Rheumatic Fever in Māori:

WHAT CAN WE DO BETTER?

Whaia te iti kahurangi – Ki te tuohu koe, me he maunga teitei, ko Aoraki anake

Pursue excellence – should you stumble, let it be to a lofty mountain

Key concepts

- In New Zealand, acute rheumatic fever is now almost exclusively a disease affecting Māori and Pacific peoples
- People who have had acute rheumatic fever require secondary prophylaxis for at least ten years to reduce the risk of developing chronic rheumatic heart disease
- Cases of acute rheumatic fever, rheumatic heart disease and the number of deaths due to chronic rheumatic heart disease are increasing in New Zealand
- Primary care clinicians need to know the risk of rheumatic fever in their community and to encourage at-risk people to seek a consultation when they have a sore throat
- In high-risk areas, all Māori and Pacific children who present with a sore throat should have a throat swab taken and antibiotics prescribed empirically if any red flags are present

Things are getting worse, not better...

The rate of Acute Rheumatic Fever (ARF) in New Zealand is increasing. Between 2005 and 2010, the rate of ARF doubled from 1.9 reported cases per 100 000 population to 3.8 per 100 000 (Figure 1).[1] The majority of cases are occurring in low socioeconomic communities in the northern and central North Island and in pockets around the Wellington region. ARF is 23 times more likely in Māori and nearly 50 times more likely in Pacific peoples than in other ethnic groups.[2] From 1996 to 2005, while ARF rates significantly decreased amongst New Zealand Europeans, rates amongst Māori and Pacific children increased significantly. Although comprising just 4.7% of the New Zealand population, this group represented almost 60% of all cases reported.[3] It is widely believed that this over representation is due to a combination of overcrowded living conditions, poverty and decreased access to treatment options.

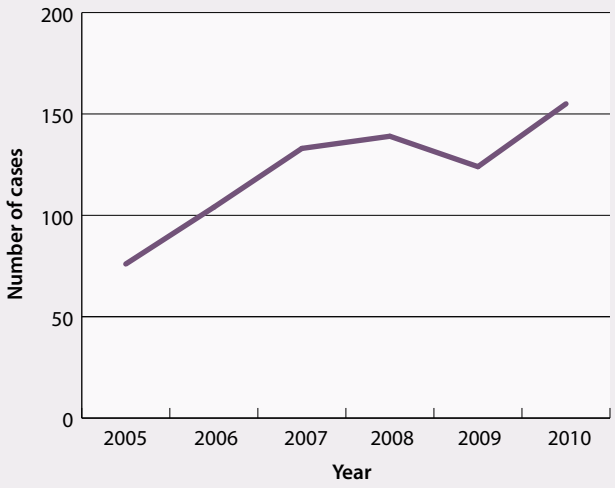


Figure 1: The number of reported cases of ARF in New Zealand (2005-2010)[1]

Other than New Zealand, and indigenous populations in Australia and the Pacific Islands, ARF is a disease of the developing world. What primary health care services in New Zealand can do is to:

1. Understand there is a problem
2. Be aware of the New Zealand guidelines and algorithms for the management of ARF
3. Assist in raising community awareness, and support early detection programmes in high-risk areas
4. Continue to lobby for improvements and equality in socioeconomic factors

What is acute rheumatic fever?

Acute rheumatic fever is an autoimmune response to a group A streptococcus (GAS) infection, usually in the upper respiratory tract. The resulting transitory, generalised, inflammatory response may affect the heart, joints, central nervous system and skin. The chief concern is the inflammation of the heart (carditis), as this may cause cardiac valve stenosis. ARF has no known genetic basis and occurs mainly in children aged five to 14 years, peaking around age eight. ARF is rare in children aged under three years, as the immune system has not developed sufficiently to allow an autoimmune response to occur.[4]

In New Zealand, ARF has been a notifiable disease since 1986. Despite this, it is likely that there are significantly more cases of ARF in New Zealand than are currently being reported to health services. A 2008 review of ARF cases (1999–2007) in the Bay of Plenty and Lakes Districts DHBs found 147 cases, where only 75 had been previously reported.[5] Factors identified as contributing to under reporting by health professionals included; difficulty of ARF diagnosis, multiple health professionals being involved at multiple institutions and health professionals being unfamiliar with reporting requirements due to a lack of experience with ARF or as a result of being trained overseas. It is also likely that some patients will not present unless the symptoms are severe.

Sore throats mean different things in different places

Rates of ARF vary throughout New Zealand. It is important that primary care clinicians know the approximate rate of ARF in their communities (Figure 2).

Practices in northern and central North Island and Porirua should be on high-alert for ARF and the group A streptococcus (GAS) throat infections that cause it. Māori and Pacific peoples have higher rates of GAS infections than New Zealand Europeans.[6] However, awareness at a national level is required given the high degree of mobility amongst the young adult population.

Overcrowding, economic deprivation and decreased treatment opportunities also increase the risk of a person developing ARF.[7] When a patient presents with a sore throat, assessing these factors during consultation indicates the risk that person may have of developing ARF.

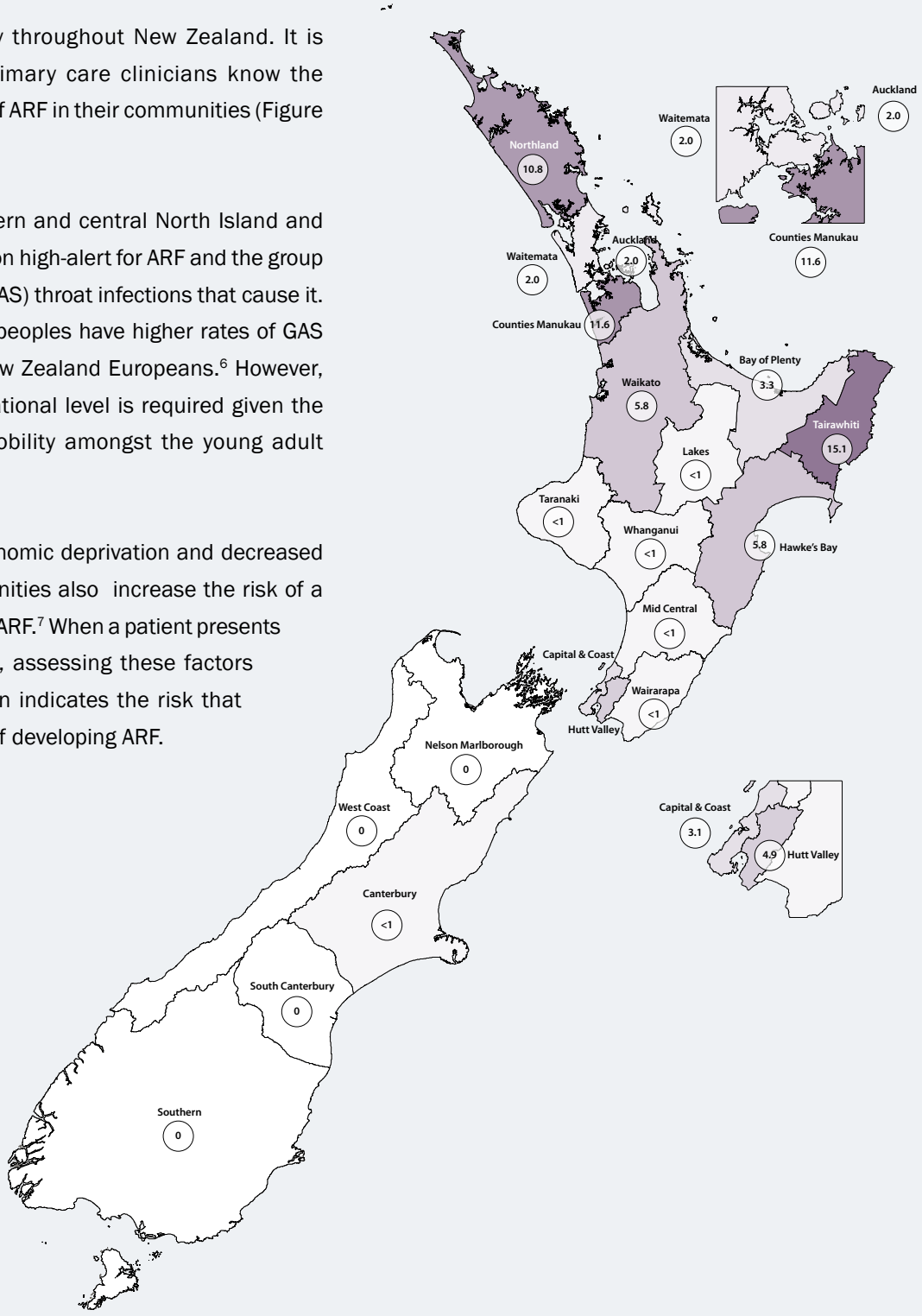


Figure 2: Annual rate of rheumatic fever first admissions by DHB in New Zealand (2010) per 100 000 population[1]

Rheumatic heart disease is a long-term consequence of acute rheumatic fever

Over 60% of people with ARF will have long-term heart damage, termed chronic rheumatic heart disease (RHD), if left untreated.[8] RHD can cause complications later in life, particularly in a person's 30s and 40s. Approximately 145 people in New Zealand die each year from RHD.[9–11]

People who have RHD have a higher risk of developing infective endocarditis and need to maintain good oral hygiene and attend regular dental check-ups. Dentists and other health professionals need to know if a patient has RHD, as it increases the risk of infection. Patients who have been identified as being at risk are given wallet cards (see Appendix H of Heart Foundation guidelines[4]) to present to health professionals. These cards contain recommended doses of additional antibiotics that may be required for various procedures, e.g. dental procedures involving bleeding of gingival tissues.

Think differently about sore throats

Most sore throats are viral in origin, but approximately 10% of sore throats in adults and 15–30% of sore throats in children are due to GAS infections.[5] GAS is droplet spread with a cross-infection rate of 19–50% within a household.[12]

Treatment of GAS sore throats with penicillin V (phenoxymethylpenicillin) or amoxicillin is considered the first-line treatment for preventing ARF.[13] Note that this is an update to the National Heart Foundation sore throat guidelines, which recommend penicillin V as first-line treatment and amoxicillin as second-line.[14] Amoxicillin is more palatable and may be taken with food, which may encourage increased compliance with treatment, compared to pencillin V.

Antibiotics, as outlined in Table 1, should be prescribed to patients for the first or second instance of GAS sore

Table 1: Recommended treatment for GAS positive sore throats (Algorithm 4 - Guide for sore throat management, National Heart Foundation, 2006, Lennon D et al, 2011)[13, 14]

Antibiotic	Route	Regimen	Duration
First choice			
Penicillin V	Oral (on empty stomach)	Children: 20 mg/kg per day in two to three divided doses. Maximum 500 mg, three times daily Adults: 500 mg, twice daily	10 days
Amoxicillin	Oral	Weight <30 kg: 750 mg, once daily Weight >30 kg: 1500 mg, once daily	10 days
Alternatives			
Erthromycin ethyl succinate (EES) if documented allergy to penicillin	Oral	Children: 40 mg/kg/day in two to four divided doses, to a maximum of 1 g/day Adults: 400 mg, twice daily	10 days
Benzathine penicillin G (BPG) if compliance with 10 day regimen an issue	IM	Weight <20 kg: 600 000 U once Weight >20 kg: 1 200 000 U once	Single dose

throat in a three month period. Other recommendations are provided for recurrent cases of GAS infection (see sore throat algorithm, Heart Foundation guidelines).[14]

Some General Practitioners may be reluctant to prescribe antibiotics empirically due to messages about the "Wise Use of Antibiotics", which have reduced antibiotic use and patient's expectations for receiving them for colds and sore throats during winter months. It is important that practices understand that wise use means carefully selecting patients who might benefit from antibiotics, rather than avoiding prescribing completely. In most cases, ARF can be prevented if antibiotics are taken within nine days of onset of symptoms.

Throat swabs

Culture of a throat swab is the best way to confirm a GAS infection. Running a standard, sterile microbiological swab over the tonsils, back of the throat and any area with exudate, while avoiding the tongue and teeth, ensures a good sample is collected. When possible the sample should be sent for culture within two hours, however a delay of up to twenty four hours is acceptable.

Red flags for a potential GAS throat infection are:

- Temperature > 38 degrees celsius
- No cough or coryza (which may suggest a viral cause)
- Swollen anterior cervical lymph nodes
- Tonsillar swelling or exudate

In high-risk areas, all Māori and Pacific people aged between three and 45 years, who present with a sore throat, should have a throat swab taken. Prescribe antibiotics empirically if a sore throat and any red flags are present. If patients later return a positive GAS culture, then prescribe antibiotics if they are not already taking them.

Swabbing and treatment of entire households should occur if more than three cases of GAS sore throat occur in the same home within a three month period. The New Zealand algorithm for sore throat management also provides guidance for managing households and whānau in high-risk areas.

A New Zealand algorithm for the management of sore throats is available from the Heart Foundation website (listed at end of article).

There is more that can be done in high-risk areas

- Raising community awareness is an important tool in combating GAS infections
- When people from high-risk groups present with sore throats, validate their attendance as this helps to reinforce good behaviour patterns
- Repeat the message – "sore throats matter" – to parents, Kaumātua, teachers, community pharmacists, Kohanga Reo, and other influential community members
- Put up posters in practices – the National Heart Foundation website has resources available in Māori, Samoan and Tongan languages

New Zealand guidelines recommend raising community awareness and implementing early detection and treatment of GAS sore throats when annualised ARF rates exceed 20 per 100 000. If possible, school-based sore throat programmes should be implemented by Public Health Units when rates exceed 50 per 100 000.[12]

Best Practice tip: To find out the incidence of ARF in a specific area, contact the Medical Officer of Health at your local Public Health Unit.

Diagnosing acute rheumatic fever

There is currently no diagnostic test for ARF so clinical judgement is important. New Zealand guidelines for the diagnosis of ARF are based on the Jones criteria and are available from the Heart Foundation website (listed at end of article).

The presence of two major symptoms, or one major and two minor symptoms, in both cases with a prior GAS infection, are required to diagnose ARF.

Major symptoms include:

- Arthritis, the most common symptom, occurs in 75% of first attacks, usually in the larger joints such as the knees and ankles[6]
- Carditis almost always affects the mitral and aortic valves and on presentation, a murmur may be heard.[6] In New Zealand, subclinical carditis confirmed by echocardiography is also considered a major symptom.
- Chorea (uncoordinated movements), often in adolescent females, especially affecting the hands, feet, tongue and face which disappear during sleep and may only affect one side of the body. Chorea may occur following a prolonged latency after streptococcus infection and generally resolves within six weeks.
- Erythema marginatum – rare (pink rings on the trunk and limbs)
- Subcutaneous nodules – rare, but highly specific to ARF

Minor symptoms include:

- Fever which accompanies most cases of ARF, except when chorea is present
- Joint pain
- Elevated CRP >30 mg/L or ESR >50 mm/h
- A prolonged P-R interval on ECG

Antistreptolysin O titre is used to confirm the diagnosis, as less than 10% of people with ARF have GAS positive throat swabs. Assuming no re-infection, antibody levels return to normal in six to 12 months.

All patients with suspected ARF should be referred to hospital for confirmation of the diagnosis. Hospitalisation also provides an opportunity to educate whānau about the condition and the importance of preventing recurrent attacks.

Recurrent acute rheumatic fever

Recurrent rheumatic fever occurs when people who have previously had ARF are re-infected with GAS. Recurrent attacks greatly increase the chance of a person developing RHD. Secondary prophylaxis reduces the incidence of recurrent ARF. However, it is of major concern that the mortality rate in New Zealand due to RHD is increasing.[10,15]

Māori and Pacific people have the highest recurrence rates of rheumatic fever. In Auckland, between the 1980s and 1999, recurrence rates dropped from 22% to 5.5%,

Rheumatic fever registers

Rheumatic fever registers are an important tool for managing patient compliance to secondary prophylaxis, particularly when people move. Registers are also necessary for analysing epidemiology and channelling financial resources into regions with the greatest need. The Auckland rheumatic fever register was established in 1981 to streamline antibiotic delivery to patients. The subsequent clinic-based prophylaxis programme saw recurrent rheumatic fever attacks fall from 20% of total hospital admissions for rheumatic fever to 6%.[18] Further, widespread reductions would be likely if a national rheumatic fever register was established.

Eradication of rheumatic fever

On 26 April 2011, a group of 40 health professionals from areas of high prevalence for rheumatic fever met to discuss a pathway to achieve eradication of the disease. The National Steering Group and the Ministry of Health are now working closely to achieve this goal.

Presentations from Professor Diana Lennon, Dr Pat Tuohy and representatives from high risk areas outlined the cross-sectoral approach to managing rheumatic fever, including what has worked and what has not. Workshop groups then discussed health promotion, the role of primary care, monitoring progress and integration with other childhood illness.

An over-arching theme was the need to bring all initiatives and knowledge together to ensure cohesion, quality processes and success.

Key recommendations from the workshops included:

- Community based and whānau centred approaches with appropriate consultation are required for successful health promotion and community awareness
- Nurse management of sore throat is an effective means of assuring accessible and affordable care in high risk areas. Flaxmere, Kaikohe and Porirua provide current best practice examples. A "walk in" basis is important with no financial barriers, with training supported by PHOs as part of existing continuing medical education (CME).
- Ensure parents know that prescriptions for children aged under six years are free of charge (or for those with a prescription subsidy card, Community Services card or High User Health card).
- Increase awareness of the importance of completing the ten day antibiotic course. Amoxicillin once daily is a good option for increasing compliance (compared to penicillin V which needs to be given two to three times daily on an empty stomach).
- For standardised care, IT support should be available in all practices with prompts for sore throat management. Local adaptation and simplification of guidelines in high risk areas such as Porirua can work well.
- Sore throat management should be audited as part of continuous quality improvement in primary care in high risk areas
- Nurses and other health workers could be used to broaden access to throat swabbing in high prevalence areas (in schools and other community settings), with appropriate linkage back into primary care for management

Following the seminar, the current management algorithms from the Heart Foundation guidelines are being reviewed and an agreed evidence-based approach to school and community clinics developed. The recent budget allocation of $12 million to assist eradication highlights the importance of rheumatic fever as a now recognised marker of child health. Minister Tariana Turia's championing and support for this successful bid and for raising the profile of rheumatic fever should be acknowledged. The current convergence of interest around this challenging issue allows guarded optimism that eradication can indeed be achieved. A follow-up seminar is planned for the end of the year.

Contributed by **Professor Norman Sharpe**, Medical Director of the National Heart Foundation of New Zealand and acting Chair of the National Steering Group.

but in 1999, all reported recurrent cases of rheumatic fever occurred in Māori or Pacific peoples.[16] In 2008, 10% of all cases of ARF in the Bay of Plenty and Lakes District DHBs were recurrences.

After contracting ARF, prevention of recurrent rheumatic fever requires intra-muscular injections of benzathine penicillin G (BPG) every four weeks for a minimum of ten years. This has been shown to be safe and effective when delivered by community nursing staff in schools, workplaces or homes.[17] Secondary prophylaxis also reduces the impact of RHD, and has been shown to be associated with a reduction in heart disease of 50–70% and decreased mortality.[4]

When intramuscular injections are not possible, oral penicillin V (250 mg, twice daily) can be prescribed. In cases of penicillin sensitivity, oral erythromycin (children 40 mg/kg/day in two to four divided doses, adolescents and adults 400 mg, twice daily) is the treatment recommended by the National Heart Foundation.[4]

Patients receiving prophylactic treatment should only stop receiving BPG injections following specialist consultation.

Further resources

New Zealand guidelines and educational materials are available from the National Heart Foundation website:

- Algorithm for sore throat management: www.heartfoundation.org.nz (key word “sore throat”)
- Educational material for patients and whānau: www.heartfoundation.org.nz (key word “rheumatic fever”)
- Guidelines for Rheumatic fever and Jones criteria for ARF diagnosis: www.heartfoundation.org.nz (key word “rheumatic fever diagnosis”)

Community success stories

There is significant evidence that community driven detection and treatment strategies do reduce ARF. A 2009 meta-analysis assessed the effectiveness of community/school based sore throat intervention programmes in Auckland, Hawaii, the United States and Cuba, and found, that on average, they reduced rates of ARF by 60%.[19]

In recent years, there have been several school-based initiatives in New Zealand. In the Northland community of Kaeo, Whangaroa, pre-intervention rates of ARF in children aged five to 14 years were 424 per 100 000. In the eight years following the intervention programme, which started in 2002, there have been no reported cases of ARF.

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

The success of the Whangaroa programme has resulted in the launch of other school-based programmes. In October 2009 a similar project was launched in Opotiki.

Details of the Opotiki programme included:

- Contacting GPs before the launch to gain support and provide guidance
- Swabbing children with sore throats three times a week to ensure no more than nine days passed between onset and treatment
- Referral of all children with positive cultures to a medical centre
- Oral antibiotics for ten days for all GAS positive students
- Raising specific community awareness of the link between sore throats and ARF

For several weeks in October 2009, when the programme started, 20% of all children with sore throats tested positive for GAS. By December 2009 the rate had dropped to 11% and further decreased to 3% by December 2010. Identical programmes have been launched in Kawarau (February 2011) and Murupara (May 2011).

Whānau Ora - Community solutions for community problems

In another example of forward thinking, the Mangere Community Health Trust PHO is overseeing the introduction of instant, on-the-spot, GAS testing at Makaurau Marae, Mangere. Trust Chair, Dr Michael Lamont explains:

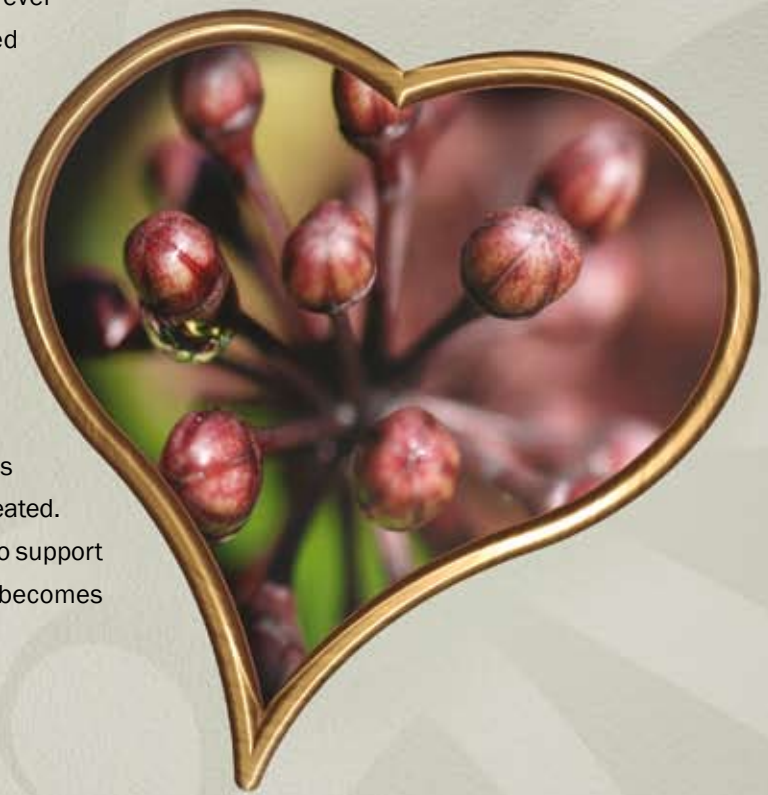
> "A major reason why some communities have high rates of ARF is that the chain of events from sore throat to antibiotic administration is much too long. The chain can be broken at any point, resulting in the child missing treatment.
>
> Our solution is to do 'near patient testing' in the community and to make swabbing a simple and easy choice. The old saying – make the decision the easiest and simplest thing to do!

The GAS throat swab we have imported is 100% sensitive and 100% specific.[20] It will be used on a marae in Mangere where many of the Kuia are on their second set of heart valves."

Examples of the media campaigns can be seen at: www.toiteorapublichealth.govt.nz/sitesearch/?k=rheumatic+fever.

Sore throats break hearts

Community programmes are not just about swabbing children's throats. Increasing community understanding of the link between GAS sore throats and ARF is also a key component of their success. In Bay of Plenty, a multi-media campaign was launched with locally-affected children fronting local campaigns. Press releases, advertorials and commissioned articles appeared in newspapers, radio adverts were aired on eight stations, newsletters distributed at schools and a rheumatic fever website created. Rheumatic fever presentations for local GPs were also conducted at CME sessions. In larger metropolitan areas, or in sparsely populated regions where school-based projects are impractical, this raising of specific community awareness assumes even greater significance.



In the 2011 Budget the Government announced an additional $12 million in funding allocated to combating ARF. It is anticipated that more school-based programmes and supporting multi-media campaigns will be created. GPs in high to medium risk areas may be asked to support such programmes as this increased funding becomes available.

ACKNOWLEDGEMENT Thank you to **Professor Norman Sharpe**, Director, National Heart Foundation and **Dr Jim Miller**, Medical Officer of Health and **Lindsay Lowe**, Communicable Disease Nurse, Toi Te Ora Public Health Service, Bay of Plenty DHB for expert guidance in developing this article.

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