



A Summary of the New Zealand Guidelines for Rheumatic Fever

Overview

Acute rheumatic fever (ARF) and rheumatic heart disease (RHD) are a significant cause of disease among Māori and Pacific children in New Zealand (affecting 1 in 3 children), with significant morbidity and mortality among young adults.

ARF, an auto-immune response to group A streptococcus (GAS) infection of the upper respiratory tract, may result in damage to the mitral and/or aortic valves resulting in RHD. Recurrences are likely in the absence of preventative measures and may cause further cardiac valve damage.

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. However access to healthcare for Māori and Pacific children is inequitable.

Acute rheumatic fever leading to rheumatic heart disease is considered to be a preventable chronic disease. A large New Zealand randomised controlled trial combined with similar studies shows a community intervention of sore throat management in schools to be effective in reducing the occurrence of acute rheumatic fever. School sore throat programmes in high risk areas are recommended.

Prevention of recurrences and therefore rheumatic heart disease prevention, with intramuscular penicillin is both effective and highly cost-effective.

Three guidelines evaluating currently available evidence have been developed outlining the approach to acute rheumatic fever/rheumatic heart disease, its diagnosis, management and control.

Background

Māori and Pacific people in New Zealand have one of the highest rates of both ARF and RHD internationally. There are ethnic differences in ARF incidences where population groups experiencing low socioeconomic status and living in overcrowded situations are particularly at risk.

ARF is now rare in industrialised countries, having 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. 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.

ARF however, has a high incidence in developing countries with RHD the most common form of paediatric heart disease in the world, and the most common cause of cardiac mortality in many countries in children and adults under 40 years old. Globally, there are at least 15.6 million people with RHD, 470,000 new cases of ARF each year and over 230,000 deaths due to RHD annually.

New Zealand has had high rates of ARF and RHD for many decades. The decline in incidence of ARF seen in other developed countries has not been evident in New Zealand. Socioeconomic deprivation is linked with higher rates of both ARF and RHD.

In 2003, the rate for ARF was 3.8 per 100,000 exceeding that in other western countries. Most of the 141 new cases reported in that year occurred in the five to 14 year age group. Auckland accounts for 60% of the cases on New Zealand registers.

New Zealand has an efficient secondary prophylaxis programme and the need for valve surgery for recurrent episodes is decreasing. However, there has been no attempt to prevent first attacks and eradicate the disease.

There is no evidence to date that Māori and Pacific people have increased genetic susceptibility to rheumatic fever. It is highly likely that a combination of overcrowded conditions, socioeconomic deprivation, high incidence of pharyngitis with GAS and inadequate access to healthcare are important factors leading to high rates of rheumatic fever and rheumatic heart disease. Most of these factors are modifiable.

Guideline 1:

Diagnosis, Management and Secondary Prevention of Rheumatic Fever

This guideline presents the evidence for best practice in ARF diagnosis and the standard of care that should be available to all people in New Zealand.

Diagnosis of acute rheumatic fever (Algorithm 1)

- The diagnosis of ARF requires health professionals to be aware of the diagnostic criteria. Hospital referral where expertise is available for accurate diagnosis particularly echocardiography, is usual.
- It is important that an accurate diagnosis of ARF is made as:
 - Misdiagnosis may result in the individual receiving benzathine penicillin G (BPG) injections unnecessarily every four weeks for a minimum of 10 years.
 - Under-diagnosis of ARF may lead to the individual suffering a further attack of ARF, cardiac damage and premature death.
- There is no single laboratory test specifically diagnostic for ARF; diagnosis is based on full clinical assessment. The pre-test probability for diagnosis of ARF varies according to location and ethnicity. e.g. 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.
- Jones' (1992) diagnostic criteria have been modified for the New Zealand guidelines and should be used to determine definite, probable and possible ARF. 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 (Table 2, 3 and 4).
- 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 two to four weeks with a repeat echocardiogram to detect any new lesions.
- All patients with suspected or definite ARF should undergo echocardiography to identify evidence of carditis (**Algorithm 2**). Patients should be hospitalised until the diagnosis is secure.

Management of acute rheumatic fever

- 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, except for heart failure management.
- Priorities for managing ARF are: admission to hospital, confirmation of diagnosis, treatment (antibiotics and management of arthritis/arthralgia, fever, carditis/heart failure and chorea), clinical follow-up and commencement of long-term preventive measures.
- Most cases of ARF without severe carditis can be discharged from hospital after approximately two weeks although in some cases it may be advisable to prolong hospital stay further.

Secondary prevention (Algorithm 3)

- This is 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 recurrent rheumatic fever.
- Secondary prophylaxis reduces the severity of RHD and is associated with improvement in heart disease in approximately 50-70%. Mortality is reduced.
- 1,200,000 U of benzathine penicillin G (BPG) four weekly for all persons weighing more than 20kg and 600,000 U for those weighing less than 20kg is the treatment of choice. This is superior to oral penicillin and compliance is assured. Where benzathine penicillin is unavailable, oral penicillin (or erythromycin for the allergic patient) can be given.
- To assist with the adherence to secondary prophylaxis, it is important to support and utilise the expertise, experience, community knowledge, culture and language skills of Māori and Pacific health workers.

Guideline 2:

Group A Streptococcal Sore Throat Management (Algorithm 4)

This guideline has been developed to define best practice for the management of group A streptococcal sore throats (pharyngitis) in three to 45 year old New Zealanders.

The guideline specifies:

- When to perform a throat swab
- When to prescribe an antibiotic
- Which antibiotic to prescribe and the length of treatment.

Sore throats are a common medical condition which are usually viral and benign. In the New Zealand population, group A streptococcal sore throats are considered to be the only clinically significant bacterial throat infection. The underlying premise is that treatment of streptococcal pharyngitis will reduce the incidence of rheumatic fever. Within the New Zealand population, not all groups are at equal risk of developing ARF as a consequence of streptococcal throat infection.

Optimal management of sore throat in those who are at greatest risk of developing rheumatic fever will minimise investigations and antibiotic use in those who are at the lowest risk. The clinical end point is rheumatic fever prevention.

Key messages include:

- In the New Zealand setting, Māori and Pacific three to 45 year olds from lower socioeconomic areas have the highest rate of acute rheumatic fever. These high rates necessitate a targeted approach to sore throat management.
- Culture of throat swabs is recommended to confirm diagnosis of group A streptococcal pharyngitis.
- Throat swabs and antibiotics should be used judiciously especially in low-risk patients using clinical parameters and rheumatic fever risk factors.
- To facilitate tracking of GAS pharyngitis, it is recommended that clusters of GAS pharyngitis, as detected in microbiology laboratories, should become a notifiable disease to local medical officers of health.
- Treatment of GAS pharyngitis infection with appropriate antibiotics (ten days of oral penicillin V) reduces the occurrence of acute rheumatic fever.
- Three or more episodes of GAS pharyngitis within a household, within a three month period, diagnosed using throat swabs, require management of the household. The entire household should have throat swabs and be treated with antibiotics if GAS positive, regardless of whether accompanied by a sore throat.
- Three or more episodes of pharyngitis for an individual inside a three month period, require treatment using antibiotics according to the 'recurrent antibiotics' table in the guideline.
- Development of GAS positive sore throat while on intramuscular benzathine penicillin prophylaxis requires differentiation of treatment depending on whether it has occurred in the first two weeks after the injection or in the second two weeks.
- Children with GAS pharyngitis should be kept home from school or day-care for 24 hours until treatment is established.
- Treatment of GAS pharyngitis can be delayed until culture results are available for up to nine days, as rheumatic fever is unlikely to occur in this time.
- Rapid streptococcal tests are not funded in New Zealand and need further investigation regarding their sensitivity and specificity in this country.
- No vaccine for GAS has yet been developed.
- There is no agreement about the role of tonsillectomy in recurrent GAS pharyngitis.

Guideline 3:

Rheumatic Fever Primary Prevention

Primary prevention is a strategy that seeks to prevent disease occurring in the first instance rather than treating it once it has developed. In the case of ARF this means treating group A streptococcus bacterial throat infections before they can initiate ARF.

Key messages include:

- Any first attack of acute rheumatic fever is potentially preventable.
- Treating group A streptococcal throat infections reduces the subsequent rate of development of acute rheumatic fever.
- School, mixed community and school-based GAS sore throat detection and treatment programmes are all effective in reducing rheumatic fever. A meta-analysis involving international community-based studies and New Zealand research on rheumatic fever primary prevention provides strong evidence to support school sore throat clinic initiatives to prevent acute rheumatic fever in high risk areas.
- Crowding in the household is associated with an increased risk of developing rheumatic fever.
- Some studies show a link between poverty and rheumatic fever; others do not.
- There is some evidence linking poor quality housing and rheumatic fever, but definitions vary between studies and it is impossible to make recommendations for minimum standards of housing.
- Māori and Pacific healthcare providers, school-based sore throat programmes and primary healthcare reforms all have a role in improving access to healthcare for patients most at risk of rheumatic fever.
- There is no convincing evidence that rheumatic fever is caused by skin infections.
- There is no convincing evidence of a genetic cause of rheumatic fever and no reliable genetic markers of susceptibility to the disease.
- The role of seasonal antibiotic prophylaxis to reduce the rate of GAS pharyngitis has not been proven.

The New Zealand guidelines on rheumatic fever include:

- **Guideline 1:** Diagnosis, management and secondary prevention
- **Guideline 2:** Sore throat management*
- **Guideline 3:** Proposed rheumatic fever primary prevention programme*
- **Algorithm 1:** Guide for diagnosing acute rheumatic fever
- **Algorithm 2:** Guide for the use of echocardiography in acute rheumatic fever
- **Algorithm 3:** Guide for the duration of secondary prophylaxis in acute rheumatic fever
- **Algorithm 4:** Guide for sore throat management

* These are only available online.

These can be ordered online, or downloaded from: www.heartfoundation.org.nz

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