

ACUTE RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE

Public health priority: Routine

PHU response time: Respond to confirmed and probable cases within 3 working days. Enter confirmed and probable cases on NCIMS within 5 working days.

Case management: Work with key contacts in the LHD to enrol cases onto the Rheumatic Heart Disease Register to enable effective long-term management.

Contact management: None

Created: October 2015

1. Reason for surveillance

- To monitor the epidemiology of acute rheumatic fever (ARF) and rheumatic heart disease (RHD) in New South Wales.
- To prevent recurrence of ARF and prevent or limit the severity of RHD by enhanced case follow up through enrolment on the NSW RHD register.

2. Case definition

Acute Rheumatic Fever

A confirmed case¹ requires

- Clinical definitive evidence **AND** Laboratory suggestive evidence **OR** Rheumatic (Sydenham's) Chorea (with other forms of chorea excluded).

A probable case requires

- Clinical definitive evidence **OR** Clinical suggestive evidence and laboratory suggestive evidence **AND** where ARF is considered the most likely diagnosis by the treating clinician.

A possible case requires

- Clinical definitive evidence **OR** Clinical suggestive evidence and laboratory suggestive evidence **AND** where the treating clinician has less confidence about ARF as the correct diagnosis, but other differential diagnoses have been excluded.

Clinical definitive evidence – initial episode

The presence of two major manifestations **OR** one major and two minor manifestations.

Clinical definitive evidence – recurrent episode²

The presence of two major manifestations **OR** one major and one minor manifestations **OR** three minor manifestations.

¹ Adapted from RHD Australia, National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edition). 2012.

² In a patient with known past ARF or RHD.

	High risk groups ³	All other groups
Major manifestations	<ul style="list-style-type: none"> • Carditis (including subclinical rheumatic valvulitis detected by echocardiogram) • Polyarthritits (classically, but not necessarily, migratory in nature) or aseptic mono-arthritis or polyarthralgia • Erythema marginatum • Subcutaneous nodules 	<ul style="list-style-type: none"> • Carditis (including subclinical rheumatic valvulitis detected by echocardiogram) • Polyarthritits (classically, but not necessarily, migratory in nature) • Erythema marginatum • Subcutaneous nodules
Minor manifestations	<ul style="list-style-type: none"> • Fever (temperature $\geq 38^{\circ}\text{C}$, or a reliably reported fever during the current illness) • Monoarthralgia (not counted if arthritis or arthralgia already a major manifestation) • Elevated acute phase reactants (CRP ≥ 30 mg/L and / or ESR ≥ 30 mm/hr) • Prolonged PR interval on ECG (not counted if carditis already a major manifestation) 	<ul style="list-style-type: none"> • Fever (temperature $\geq 38^{\circ}\text{C}$, or a reliably reported fever during the current illness) • Polyarthralgia or aseptic monarthritits (not counted if polyarthritits already a major manifestation) • Elevated acute phase reactants (CRP ≥ 30 mg/L and / or ESR ≥ 30 mm/hr) • Prolonged PR interval on ECG (not counted if carditis already a major manifestation)

Clinical suggestive evidence – initial or recurrent

A clinical presentation that falls short by either 1 Major or 1 Minor manifestation.

Laboratory suggestive evidence

Supporting evidence of preceding group A Streptococcal (GAS) infection includes the following:

- GAS bacteria isolated from a throat swab
- Elevated or rising streptococcal antibody (ASOT, anti-DNase B) titres.

Streptococcal titres vary according to a number of factors, including age. The following titres are the upper limit of normal (ULN) for the given age groups:

Age group (years)	ASO titre (IU/mL)	Anti-DNase B titre (IU/mL)
1-4	170	366
5-14	276	499
15-24	238	473
25-34	177	390
≥ 35	127	265

Epidemiological evidence

Not applicable.

Rheumatic Heart Disease

A confirmed case (requiring notification)

- Clinical definitive evidence in a person less than 35 years of age

Clinical definitive evidence

An echocardiogram with valve changes consistent with RHD as defined by the World Heart Federation criteria.⁴

3. Notification criteria and procedure

ARF and RHD in people aged less than 35 years are to be notified by:

- Medical Practitioners and Hospital CEOs on diagnosis (ideal reporting by telephone or routine mail)

³ In NSW, Aboriginal people and Torres Strait Islanders, Maoris, Pacific Islanders, and immigrants from countries with a high prevalence of RHD should be considered high risk. Low risk groups include all other populations.

⁴ Reményi, B. et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease— an evidence-based guideline. Nat. Rev. Cardiol. 9, 297–309 (2012).

Only confirmed and probable cases should be entered onto NCIMS.

4. The disease

Infectious agent

ARF is an immune-mediated multi-system inflammatory disease that follows an infection by the GAS bacterium. Episodes of ARF can cause permanent damage to the heart valves leading to RHD, and the risk of permanent damage increases with each episode.

ARF remains the most common cause of acquired heart disease in children around the world. ARF incidence and RHD prevalence in remote Indigenous communities of Northern and Central Australia, Pacific Islanders and Maori are among the highest reported in the world. However, cases of ARF and RHD can occur in all population subgroups in Australia.

ARF and RHD are more common in females. They tend to run in families and a genetic susceptibility to the inflammatory response to GAS is recognised. The incidence of ARF peaks in the 5 to 14 year age group.

Mode of transmission

ARF and RHD are not communicable.

GAS infection is spread by large respiratory droplets or direct contact with infected individuals or carriers, and rarely via indirect contact through objects. Individuals with acute upper respiratory tract (especially nasal) infections are particularly likely to transmit infection.

Timeline

- GAS infection: the incubation period is usually 1 to 3 days, rarely longer.
- ARF: symptoms usually develop 1 to 5 weeks after the initial GAS infection.
- RHD: damage to the heart valves occurs during ARF and becomes apparent once the inflammation subsides. The severity of RHD increases with repeated episodes of ARF.

Infectious period

There is no infectious period for ARF and RHD, which are not communicable. GAS infection, the underlying cause, is infectious for 10 to 21 days in untreated, uncomplicated cases.

However, asymptomatic carriage may last for weeks to months in some individuals. Infectiousness decreases sharply 2 to 3 weeks after onset of infection. Adequate penicillin therapy will generally terminate transmissibility within 24 hours.

Clinical presentation

ARF is characterised by an acute, generalised inflammatory response that mainly affects the heart, joints, brain and skin. Individuals with ARF often require hospitalisation.

After the acute episode of ARF has resolved, the range of outcomes include: resolution without ongoing cardiac involvement, resolution with cardiac involvement ranging from mild to severe, death from fulminant carditis, and / or repeat episodes of ARF. Without antibiotic prophylaxis, more than 60 percent of ARF patients develop RHD within 10 years of their first episode of ARF⁵.

People who have had ARF previously are much more likely than the general community to have subsequent episodes in the absence of preventive measures. Recurrences of ARF may cause further cardiac valve damage. Hence, RHD steadily worsens in people who have multiple episodes of ARF. Outcomes of RHD include cardiac surgery to replace or repair faulty heart valves, cardiac failure, endocarditis and premature death.

The major and minor manifestations used for ARF diagnosis are described above (see section 2. Case Definition).

⁵ Lawrence JG, Carapetis JR, Griffiths K, Edwards , Condon JR. Acute Rheumatic Fever and Rheumatic Heart Disease: Incidence and Progression in the Northern Territory of Australia 1997 – 2010. *Circulation* 2013.

Diagnosis

The diagnosis of ARF is primarily a clinical one supported by evidence of recent group A streptococcal infection and inflammatory markers (see section 2. Case Definition).

RHD is also a clinical diagnosis, using an echocardiogram to detect valve changes consistent with RHD (see section 2. Case Definition).

5. Managing single notifications

Response time

Investigation

Within 3 days of notification of confirmed and probable cases begin follow-up investigation.

Data entry

Within 5 working days of notification enter confirmed and probable cases on NCIMS.

Response procedure

The response to a notification will normally be carried out in collaboration with the case's health carers, the LHD RHD Coordinator and case manager. A culturally appropriate health worker should also be involved in the response if needed. PHU staff should work with their LHD RHD Coordinator to ensure the following action is undertaken:

- Contact the treating doctor to confirm the onset date, and to confirm the illness meets the case definition.
- Ensure the treating doctor is aware of the RHD National ARF/RHD clinical guidelines and offer to provide the link to the guidelines if appropriate (<http://www.rhdaustralia.org.au/resources/arf-rhd-guideline>).
- For confirmed and probable cases, coordinate the enrolment of the case onto the RHD Register. This may be done by the treating doctor, the LHD Coordinator, or the case manager.
 - Explain the disease and the benefits of prophylaxis to the patient / carer.
 - Explain the role of the Register, provide the relevant fact sheets and other patient education material. See Appendices 1-3.
- Assist in consenting the patient. If the doctor is unable to follow up with the patient to obtain consent, offer to arrange for the patient to be consented on the doctors' behalf, with his/her permission. For Aboriginal and Torres Strait Islander people, consent should be sought with the support of Aboriginal Health Workers where possible.
 - If consent is given, arrange for the consent form and enrolment form to be returned to the PHU. On receipt, scan the forms, attach to the NCIMS record and mark the NCIMS record to enrol them onto the RHD Register, then send the originals to the Rheumatic Heart Disease Coordinator.
 - If no consent is given, ensure the treating doctor and patient / carer are aware of the recommendations within the national guidelines for on-going management of ARF cases (see under Case Management, below).

Case management

Roles and responsibilities

Primary and secondary care providers responsibilities include:

- 1) The initial investigation and diagnosis of new cases of ARF and RHD.
- 2) Notification of all new and recurrent cases of ARF and RHD aged ≤35 years to the local PHU.
- 3) Obtaining consent to enrol patients with newly diagnosed ARF or RHD onto the RHD Register.
- 4) Provision of ongoing clinical management for patients with ARF and RHD.

LHD RHD Coordinator responsibilities include:

- 1) Coordination of discharge planning with hospital staff when clients are in-patients and ensuring information about follow-up appointments are sent to the Central RHD Coordinator.
- 2) Identification of an appropriate case manager for each case.
- 3) Work with clinical networks on active case finding.
- 4) Inform NSW RHD coordinator when patients complete the treatment period, move between LHDs or interstate, or wish to leave the register.
- 5) Work with the treating doctor and case manager to consent cases to the register.

- 6) Assist the case manager with strategies to retain patients in care when cases have missed appointment or are lost to follow-up.

Case Manager responsibilities include:

- 1) Liaison between patients and primary care providers to facilitate monthly penicillin injections and specialist appointments.
- 2) Provision of feedback on adherence to monthly prophylaxis and follow-up appointments to the register in response to reminders by the Central RHD Coordinator.
- 3) Alert the LHD Coordinator of patients who are lost to follow-up, moving between LHDs/interstate or wishing to leave the register.

The Central RHD Coordinator responsibilities include:

- 1) Sends monthly reminders of penicillin injections to case managers.
- 2) Receive responses from case managers on follow-up appointments and enters onto register.
- 3) Liaison with the LHD Coordinator regarding patients who are lost to follow-up or not accessing care.
- 4) Remove patients from register if requested to do so.
- 5) Supporting public health units in case classification.
- 6) Undertake active surveillance for people newly diagnosed with ARF.

Investigation and treatment

The responsibility for ongoing management of patients with ARF and RHD rests with the treating doctor.

It is recommended that all patients with suspected ARF are admitted to a hospital for specialist paediatric or cardiology review and echocardiography, in order to maximise the likelihood of an accurate diagnosis, and to ensure prompt and optimal treatment.

Secondary prophylaxis and RHD control

Secondary prophylaxis with benzathine penicillin G (BPG) by intra-muscular injection (IMI) is recommended for all people with a history of ARF or RHD. Currently the recommended first line treatment is a BPG injection given every 21-28 days, except in patients considered to be at high risk⁶, for whom 21 day administration is recommended. Adherence to the recommended frequency is essential to prevent recurrence and disease progression. Oral antibiotics are not recommended, except following a documented penicillin allergy.

All people with ARF or RHD should continue secondary prophylaxis for a minimum of 10 years after the last episode of ARF, or until the age of 21 years (whichever is longer). Those with moderate or severe RHD should continue secondary prophylaxis up to the age of 35–40 years.⁷

ARF patients without evidence of RHD or mild RHD should have yearly medical and dental reviews, including echocardiograms every two years for children and every two to three years for adults still receiving secondary prophylaxis. Patients with evidence of moderate or severe RHD require more intensive follow-up schedules depending upon the severity of the disease. This will include regular specialist review and echocardiography.

Education

Provision of patient education is the responsibility of the treating doctor, but the case manager or other appropriate health care worker could be used to deliver it. PHUs should provide the treating doctor with the ARF factsheet as part of the procedure to seek consent from the patient to be entered onto the RHD Register and link the doctor to the resources available on the Rheumatic Heart Disease Australia website <http://www.rhdaustralia.org.au/>.

Exposure investigation

Not applicable.

Isolation and restriction

⁶ Patients at high risk are those with moderate or severe carditis or a history of valve surgery, or those who have confirmed breakthrough ARF, despite full adherence to 4-weekly BPG.

⁷ RHD Australia, National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edition). 2012.

None

Environmental evaluation

The PHU should consider involvement of the environmental health team, particularly if more than one case has occurred in a community. The environmental health team should seek to identify rectifiable environmental factors predisposing to GAS infection, and discuss the potential for a Housing for Health or other community-level initiative with the Aboriginal Environmental Health Unit (if an Aboriginal community) or local council, housing provider/manager, relevant community leaders or other relevant stakeholders/service providers.

Appendix 1: ARF/RHD and RHD Register Factsheet

Appendix 2: RHD Register Consent Form

Appendix 3: RHD Register Patient Information Sheet

Appendix 4: NSW ARF plan