



# Rheumatic fever: the neglected disease

## Key concepts

- New Zealand continues to have high rates of rheumatic fever, particularly among Pacific peoples
- 80% of cases occur in young people aged less than 15 years
- The majority of areas with a high incidence of rheumatic fever are in the North Island
- Acute rheumatic fever can be prevented by effective treatment of Group A streptococcal throat infection
- Management of people presenting with sore throat should be guided by age, ethnicity and location
- Guidelines have been developed for the diagnosis and management of sore throat and acute rheumatic fever and also for primary and secondary prevention of rheumatic fever

## The problem

Pacific peoples have the highest rate of rheumatic fever in New Zealand and one of the highest rates in the world. In 2009, there were 53 notified cases of rheumatic fever among Pacific peoples in New Zealand, a rate of 23 per 100 000. This is over six times the overall rate for all New Zealanders of 3.5 per 100 000 (a total of 140 cases).<sup>1</sup>

New Zealand stands out from most other developed countries in continuing to have high rates of acute rheumatic fever (ARF) and rheumatic heart disease (RHD). It is estimated that 97% of cases of RHD worldwide occur in developing countries and in the indigenous populations of countries such as New Zealand and Australia.<sup>2</sup>

Since 1984, ARF has been a notifiable disease in New Zealand. However, it continues to be under-notified despite increasing rates each year.<sup>3,4</sup>

There is significant geographical variation in the rates of ARF in New Zealand, with the highest rates in the North Island, e.g. Tairāwhiti, Hawke's Bay and Northland.<sup>4,5</sup> However, clusters of cases occur in a number of communities across New Zealand.

 **Best Practice Tip:** Check the incidence of rheumatic fever in your DHB area. A map of New Zealand showing rates per DHB is available in the Heart Foundation Rheumatic Fever Guidelines (see sidebar).

The majority of cases of ARF (approximately 80%) occur in young people aged less than 15 years.<sup>1</sup> The high rates of ARF in Pacific peoples have been widely attributed to socioeconomic factors such as overcrowding, poverty and poor nutrition, but also to delayed diagnosis and treatment of streptococcal throat infection.

### Group A streptococcal throat infection

Appropriate diagnosis and treatment of streptococcal sore throat in high risk populations is required to reduce the incidence of ARF. A guideline for the management of sore throats in New Zealand (see sidebar) has been developed to assist with targeted treatment of streptococcal throat infection and includes algorithms for individual and household management.

Community pharmacists, particularly those in areas of high ARF incidence, can assist by encouraging patients with sore throat to see their GP.

### Approach to treatment of sore throat in high risk groups

A key message from the Auckland Regional Public Health Service is to: “Think differently about sore throats in different population groups”.<sup>6</sup>

All children presenting with sore throat who are of Pacific or Māori ethnicity, aged three years and over and who live

in areas with a high incidence of rheumatic fever, should have a throat swab taken.

If the child has any of the following clinical features, empirical antibiotics should be prescribed:<sup>6</sup>

- Tonsillar swelling or exudate
- Anterior cervical lymphadenopathy
- No cough or coryza (which may suggest viral cause)
- Temperature  $\geq 38^{\circ}\text{C}$

If none of the clinical features are present, wait for the results of the throat swab. If the swab is positive for group A streptococcus, a ten day course of antibiotics, e.g. penicillin V, amoxicillin or erythromycin, should be prescribed.

### Acute rheumatic fever and rheumatic heart disease

ARF arises from an autoimmune response to group A streptococcal throat infection. On average there is a latent period of three weeks between the initial infection and the development of symptoms of ARF. The majority of people with ARF are very unwell, in considerable pain and require hospitalisation for confirmation of diagnosis and treatment. ARF causes a widespread inflammatory response that affects the heart, joints, skin and brain.

## The Heart Foundation Rheumatic Fever Guidelines

The Heart Foundation of New Zealand has developed a three part guideline for rheumatic fever;

1. Diagnosis, management and secondary prevention
2. Group A streptococcal sore throat management
3. Proposed rheumatic fever primary prevention programme

These guidelines provide key information including:

- A geographical map of rheumatic fever incidence
- Guidelines for the management of sore throat
- Clinical features and diagnosis criteria for rheumatic fever

The full guidelines are available from the Heart Foundation website: [www.heartfoundation.org.nz](http://www.heartfoundation.org.nz) Keyword search: Rheumatic fever



The heart (specifically the mitral and/or aortic valves) is the only organ that suffers long term damage, particularly after recurrent attacks of ARF. In some people ARF may be silent and symptomless, but still affects the heart i.e. causing subclinical carditis.<sup>4,7</sup>

ARF is diagnosed clinically because there is no single diagnostic test available. Diagnosis is based on the Jones criteria although these may not be sensitive enough to detect ARF in populations with a high incidence such as Pacific peoples.<sup>8</sup>

A modified version of the Jones criteria and a full description of the clinical features of the major and minor manifestations of ARF are detailed in the Heart Foundation guidelines for rheumatic fever (see sidebar). Criteria for diagnosing ARF include the presence of two major, or one major and two minor, manifestations, plus a preceding group A streptococcal infection. Major manifestations include carditis, polyarthrititis, chorea, erythema marginatum and subcutaneous nodules. In New Zealand, evidence of subclinical carditis on echocardiogram is also accepted as a major manifestation.<sup>8</sup> Minor manifestations include fever, raised CRP, polyarthralgia and prolonged P-R interval on ECG. If these signs are not present but there is strong clinical suspicion, ARF remains a possible diagnosis.<sup>8</sup>

Diagnostic certainty may vary according to location and ethnicity. It is recommended that a lower threshold for diagnosis be applied to people who:<sup>8</sup>

- Are in high risk groups (such as Māori and Pacific peoples)
- Live in lower socioeconomic areas
- Have delayed presentation
- Have atypical clinical features at presentation

Refer all patients with suspected ARF to hospital. Clinical follow-up of patients and their close contacts, and the ongoing use of prophylactic antibiotics after an attack of ARF are important in preventing recurrence of ARF and RHD.

It has been estimated that over 60% of patients with ARF will develop RHD,<sup>9</sup> which remains a significant cause of premature death in New Zealand (responsible for up to 200 deaths each year).<sup>10</sup> Adult patients may present with RHD that is a legacy of ARF from decades previously.

## The difficulties

Not all streptococcal throat infections cause symptoms and many children with sore throat do not present to primary care. Therefore there should be a low threshold for swabbing and treating sore throats in people who live in areas of high incidence of ARF.

Pacific people are often stoical, putting up with a sore throat or a sore joint and not presenting for medical care. Pacific families may prefer to use traditional health remedies rather than visit a doctor. Children may present later, so in high incidence areas antibiotics should be prescribed empirically rather than waiting for swab results (if the child has a sore throat and clinical features as detailed previously). A sore, swollen joint in a child should never be ignored and a possible diagnosis of ARF should always be considered.

Populations that are transient are likely to be more at risk. There may be a lack of continuity within primary care which can result in delayed diagnosis or treatment or stopping antibiotics needed for secondary prevention. Irregular school attendance may jeopardise school-based detection programmes. Multiple caregivers may result in a child attending multiple GPs. Secondary prevention programmes are also only effective with consistent long-term follow up.

## The solutions

Targeted interventions are important. New Zealand-wide approaches include:

- Ongoing awareness and education about the Heart Foundation rheumatic fever treatment guidelines for all medical care staff both at a primary and secondary care level. The goal is for a reduced incidence of ARF through effective treatment of sore throat.

- Secondary prevention programmes to prevent recurrence in people who have had confirmed ARF or RHD. These programmes rely on effective follow up to ensure regular administration of prophylactic antibiotics over a minimum of ten years.

School-wide regular throat swabbing programmes have successfully reduced the incidence of ARF in some regions. The use of portable echocardiograms to detect previously undiagnosed RHD in school children has also been initiated in some areas.

Solutions aimed at improving housing, reducing overcrowding and improving the socioeconomic situation of Pacific peoples will require a longer time frame and a co-ordinated approach with other sectors, e.g. education, welfare and housing, at both local and national levels.

Some recent regional approaches include:

**The Opotiki Rheumatic Fever prevention project** led by Te Ao Hou PHO was initiated in October 2009. The message was: “sore throats matter”, and the project involved community health workers visiting primary schools, three times a week, to take throat swabs (with parental consent) from children who reported sore throat.

Any child who tested positive for group A Streptococcus received a ten day course of antibiotics. This campaign has raised public awareness and has increased the number of parents requesting throat swabs for children with sore throat.

More information is available from:

**[www.toiteorapublichealth.govt.nz/Rheumatic\\_Fever\\_GP](http://www.toiteorapublichealth.govt.nz/Rheumatic_Fever_GP)**

**The “Say Aah” campaign** in Flaxmere, Hawkes Bay, is fronted by All Black Israel Dagg. This campaign aims to obtain parental permission to take throat swabs from all school children in Flaxmere, an area with a rheumatic fever rate of 32 per 10 000.

In 2002, a successful community based primary prevention programme for rheumatic fever was initiated in **Whangaroa, Northland**. For more information on this programme, see “How a community controlled the Streptococcus”, BPJ 13 (May, 2008).

 For further information about rheumatic fever see “Why we still need to think of rheumatic fever”, BPJ 13 (May, 2008).

## References

1. ESR. Public Health Surveillance. Notifiable and Other Diseases in New Zealand. Annual Surveillance Report. ESR, 2009. Available from [www.surv.esr.cri.nz](http://www.surv.esr.cri.nz) (Accessed Oct, 2010).
2. White H, Walsh W, Brown A et al. Rheumatic Heart Disease in Indigenous Populations. *Heart Lung Circ* 2010;19:273-81.
3. Loring B. Rheumatic Fever in the Bay of Plenty and Lakes District Health Boards. A review of the evidence and recommendations for action. Summary Report. 2008. Available from: [www.toiteorapublichealth.govt.nz/vdb/document/150](http://www.toiteorapublichealth.govt.nz/vdb/document/150) (Accessed Oct, 2010).
4. Wilson N. Rheumatic heart disease in indigenous populations – New Zealand experience. *Heart Lung Circ* 2010;19:282-8.
5. Heart Foundation of New Zealand (HFNZ). New Zealand guidelines for rheumatic fever. 2. Group A Streptococcal Sore throat Management. HFNZ, 2008. Available from: [www.heartfoundation.org.nz](http://www.heartfoundation.org.nz) (Accessed Oct, 2010).
6. Auckland Regional Public Health Service (ARPHS). Sore throats and acute rheumatic fever: advice for GPs and Practice Nurses. Available from: [www.arphs.govt.nz/Guidelines/SoreThroats\\_ARF.asp](http://www.arphs.govt.nz/Guidelines/SoreThroats_ARF.asp) (Accessed Oct, 2010).
7. Tubridy-Clark M, Carapetis JR. Subclinical carditis in rheumatic fever: A systematic review. *Int J Cardiol* 2007;119(1):54-8.
8. Heart Foundation of New Zealand (HFNZ). New Zealand guidelines for rheumatic fever. 1. Diagnosis, management and secondary prevention. HFNZ, 2006. Available from [www.heartfoundation.org.nz](http://www.heartfoundation.org.nz) (Accessed Oct, 2010).
9. Carapetis JR, Steer AC, Mulholland EK, Weber M. The global burden of group A streptococcal diseases. *Lancet Infect Dis* 2005;5(11):665-732.
10. Ministry of Health. New Zealand mortality statistics: 2006-2009. Ministry of Health. Wellington, 2010. Available from: [www.moh.govt.nz](http://www.moh.govt.nz) (Accessed Oct, 2010).