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 and chronic rheumatic heart disease. Most cases of acute rheumatic fever are in children aged 5 to 14 years although cases can occur in older teens and young adults.

Acute rheumatic fever is a notifiable disease, and this includes recurrence of the condition. Presumed rheumatic heart disease under the age of 20 years is also notifiable, even in the absence of a confirmed acute episode; this should be classified separately (see ‘Case classification’ below).

More detailed epidemiological information is available on the Institute of Environmental Science and Research (ESR) surveillance website at www.surv.esr.cri.nz

Case definition

Clinical description

Acute rheumatic fever (ARF) is an autoimmune consequence of throat infection with the bacterium group A Streptococcus (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. Individuals with ARF are often severely unwell, in great pain and require hospitalisation.

The diagnosis of ARF relies on health professionals being aware of the diagnostic features, particularly when presentation is delayed or atypical. Currently there is no single laboratory test for ARF, so diagnosis remains a clinical decision. Diagnostic certainty may vary according to location and ethnicity.

The Jones criteria for diagnosing ARF divide the diagnostic features of ARF 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 (WHO) in 2004 (WHO 2004).

Major manifestations are:

• carditis
• polyarthritis
• erythema marginatum
• chorea
• subcutaneous nodules.

Minor manifestations are:
• arthralgia
• fever
• elevated acute phase reactants: erythrocyte sedimentation rate; C-reactive protein; prolonged PR interval.

**Laboratory test for diagnosis (of group A streptococcal infection)**

Diagnosis of ARF is based on the Jones criteria and evidence of a preceding GAS throat infection (see ‘Case classification’). Currently there is no single laboratory test for ARF.

Tests for diagnosing GAS throat infection include:
• elevated or rising anti-streptococcal serology: all of antistreptolysin O and antiDNase B titres
• positive throat culture for GAS
• elevated rapid diagnostic test for GAS.

**Case classification**

- **Under investigation**: A case that has been notified, but information is not yet available to classify it as probable or confirmed.
- **Probable**: Initial attack; evidence of a preceding GAS throat infection and a major manifestation but does not fulfil the complete Jones criteria.
- **Confirmed**: Initial attack; evidence of a preceding GAS throat infection (preferably elevated or rising antistreptolysin O or other streptococcal antibody, National Heart Foundation 2006, p 15) and fulfils the Jones criteria – that is, two major manifestations, or one major and two minor manifestations, are present.
- **Not a case**: A case that has been investigated and subsequently found not to meet the case definition.

Cases can be also be classified as follows:

- **Recurrent attack**: A case with a known past history of ARF that fulfils the criteria as for an initial attack (either probable or confirmed).
- **Presumed rheumatic heart disease (under the age of 20)**: not a case of acute rheumatic fever but a diagnosis or strong clinical suspicion of rheumatic heart disease.
Exceptions

Even in the absence of evidence of a preceding group A streptococcal infection the following should be reported as they may be the only manifestation of rheumatic fever:

- chorea – this can be readily diagnosed on the basis of history, physical examination and laboratory evaluation (National Heart Foundation 2006)

- indolent carditis – defined as carditis of insidious onset and slow progression (Gaasch 1992), especially in cases who present for medical diagnosis months after the onset of rheumatic fever.

Spread of infection

Incubation period

GAS pharyngo-tonsilitis begins 1–5 days after exposure. Scarlet fever typically starts 1 day after the throat symptoms. Rheumatic fever occurs 1–5 weeks (mean 19 days) after the throat infection.

Mode of transmission

GAS transmission is by respiratory droplets or direct contact with a case or carrier. Individuals with acute upper respiratory tract infections are particularly likely to transmit infection. It is rarely transmitted by indirect contact through fomites. There have been well-documented explosive outbreaks of streptococcal pharyngo-tonsilitis associated with contaminated food or milk, and some of these may be due to rheumatogenic strains.

Period of communicability

Cases with untreated GAS pharyngo-tonsilitis may carry the organism for weeks or months, but contagiousness decreases sharply in the 2–3 weeks after onset of infection. Penicillin treatment generally reduces transmissibility within 24 hours.

Notification procedure

Attending medical practitioners or laboratories must immediately notify the local medical officer of health of suspected cases. This includes acute or recurrent cases of rheumatic fever, or presumed rheumatic heart disease under the age of 20. Notification should not await confirmation.
**Management of case**

**Investigation**

- **Initial attack:** In the event of an outbreak, attempt to identify the M-type of the strain as this is most often associated with rheumatic fever. Obtain a history of possible household contacts and recent throat infection.

- **Recurrent attack:** As above for initial attack but also investigate the reason for recurrence. Recurrent attacks may represent a treatment failure or systems failure and should be investigated.

**Restriction**

All individuals with known or suspected GAS pharyngo-tonsilitis require droplet isolation precautions until 24 hours after antibiotic treatment has been started. Individuals with GAS pharyngo-tonsilitis should not handle milk or food for other people until 24 hours after antibiotic treatment has been started.

**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.

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).

The notifying medical practitioner should also make direct contact with community cases to ensure that they are aware of the diagnosis, the need for secondary prophylaxis and any other specific follow-up requirements. Where relevant, it is also important for consent to be obtained from the case (or caregiver) to inform other health care providers, such as their local Māori or Pacific provider, about the illness.

One episode of rheumatic fever significantly increases the risk of recurrence, 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 that the disease process is explained 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. They should also be reminded of the importance of additional antibiotic prophylaxis for dental and other procedures to protect against endocarditis.

**Management of contacts**

Identify contacts for investigation, treatment and counselling where indicated.

**Definition**

All people in close contact with a case (for example, the case’s household) during the period up to 1 month before the onset of illness in the case.

**Investigation**

Streptococcal acquisition rates of 25% or greater have been recorded in family contacts of GAS pharyngitis.

All household contacts (regardless of symptoms) of the index case of ARF, who are aged 3 years and older should have a throat swab for GAS if the contact was no longer than one month after onset of ARF in the index case.

If there are three or more cases of GAS pharyngitis within a household in a 3-month period, throat swabs for GAS should be considered for all household members (regardless of symptoms).

Refer to the algorithms in the National Heart Foundation’s *Guidelines for Rheumatic Fever*.

**Restriction**

Children with GAS pharyngo-tonsilitis should not attend school or an early childhood service or have close contact with other children, if possible, until 24 hours after antibiotic treatment has started. Individuals with GAS pharyngo-tonsilitis should not handle milk or food for other people until 24 hours after antibiotic treatment has been started.
**Treatment**

In partnership with the primary health care provider, treat contacts identified with positive GAS cultures with an appropriate antibiotic. See Algorithm 4 of the *New Zealand Guidelines for Rheumatic Fever: 2: Group A streptococcal sore throat management* (National Heart Foundation 2008). Ten days of treatment is required for oral antibiotics.

Post-treatment throat swabs are not recommended unless:

- the case has a history of rheumatic fever and is not receiving prophylactic IM penicillin
- the case develops GAS pharyngitis during an outbreak of ARF or post streptococcal glomerulonephritis
- the case developed GAS pharyngitis during an outbreak in a closed or partially closed community
- there is recurrent GAS pharyngitis within the family/household
- the case remains symptomatic after completing their full course of antibiotics.

**Counselling**

Contacts should be advised on sore throats, GAS throat infection and its mode of transmission and the relationship of untreated disease with ARF. Educate on respiratory hygiene. Advise all contacts to seek early medical attention if symptoms develop.

**Other control measures**

High incidence rates of rheumatic fever in a community or school may constitute an outbreak or epidemic that warrants a range of control measures. More information about when and how to implement community-wide strategies to reduce rheumatic fever rates is contained in the National Heart Foundation’s *Guidelines for Rheumatic Fever*.

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

- primary prevention of GAS infections, for example, by improving wider socioeconomic factors, such as housing conditions
- early treatment of GAS infections, for example, by improving health service access and early diagnosis and treatment (community- or school-based interventions may be useful)
- ensuring good follow-up for antibiotic prophylaxis (secondary prevention).

**Identification of source**

Not applicable.
Disinfection
Clean and disinfect surfaces and articles soiled with respiratory secretions from infectious GAS cases.

Health education
Encourage early presentation for streptococcal sore throat, especially in families or other groups where there is a history of rheumatic fever. Educate the public on the relationship between streptococcal infection and subsequent ARF and rheumatic heart disease. Reinforce the importance of completing a full course of antibiotics (for example, 10 days of penicillin) for streptococcal pharyngitis. Promote the message ‘sore throats matter’ with population groups that have a high incidence of ARF.

Reporting
Ensure complete case information is entered into EpiSurv.
If a cluster of cases occurs, discuss with the Communicable Diseases Team at the Ministry of Health and outbreak liaison staff at ESR, and complete the Outbreak Report Form.

References and further information

