
Rheumatic fever

Epidemiology in New Zealand

The incidence of rheumatic fever in New Zealand is much higher than in comparable countries and regions such as North America and the United Kingdom. Within New Zealand, the incidence varies greatly by geographic region and ethnicity. Māori and Pacific peoples, in particular, are disproportionately affected, for both acute rheumatic fever (ARF) and chronic rheumatic heart disease (RHD). Most cases of ARF are in children aged 5–14 years, although about one-third of cases occur in older teens and young adults.

For more detailed epidemiological information, see the Institute of Environmental Science and Research surveillance website (www.surv.esr.cri.nz).

ARF (including recurrence) is a notifiable disease; however rheumatic heart disease, in the absence of signs and symptoms of ARF, is not.

The purpose of ARF notification is to facilitate public health investigation and community education and to inform prevention strategies for addressing causative factors for cases and high-risk populations. Causative factors include economic deprivation, household crowding, poor health literacy and lack of access to health care. These factors prevent rapid investigation and effective treatment of group A *Streptococcus* (GAS) pharyngitis and access to secondary prevention of recurrences.

When rheumatic fever first became notifiable in 1986, guidance was given to medical professionals that presumed rheumatic heart disease in patients under the age of 20 years should be notified to the local medical officer of health (Department of Health circular letter to Medical Practitioners HP 1/87, January 1987). Notification of rheumatic heart disease under the age of 20 years is no longer required as the diagnosing medical professional is responsible for ensuring cases of rheumatic heart disease that require secondary prophylaxis receive active clinical follow-up. Local registers are useful to facilitate active follow-up and help prevent cases from being lost to follow-up. ARF registers in New Zealand have been shown to be effective at reducing admissions for ARF recurrences (National Heart Foundation 2006).

Case definition

Clinical description

ARF is an autoimmune consequence of a throat infection caused by the bacterium GAS, that is, *Streptococcus pyogenes*. 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. All suspected cases of ARF should be referred to hospital for specialist assessment, investigation, education and treatment.

Laboratory test for diagnosis

ARF is a clinical diagnosis (see ‘Case classification’). Currently, there is no single laboratory test for ARF. Laboratory tests for evidence of preceding GAS infection are described below.

Case classification

The diagnosis of ARF relies on health professionals being aware of the diagnostic features of the condition, particularly when presentation is delayed or atypical. Diagnostic certainty may vary according to location and ethnicity. Diagnosis is largely based on the Jones criteria, which are divided into major and minor manifestations based on their prevalence and specificity. The original Jones criteria were modified in 1992 and reconfirmed by the World Health Organization in 2004 (WHO 2004) (see Table 1).

ARF episodes can be classified as initial attacks (no known past history of ARF) or recurrent attacks (an episode in a person with a known past history of ARF that fulfils the criteria for a suspect, probable or confirmed case or previously diagnosed rheumatic heart disease).

The case classification for both initial and recurrent attacks is described in Table 2 below. The table also describes how the classification aligns with the categories used in the *New Zealand Guidelines for Rheumatic Fever: 1. Diagnosis, management and secondary prevention* (National Heart Foundation 2006). Referral to a rheumatic fever register may still be recommended for some people who do not meet the case definitions (see ‘Reporting’).

Table 1: Jones criteria for acute rheumatic fever

Manifestation	Criteria
Major manifestations ² modified from Jones 1992	Carditis (including evidence of subclinical rheumatic valve disease on echocardiogram) ¹ Polyarthritis ² (or aseptic monoarthritis; refer to National Heart Foundation 2006 for further information) Chorea (can be stand-alone for definite/confirmed initial or recurrent ARF diagnosis) Erythema marginatum Subcutaneous nodules
Minor manifestations	Fever Raised ESR or CRP ³ Polyarthralgia Prolonged P-R interval on ECG ⁴

Note:

- 1 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.
- 2 Other causes of arthritis/arthralgia should be carefully excluded, particularly in the case of monoarthritis, eg, septic arthritis (including disseminated gonococcal infection), infective or reactive arthritis and auto-immune arthropathy (eg, 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. References from National Heart Foundation (2006).
- 3 ESR = Erythrocyte sedimentation rate; CRP = C-reactive protein.
- 4 ECG = electrocardiogram.

Table 2: Case classification and diagnostic criteria for acute rheumatic fever

Case classification	Heart Foundation guidelines' diagnostic category	Diagnostic criteria
Under investigation	n/a	<ul style="list-style-type: none"> A case that has been notified, but information is not yet available to classify it as suspect, probable or confirmed
Suspect	Possible ARF	<ul style="list-style-type: none"> Strong clinical suspicion of ARF Insufficient signs and symptoms to fulfil diagnosis of confirmed or probable ARF
Probable	Probable ARF	<ul style="list-style-type: none"> Evidence of preceding group A streptococcal infection from positive throat culture or rapid antigen test Two major, or one major and two minor manifestations in the Jones criteria (see Table 1) <p>Or</p> <ul style="list-style-type: none"> Serological evidence of a preceding group A streptococcal infection One major and one minor manifestation
Confirmed	Definite ARF	<ul style="list-style-type: none"> Serological evidence of preceding group A streptococcal infection¹ Two major, or one major and two minor manifestations in the Jones criteria (see Table 1) are present <p>Or</p> <ul style="list-style-type: none"> Chorea (other major manifestations or evidence of group A streptococcal infection not required)
Not a case	n/a	<ul style="list-style-type: none"> A case that has been investigated and subsequently found not to meet the case definition

Note:

- 1 Elevated or rising streptococcal antibody titres are essential for confirming preceding GAS infection. Other laboratory tests, including culture and rapid antigen test, cannot distinguish between infection and carriage.

Spread of infection

ARF is not infectious but the precursor condition GAS pharyngitis is moderately infectious within households. For further information on GAS management, see the *New Zealand Guidelines for Rheumatic Fever: 2. Group A streptococcal sore throat management* (National Heart Foundation 2008).

Notification procedure for ARF

It is expected that the attending medical practitioner will notify the local medical officer of health of suspected initial or recurrent cases of ARF within seven days. Notification should not await a confirmed diagnosis (see also 'Reporting').

If cases of ARF are identified through other processes, such as audits, there is no legal requirement for the audit team to notify these cases to the local medical officer of health or for these cases to be recorded on EpiSurv (although cases identified through audit activities should still be entered into the local register if prophylaxis or follow-up is indicated). However, if a case identified via audit is notified to a medical officer of health by the attending medical practitioner, this case should be recorded on EpiSurv.

Management of a case of ARF

Investigation

- **Initial attack:** Ascertain if the case has had sufficient investigation to confirm diagnosis (ie, throat swab, serology, ESR/CRP, echocardiogram, ECG). Any GAS isolated from the throat of a person suspected of having ARF should be referred to the Institute of Environmental Science and Research for emm typing. Obtain a history of possible household contacts and recent throat infection. See also 'Reporting'.
- **Recurrent attack:** Follow the procedure as above for initial attack but also investigate the reason for recurrence. Recurrent attacks may represent a treatment or systems failure and should be investigated.

Restriction

Acute rheumatic fever

Cases of ARF do not require isolation unless they have known or suspected acute GAS pharyngitis. For information on GAS management, see *New Zealand Guidelines for Rheumatic Fever: 2. Group A streptococcal sore throat management* (National Heart Foundation 2008).

Treatment

Ideally all those with suspected ARF (first episode or recurrence) should be hospitalised as soon as possible after onset of symptoms, and should be under the care of a specialist paediatrician or physician. The main priority in the first few days after presentation is confirmation of the diagnosis. The treating clinician is responsible for treatment, prophylaxis, education, dental referral, notification to public health, and informing the case's general practitioner.

Treatment options for arthritis/arthralgia, fever, carditis/heart failure and chorea are outlined in the *New Zealand Guidelines for Rheumatic Fever: 1. Diagnosis, management and secondary prevention* (National Heart Foundation 2006).

One episode of rheumatic fever significantly increases the risk of further episodes, often with further cardiac damage. Antibiotic prophylaxis to prevent recurrent attacks of rheumatic fever should therefore be started before discharge from hospital. The appropriate duration of secondary prophylaxis depends on a number of factors, including age, clinical pattern, environment and time elapsed since the last episode of ARF.

All cases should receive regular primary care review, and outpatient follow-up should be initiated before discharge from hospital.

Rheumatic heart disease leads to a lifelong increased risk of bacterial endocarditis, and antibiotic prophylaxis may be required at the time of dental, oral, respiratory tract, oesophageal, gastrointestinal and genitourinary procedures. Ongoing dental care is essential, and each case should be notified to the appropriate school dental service or dentist.

Counselling

At the time of diagnosis, it is essential to explain the disease process to the case and their family in a culturally appropriate way. On discharge, all cases should have a good understanding of the cause of rheumatic fever and the need for any family member to have sore throats treated early. Cases and their families should understand the consequences of missing antibiotic doses. Also remind them of the importance of additional antibiotic prophylaxis for dental and other procedures to protect against endocarditis.

Management of contacts

Clustering of cases of rheumatic fever in families has been documented for more than a century. Familial clustering persists when socioeconomic factors and environment are controlled for, suggesting there is some inherited susceptibility to rheumatic fever.

Definition

Contacts include all people in close contact with a case (for example, members of the case's household) during the period up to one month before the onset of illness in the case.

Investigation

All household contacts of the index case should have a throat swab if the contact was within one month of onset of ARF. GAS isolated from a household contact should be referred to the Institute of Environmental Science and Research for emm typing.

Emm typing of GAS isolated from household contacts may aid understanding of circulating GAS strains in household contacts of cases of ARF and may inform our knowledge of rheumatogenic strains in New Zealand.

Note: There is little evidence available with which to evaluate how effective contact tracing is in preventing future cases of ARF. However, streptococcal acquisition rates of 25% or greater have been recorded in family contacts of GAS pharyngitis.

For further information on GAS management, see the *New Zealand Guidelines for Rheumatic Fever: 2. Group A streptococcal sore throat management* (National Heart Foundation 2008).

Restriction

Asymptomatic contacts do not need to be restricted.

Treatment

For information on treatment of contacts diagnosed with GAS pharyngitis, see the *New Zealand Guidelines for Rheumatic Fever: 2. Group A streptococcal sore throat management* (National Heart Foundation 2008).

Counselling

Advise contacts about GAS throat infection as well as its mode of transmission and the relationship of untreated disease with ARF. Also provide education on respiratory hygiene. Advise all contacts to seek early medical attention if a sore throat develops.

Other control measures

A case of ARF can be an indicator of high GAS load in the case's community. Therefore, a case of ARF in a community may warrant a range of control measures aimed at addressing GAS transmission. For more information about when and how to implement community-wide strategies to reduce rheumatic fever rates, see the National Heart Foundation's *New Zealand Guidelines for Rheumatic Fever: 3. Proposed rheumatic fever primary prevention programme* (National Heart Foundation 2009).

Strategies that address the multiple determinants of rheumatic fever are more likely to have long-term success, including:

- prevention of transmission of GAS infections, for example, by addressing household crowding and socioeconomic factors that predispose to it
- early detection and treatment of GAS infections, for example, by improving health literacy, health service access and early diagnosis and treatment (community- or school-based interventions may be useful)
- early diagnosis of ARF to reduce the risk of severe rheumatic heart disease
- ensuring good follow-up for antibiotic prophylaxis (secondary prevention) for those with a diagnosis of ARF.

Health education

Schools and general practitioners should be alerted to a case of ARF in the community. The community should be educated on the relationship between streptococcal sore throats, ARF and RHD. Public health providers should promote the key messages of the rheumatic fever prevention programme with population groups that have a high incidence of ARF. Additionally, such professionals should educate on respiratory hygiene, referral systems between health, housing and social welfare sectors and the importance of completing a full course of antibiotics.

Reporting

Ensure complete case information is entered into EpiSurv. Demographic and other risk factor/exposure information on the case report form is used to inform the public health response. For instructions on completing the case report forms, see the EpiSurv website (www.surv.esr.cri.nz/episurv/crf.php).

If a cluster of cases occurs, discuss it with the Director of Public Health at the Ministry of Health.

In addition to public health notification and recording on EpiSurv, all cases of ARF (suspect, probable and confirmed) should be referred to and recorded on a clinical register to ensure appropriate follow-up and any necessary antibiotic prophylaxis.

Cases of RHD that require secondary prophylaxis should also be recorded on a register.

References and further information

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