF, as up to 50% of those with a positive throat culture will be carriers only.[19]

Management of ARF—With very few exceptions, all patients with definite or possible ARF should be admitted to hospital as soon as possible after onset of symptoms. This ensures that all investigations (particularly echocardiography) are performed and, if necessary, the patient observed for a period prior to commencing treatment to confirm the diagnosis.

Hospitalisation also provides an ideal opportunity to provide information to patients and families, to notify the disease to public health (and preferably also to a local rheumatic fever register), to organise a dental check and ongoing dental care, and to initiate secondary prevention. All cases should also receive regular review, and outpatient follow-up should be organised 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. To ensure continuity of follow-up, a case can be made for maintaining less severe cases in the paediatric services until discharge at age 21 years.

Except in the case of heart failure management, none of the treatments offered to patients with ARF has been proven to alter the outcome of the acute episode or the amount of damage to heart valves.[31,32] Thus, there is no urgency to begin definitive treatment.

Salicylates or NSAID medications provide only symptomatic relief, and they should be withheld (with paracetamol used if required) until the diagnosis is confirmed to avoid masking the evolution of polyarthritis. Oral penicillin V (250 mg twice daily in children; 500 mg twice daily in adolescents and adults) should be commenced in all cases while the diagnosis is being established, and this should be continued until the first dose of intramuscular BPG for secondary prevention is delivered (also in hospital).

Many cases of chorea can be managed without medication. Where necessary, carbamazepine or valproic acid are recommended. Corticosteroids have not been proven to alter the likelihood of developing, or the severity of RHD.[32] Further details regarding the priorities in managing ARF and medication use are outlined in the Guideline.[1]

Secondary prevention—Secondary prevention of rheumatic fever is defined as the continuous administration of antibiotics to cases with a previous attack of ARF, or well-documented RHD. The purpose is to prevent colonisation or infection of the upper respiratory tract with GAS, and the development of recurrent rheumatic fever.[7,21] Secondary prophylaxis reduces the severity of RHD and is associated with regression of heart disease in approximately 50–70% of those with adequate adherence over a decade,[33–35] and reduces RHD mortality.[36]

Penicillin—The regular administration of antibiotics to prevent infection with GAS and recurrent ARF is recommended for all people with a history of ARF or RHD. A recent Cochrane meta-analysis 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).[37]

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.[38,39] In addition, oral penicillin V incurs a cost to the patient, while BPG is free when provided through an ARF prevention programme.

The internationally accepted standard dose of BPG for the secondary prevention of ARF in adults is 1,200,000 U.[40] In New Zealand, it is recommended that 1,200,000 U of BPG should be used for secondary prophylaxis for all persons weighing 20 kg or more, and 600,000 U for those weighing less than 20 kg.[41] Four-weekly (28-day) BPG delivery is recommended for all cases as prospective data has showed that few, if any, recurrences occurred among people who were fully adherent to a 28-day BPG regimen.[13]

Three-weekly (21-day) BPG is recommended only for those who have confirmed recurrent ARF despite full adherence to 28-day BPG. Oral penicillin should be reserved for patients who refuse intramuscular BPG. If a patient is offered oral penicillin, the consequences of missed doses must be emphasised, and adherence carefully monitored.

The Auckland Acute Rheumatic Fever Register covers 60% of New Zealand ARF registrations. The overall programme failure rate is very low at 1.4 per 100 patient years and the penicillin failure rate 0.07 per 100 patient years.[13]

Duration of secondary prophylaxis

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

- Age (ARF recurrence is less common after the age of 25, and uncommon after the age of 30);[13,42]
- Clinical pattern (presence or absence of carditis or RHD, and severity of carditis or RHD); and
- Environment (particularly the likelihood of ongoing exposure to GAS), and time elapsed since last episode of ARF (the most vulnerable period for ARF recurrence are the years closest to the last episode).[12]

Based on these factors, the recommended duration of secondary prophylaxis is outlined in Table 3. See also the full guideline text[1] and *Algorithm 3: Guide for the duration of secondary prophylaxis*.[1]

Table 3. Duration of secondary prophylaxis

Category	Duration of prophylaxis
All persons with ARF with no or mild carditis	Minimum of 10 years after most recent episode ARF or until age 21 years (whichever is longer)
All persons with ARF with moderate carditis	Minimum of 10 years after most recent episode ARF or until age 30 years (whichever is longer).
All persons with ARF with severe carditis	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.

Individuals working or living with children, or in a living situation where there is overcrowding or close proximity to others (such as boarding schools 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.

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

Before stopping prophylaxis, recipients should be evaluated for symptomatic deterioration and the stability and severity of valve lesions. This should include echocardiographic assessment. Where limited echocardiography is available, priority should be given to patients with a history of moderate or greater carditis, a history of one or more ARF recurrences, or clinical evidence of carditis such as a murmur. 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.

Improving adherence to secondary prophylaxis—Improved adherence to secondary prevention 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.

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 and community members to assist compliance to BPG. Three key methods for improved adherence to prophylaxis (which are covered in more detail in the full guideline text)[1] are: effective rheumatic fever registers; education of families and children affected; and attempting to reduce the pain of the BPG injection.

The efficacy of secondary prophylaxis achieved by the Auckland Register sets a benchmark for other registers in New Zealand and indeed, internationally.

Conclusion

Although ARF is rare in industrialised countries, it is a significant cause of disease among Māori and Pacific children in New Zealand. The prevalence of RHD is also high among these populations. There is often significant variability in the diagnosis and management of ARF cases and the persistence of recurrent ARF in New Zealand highlights that implementation of prevention strategies has been inconsistent.

The New Zealand Guideline for Rheumatic Fever: Diagnosis, Management and Secondary Prevention[1] summarised here increases the sensitivity for the diagnosis of ARF and provides opportunity for consistent management of ARF. The Guideline details effective and cost-effective secondary prevention strategies. Implementation of the Guideline should reduce ARF recurrences and reduce the burden of RHD in high risk populations in New Zealand.

Competing interests: No competing interests were involved in the development of the Guideline.

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Acknowledgements: The authors thank:

- Other members of the New Zealand Writing Group—Bruce Arroll, Elizabeth Farrell, Jonathan Jarman, Melissa Kerdemelidis, Henare Mason, Johan Morreau, Ross Nicholson, Briar Peat, Heather Spinetto, and Lesley Voss.
- National Heart Foundation of New Zealand.
- National Heart Foundation of New Zealand and the Australasian Faculty of Public Health Medicine for funding Polly Atatoa-Carr (who coordinated the writing of this Guideline).
- The Chairs of the New Zealand Guideline Writing Committee were also involved in the development of the Australian guideline,[2] with the understanding that this would be adapted for the New Zealand setting. We are grateful for the contribution of our Australian colleagues.

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