



Acute Rheumatic Fever (ARF) & Rheumatic Heart Disease (RHD)

CDNA National Guidelines for Public Health Units

Revision history

Version	Date	Revised by	Changes
1.0	30 April 2014	Acute Rheumatic Fever & Rheumatic Heart Disease Working group	Developed by the Acute Rheumatic Fever & Rheumatic Heart Disease Working Group
2.0	4 December 2017	Acute Rheumatic Fever & Rheumatic Heart Disease Working group Chair	Responded to CDNA member feedback

The Series of National Guidelines ('the Guidelines') have been developed by the Communicable Disease Network Australia and noted by the Australian Health Protection Principal Committee. Their purpose is to provide nationally consistent guidance to public health units (PHUs) in responding to a notifiable or significant disease event.

These guidelines capture the knowledge of experienced professionals and provide guidance on best practice based upon the best available evidence at the time of completion.

Readers should not rely solely on the information contained within these guidelines. Guideline information is not intended to be a substitute for advice from other relevant sources including, but not limited to, the advice from a health professional. Clinical judgment and discretion may be required in the interpretation and application of these guidelines. Wherever possible, local knowledge and community priorities should always be considered when implementing these guidelines.

The membership of the CDNA and the Australian Health Protection Principal Committee (AHPPC), and the Australian Government as represented by the Department of Health do not warrant or represent that the information contained in the Guidelines is accurate, current or complete. The CDNA, the AHPPC and the Australian Government do not accept any legal liability or responsibility for any loss, damages, costs or expenses incurred by the use of, or reliance on, or interpretation of, the information contained in the guidelines.

Acute rheumatic fever and rheumatic heart disease are sentinel conditions of poverty and of health inequality; their persistence mark the failure of our health and other systems to address the non-communicable diseases of the disadvantaged. While this document does not attempt to address all the social determinants of health there are important considerations for a public health response which should be taken into consideration when planning public health interventions (including but not limited to Environmental Health) specifically in addressing the Closing the Gap National targets. Recommendations made with regards to the social determinants of health, including Environmental Health may be beyond the scope of the Public Health workforce responding to an individual case or an outbreak, but have been included for consideration.

Endorsed by CDNA: 27 November 2017

Endorsed by AHPPC: 03 July 2018

Release of Health: 01 August 2018

ACUTE RHEUMATIC FEVER (ARF) & RHEUMATIC HEART DISEASE (RHD)

CDNA National Guidelines for Public Health Units

Contents

Revision history.....	1
1. Summary	5
Public health priority	5
Routine	5
Case management	5
Contact management	6
2. The disease	6
Infectious agent.....	6
Reservoir.....	6
Mode of transmission	6
Incubation period.....	7
Infectious period	7
Clinical presentation and outcome.....	7
Diagnosis of primary ARF	7
Major manifestations.....	8
Minor manifestations.....	8
Evidence of group A streptococcal infection	10
Quality of life impact of an acute episode	10
Outcome.....	10
Diagnosis of ARF recurrence.....	11
Persons at increased risk of disease	11
Disease occurrence and public health significance.....	11
3. Routine prevention activities	13
4. Surveillance objectives.....	18
Data management.....	18
Reporting requirements	18
5. Communications	18
6. Case definitions	19
7. Laboratory testing	20
8. Case management.....	21

Response times.....	21
Response procedure	21
Case investigation.....	21
Case treatment	21
Education.....	22
Isolation and restriction	22
Active case finding	22
9. Environmental evaluation.....	22
10. Contact management	23
Contact definition.....	23
Management of identified contacts	23
Education	23
Isolation and restriction	23
11. Special situations	23
ARF possible outbreak/clustering response.....	23
Components of outbreak response	24
12. References and additional sources of information	25
References	27
Appendices	30
Appendix 1: Sample Acute Rheumatic Fever Notification Form for jurisdictions where ARF is notifiable and an ARF/RHD register exists	30
Appendix 2: Example of an Enhanced Surveillance form for person diagnosed with ARF 32	
Appendix 3: World Heart Federation Criteria for the Echocardiographic Diagnosis of RHD in Individuals Aged ≤20 Years	34
Appendix 4: Public Health Unit checklist (details will vary significantly according to jurisdiction). Ensure all criteria are marked	35
Contact the patient (or caregiver) to:	35
Contact laboratory to:.....	36
Contact local Housing Authority to:	36
Other issues:.....	36
Jurisdictional specific issues	36

1. Summary

The priorities in the control and prevention of acute rheumatic fever (ARF) are:

1. Timely diagnosis with accurate recognition of clinical symptoms
2. Notification and reporting:
 - a. For ARF – Notification to Public Health Unit by clinician in Western Australia (WA), Northern Territory (NT), Queensland (QLD), South Australia (SA)* and New South Wales (NSW)
 - b. For Rheumatic Heart Disease (RHD) – Notification to Public Health Unit by clinician in Western Australia (WA), New South Wales (NSW) and South Australia (SA)*
3. Secondary prevention with regular benzathine penicillin G after ARF diagnosis.

The aim of this guideline is to summarise the public health aspects of ARF case management, including mitigation of progression to RHD, the chief complication of ARF. It is based on the existing Australian National Guideline (Second Edition, 2012).¹

Public health priority

Routine

ARF is a sporadic condition in the majority of instances. It has been estimated that a minority of individuals (approximately 3-5 per cent) infected with the causative organism, i.e. rheumatogenic strains of group A streptococcus (GAS), have an inherent susceptibility to developing the autoimmune sequelae which constitute ARF.

Although rare, ARF case clusters or outbreaks can occur and a public health response is required (refer to [Section 12](#) – References and additional sources of information).

Case management

Based on the Australian National Guideline¹ and 2015 Revised Jones Criteria,² case management includes:

- Assignment of ARF as a primary or recurrent case.
- Assignment of ARF diagnosis as Definite, Probable or Possible.
- Assignment of RHD diagnosis based on echocardiogram result as absent, mild, moderate or severe, then assignment of clinical priority status as Priority 1 (most severe), Priority 2 or Priority 3. These are denoted P1, P2, P3.
- Notification and reporting:
 - There is no ARF-specific diagnostic laboratory test. Notification therefore relies on clinical recognition and clinician-initiated reporting. Laboratory evidence of GAS infection is required for definite diagnosis of non-chorea forms of ARF, but this test is not specific for ARF, hence this is not a laboratory-notifiable condition.
 - Under-reporting is a recognized problem, due both to missed diagnosis, and a lack of recognition of the need to notify.
- Education for case and household/family by healthcare staff including the notifying clinician about primordial and primary preventive strategies to reduce future GAS infections and the subsequent impacts. Commencement of secondary preventive strategies

* Notification to Communicable Disease Control Branch in SA with inclusion on the RHD Register upon patient consent.

^a The 2012 National Guidelines¹ use the terminology 'probable - highly suspected' for probable, and 'probable – uncertain' for possible. These terms are interchangeable.

- Benzathine penicillin G intramuscular injection every 21 - 28 days 450mg for individuals <20kg, 900mg for individuals ≥20kgb.
- Ensuring follow up with appropriately-timed future echocardiograms, specialist and dental reviews, in accordance with established care plans based on priority status.¹

Contact management

- The secondary attack rate for sporadic ARF is well below the threshold required for instigation of contact treatment,³ therefore, treatment of household and/or close contacts is not required in sporadic cases.
- Group A streptococcal infections cluster in households or other living conditions characterised by over-crowding, hence education for households regarding prevention, and ensuring the availability of adequate health hardware, is strongly recommended.
- Contact management in potential outbreak scenarios is addressed in – [Sections 10 & 11](#) = Contact Management and Special Situations.

2. The disease

ARF is a preventable disease of socioeconomic disadvantage. Repeated episodes of ARF lead to cumulative cardiac valve damage, resulting in RHD. RHD is an important cause of premature morbidity and mortality in selected Australian populations, particularly Aboriginal and Torres Strait Islander people, Maori, and migrants from Pacific nations. In addition to cardiac involvement other major manifestations include: carditis, arthritis, Sydenham's chorea, erythema marginatum, and subcutaneous nodules. Minor manifestations include arthralgia, fever, and elevated acute-phase reactants.

Infectious agent

The infectious agent is Group A-haemolytic streptococcus (*Streptococcus pyogenes* or GAS).

- The potential for non-A-haemolytic streptococci to cause ARF is acknowledged but remains speculative.^{4, 5}
- ARF is classically understood to follow GAS pharyngitis; however, circumstantial evidence from Northern Australia demonstrates that GAS skin infections (impetigo, pyoderma, skin sores) may be antecedents of ARF.^{1, 6-8}

An abnormal autoimmune host response to GAS is required for the development of ARF.

- Immune responses to GAS antigens cross-react with host tissues such as cardiac antigens.
- This occurs only in people with an inherent susceptibility (estimated to occur in 3 to 5 per cent of people), which may be at least partly genetically mediated. Understanding genetic susceptibility to ARF/RHD is complex⁹ but evidence is growing.¹⁰
- The autoimmune response leading to clinically apparent ARF probably only develops after repeated GAS exposures have occurred, accounting for the absence of ARF in infancy.

Reservoir

Humans are the sole reservoir for GAS.

Mode of transmission

GAS is transmitted human-to-human via:

- direct contact

^b See Section 9 for details including antibiotic option in confirmed penicillin allergy

- droplet spread from people with upper airways colonisation or carriage (pharynx, tonsils, nasal passages)
- contaminated fomites or surfaces (less common)

The importance of crowded living conditions to facilitate transmission has been well documented, e.g., the risk of GAS pharyngeal infection has been shown to be inversely proportional to the distance between a subject's bed and that of a colonised or infected case (see review¹¹). Poor skin health and household crowding is thought to contribute to the overall burden of GAS infection and carriage.

Poverty, household crowding, poor household and community hygiene as risk factors for ARF has recently been confirmed in a large ecological study of over 1000 rheumatic fever cases in New Zealand.¹²

Incubation period

The interval between GAS exposure and pharyngitis (which may be asymptomatic) or clinically apparent impetigo/pyoderma is 1-10 days.

The interval between GAS infection and onset of ARF varies depending on:

- ARF type which can manifest as one or a combination of carditis, arthritis, chorea, erythema marginatum or subcutaneous nodules.
- factors such as host immune response and whether the episode is primary or a recurrence.

In general, the following applies:

- Rheumatic carditis or arthritis – 2-3 weeks after GAS infection but can be as early as 1 week in recurrent ARF
- Sydenham's chorea – 6-9 weeks after GAS infection.

Infectious period

- The infectious period for GAS infection is 10 to 21 days in untreated, uncomplicated cases.
- Individuals with untreated streptococcal pharyngitis or asymptomatic carriage may carry the organism for weeks to months.
- Contagiousness of GAS decreases 2 to 3 weeks after onset of infection.
- Adequate penicillin therapy reduces the infective period to within 24 hours.

Clinical presentation and outcome

Diagnosis of primary ARF

A diagnosis of ARF can be made (a) after exclusion of differential diagnoses, such as disseminated gonococcal infection or systemic lupus erythematosus,¹ and (b) if the clinical and laboratory features fulfil the Australian modification of the Jones Criteria (now largely incorporated into the 2015 Revised Jones criteria²) (Refer to [Table 1](#)).

A high index of suspicion for ARF is required, especially in at-risk individuals. The Revised Jones Criteria recognise the need for a lower diagnostic threshold in high-risk groups (see [Table 1](#)). In Australia, the high-risk populations are:

- Aboriginal and Torres Strait Islander people, particularly those from northern Australia
- People of Maori or Pacific Islander background (Melanesia, Micronesia and Polynesia)
- Immigrants from developing countries

Although the recorded primary ARF episode is by definition the first episode to have been recognised and notified, many individuals have already had previous unrecognised ARF, evidenced by the finding of established RHD (as seen on echocardiogram) at the time of

'primary' ARF, or by performing a retrospective chart review and finding presentations consistent with ARF where the diagnosis was missed. Prior episodes may also have been inadequately symptomatic for the individual to have sought medical care. (See [Table 1](#))

Major manifestations

Carditis: active inflammation of the myocardium, endocardium and pericardium (i.e., pericarditis). The predominant manifestation of rheumatic carditis is involvement of the mitral and/or aortic valve endocardium presenting as a valvulitis. Echocardiography is the gold standard diagnostic modality, and echocardiogram criteria for rheumatic carditis diagnosis (and RHD diagnosis) are clearly defined.^{1, 13}

Arthritis: swollen and hot joint(s) with pain on movement. This is the most common presenting symptom of ARF. Rheumatic arthritis may have an abrupt onset and last for a few days to weeks and commonly involves the large joints either as a mono-arthritis (single joint) or polyarthritis (multiple joints). The inflammation is typically migratory (moves from one joint to another) with possible resolution of pain in one joint before onset in the next. Mono-arthritis is a major manifestation in high-risk groups (see below) but lower-risk groups require polyarthritis to be a major manifestation; for them monoarthritis is classified as a minor manifestation¹⁴.

Sydenham's chorea: abrupt, jerky, involuntary movements +/- associated muscular weakness and emotional lability. These uncoordinated movements especially affect the hands, feet and facial muscles, are often more severe on one side of the body and disappear during sleep. Chorea has the latest onset of timing of all ARF manifestations, with onset from weeks to months after initial streptococcal infection.

Erythema marginatum: rare (<2%); highly specific for ARF (considered pathognomonic), but other rashes can be easily mistaken for it. The rash can be difficult to detect in dark-skinned people. The rash appears early in the course of ARF and appears as bright, pink macules or papules that are non-pruritic, blanch under pressure, spread outwards in a circular pattern and may wax and wane over the course of a day. The rash may be found on the trunk or limbs but not on the face.

Subcutaneous nodules: also a rare (<2% of cases) but highly specific manifestation of ARF in Aboriginal people.¹⁵ They are 0.5–2 cm in diameter, round, firm, mobile, painless nodules occurring in crops of up to 12 over the elbows, wrists, knees, ankles, Achilles tendons, occiput and posterior spinal processes of the vertebrae. The nodules usually appear in a symmetric distribution, usually present in the first weeks of illness and may be associated with more severe forms of carditis.

Minor manifestations

Arthralgia: pain on joint movement without evidence of swelling or heat. It usually occurs in the same pattern as rheumatic polyarthritis (migratory, asymmetrical, affecting large joints). Polyarthralgia is a major manifestation in high-risk groups (see below) but classified as a minor manifestation in others. Monoarthralgia is a minor manifestation but only for high-risk groups.

Fever: accompanies most manifestations of ARF, with the exception of chorea.

Elevated acute-phase reactants: e.g., CRP, ESR: typically seen in ARF; less so in chorea.

Prolonged P-R interval on electrocardiogram (ECG): transient first-degree heart block or other conduction abnormalities (e.g. junctional rhythm) are hallmarks of rheumatic carditis; however, only P-R prolongation is included as a minor criterion. For P-R interval upper limits

of normal for different age groups, see [Table 1](#), last footnote. An ECG is required in all cases of suspected ARF.

Table 1 Australian Guidelines for the diagnosis of ARF (from Table 3.2, National Guideline) ¹

	High Risk†	All other groups
Definite initial episode of ARF	2 major or 1 major and 2 minor manifestations plus evidence of a preceding GAS infection‡	
Definite recurrent episode of ARF in a patient with known past ARF or RHD	2 major or 1 major and 1 minor or 3 minor manifestations plus evidence of a preceding GAS infection‡	
Probable ARF (first episode or recurrence)	A clinical presentation that falls short by either one major or one minor manifestation, or the absence of streptococcal serology results, but one in which ARF is considered the most likely diagnosis. Such cases should be further categorised according to the level of confidence with which the diagnosis is made: <ul style="list-style-type: none"> • Probable ('highly-suspected ARF') • Possible ('uncertain ARF') 	
Major manifestations	<ul style="list-style-type: none"> • Carditis (including subclinical evidence of rheumatic valvulitis of echocardiogram) • Polyarthritist†† or aseptic • mono-arthritis or polyarthralgia • Chorea§ • Erythema marginatum • Subcutaneous nodules 	<ul style="list-style-type: none"> • Carditis (excluding subclinical evidence of rheumatic valvulitis on echocardiogram) • Polyarthritist†† Chorea§ • Erythema marginatum* • Subcutaneous nodules
Minor manifestations	<ul style="list-style-type: none"> • Monoarthralgia • Fever‡‡ • ESR ≥30 mm/h or CRP ≥30 mg/L • Prolonged P-R interval on ECG§ 	<ul style="list-style-type: none"> • Fever‡‡ • Polyarthralgia or aseptic monoarthritis • ESR ≥30 mm/h or CRP ≥30 mg/L • Prolonged P-R interval on • ECG§§

†High-risk groups are those living in communities with high rates of ARF (incidence >30/100,000 per year in 5 to 14 year olds) or RHD (all-age prevalence >2/1000). Aboriginal people and Torres Strait Islanders living in rural or remote settings are known to be at high risk. Data are not available for other populations, but Aboriginal people and Torres Strait Islanders living in urban settings, Maori and Pacific Islanders, and immigrants from developing countries, may also be at high risk.

‡Elevated or rising antistreptolysin O or other streptococcal antibody, or a positive throat culture or rapid antigen test for GAS.

††A definite history of arthritis is sufficient to satisfy this manifestation. Note that if polyarthritis is present as a major manifestation, polyarthralgia or aseptic mono-arthritis cannot be considered an additional minor manifestation in the same person.

§Chorea does not require other manifestations or evidence of preceding GAS infection, provided other causes of chorea are excluded.

*Care should be taken not to label other rashes, particularly non-specific viral exanthemata, as erythema marginatum. ‡‡Oral, tympanic or rectal temperature ≥38°C on admission, or a reliably reported fever documented during the current illness.

§Upper limit of normal P-R interval is: ages 2-12 years: 0.16 seconds; ages 13-16 years: 0.18 seconds; ages 17+ years: 0.20 seconds. If carditis is present as a major manifestation, a prolonged P-R interval cannot be considered an additional minor manifestation.

Evidence of group A streptococcal infection

This can comprise positive antistreptolysin O titre (ASOT) or anti-DNAseB titre or isolation of group A-haemolytic streptococcus from throat swab; Refer to [Section 8](#) Case Management.

Quality of life impact of an acute episode

Although ARF symptoms may be subtle, and the key importance of ARF is the potential for progression to RHD, ARF itself is often a very burdensome illness.

- ARF requires hospitalisation, to facilitate diagnosis and commence appropriate management and education for the patient and the family.
- Joint pain and chorea can be very painful and temporarily disabling.
- Management is burdensome, comprising intramuscular benzathine penicillin G injections every 21-28 days for a minimum of 10 years, requiring a high level of ongoing engagement with health services.

Outcome

After primary ARF, outcomes range from full recovery with no permanent sequelae, to a fulminant disease course resulting in death. A recent NT study demonstrated that after a first ARF diagnosis, 61 per cent of people developed RHD within ten years. After RHD diagnosis, 27 per cent developed heart failure within 5 years¹⁶. Careful pregnancy planning in women with RHD of child-bearing age is an important consideration, given elevated foetal and maternal risks.¹⁷

The range of outcomes after a primary ARF episode includes:

- Resolution of acute episode with no cardiac involvement
 - A rebound phenomenon (flare or relapse of inflammatory and articular features) can occur on weaning or cessation of salicylate/non-steroidal anti-inflammatory drugs (NSAIDs)
- Resolution of acute episode with cardiac involvement ranging from mild to severe
- Subsequent ARF recurrence(s) after resolution of primary episode with development of new RHD or progression of existing RHD leading to the potential for early morbidity/mortality related to RHD complications
 - Atrial fibrillation
 - Heart failure
 - Endocarditis
 - Thromboembolic or haemorrhagic complications including stroke
- Death from fulminant carditis

ARF outcomes with regards to treatment requirements include:

- All individuals with definite or probable ARF: intramuscular benzathine penicillin G every 21-28 days for a minimum 10 years as per [Section 8](#) – Case Management
- Treatment options for RHD, depending on type and severity, which may include:
 - medical management of heart failure including ongoing echocardiograms and intramuscular Benzathine penicillin G injections requiring high level of ongoing engagement with health services
 - surgical management of valvular disease (repair or replacement)
 - life-long anticoagulation for certain prosthetic valve types or for atrial fibrillation

Diagnosis of ARF recurrence

Definition of ARF recurrence: new ARF episode occurring >90 days after a prior episode.

Diagnosis of ARF recurrence: there is a lower threshold for diagnosis of a recurrence compared with primary episode, as per line 3 of [Table 1](#).

Timing of recurrences: most recurrences occur within the first 10 years after the primary ARF diagnosis which is the basis for the recommendation to give benzathine penicillin G for this duration. The recurrence rate is highest in the first year, and drops annually thereafter.¹⁶

Persons at increased risk of disease

In Australia, the high-risk populations are:

- Aboriginal and Torres Strait Islander people, particularly those from northern Australia
- People of Maori or Pacific Islander background
- Immigrants from developing countries

People at risk of ARF are:

1. Children aged 5–14 years, with a peak at around 8 years
 - a. It is rare for ARF to occur in <3 year-olds ¹⁸ or > 40 year-olds, although recurrent ARF can occur beyond 40 years-old.
2. People living in a high-risk community
 - b. A high-risk community is one where high rates of ARF (incidence >30/100,000 per year in 5–14 year olds) or RHD (all-age prevalence >2/1000) are present ([Table 2](#))
3. Those with increased risk of exposure to GAS infection
 - a. Crowded living conditions
 - b. Inadequate 'health hardware' within homes or communities i.e., the physical equipment necessary for healthy, hygienic living
 - c. Lower levels of health literacy
 - d. Socioeconomic disadvantage
4. Those with an inherent susceptibility to the autoimmune response
 - e. As indicated in [Section 2](#) (Infectious agent), evidence for genetic susceptibility is growing, and remains under investigation in the Australian context and globally.

Disease occurrence and public health significance

Australian ARF rates are available for jurisdictions in which ARF is notifiable or where specific research on this question has been conducted.

There are several reasons why ARF and RHD are leading causes for public health concern. Firstly, ARF and RHD are indicators for socioeconomic disadvantage and highlight the requirement for major improvements in the social determinants of health. Secondly, ARF and RHD cause a major burden of premature morbidity and mortality for young Indigenous people in Australia. In the 2006 Australian Institute of Health and Welfare report on mortality in Australia, rheumatic and other valvular disease had the highest differential mortality ratio between Indigenous and non-Indigenous Australians for any clinical category; 19.1, in comparison to 18.2 for nephritis and nephrosis, 18.1 for diabetes, 4.3 for ischaemic heart disease and overall ratio 3.9.

Table 2: ARF incidence rates in selected Australian jurisdictions

Jurisdiction	ARF annual incidence (primary cases and recurrences)	Reference and details
Non-Indigenous	Almost non-existent	No national data on ARF incidence are

Jurisdiction	ARF annual incidence (primary cases and recurrences)	Reference and details
Australian born population		available. ¹⁹ Number of cases reported via the Australian Paediatric Surveillance Unit in non-Indigenous Australian children in 2007-10 was 10. ²⁰
Northern Territory (whole Top End and Central)	150-380 / 100,000	ARF incidence for NT-wide Aboriginal school age children, 2005-9. ¹⁹
Northern Territory (single Central Australian community)	815 / 100,000	ARF incidence for children aged 5-14 in one community, 1978-1987. ²¹
Kimberly, Western Australia	375 / 100,000	ARF incidence for Aboriginal children aged 5-14 in the Kimberley, 1988-1992. ²²
North Queensland	156 to 319 per 100,000	ARF incidence in north Queensland, 2004-2009. ²³
NSW	1.5-2 per 100,000*	ARF in 5 – 14-year-olds
South Australia	30 to 43 per 100,000	ARF incidence for SA Aboriginal Heart and Stroke Plan 2015. ²⁴

* NSW Admitted Patient Data Collection, 2003 – 2012, ICD-10 AM codes (100-102), excludes repeat admissions for individuals.

Source: SaPHaRI Centre for Epidemiology and Evidence, NSW Ministry of Health. (excludes recurrences)

3. Routine prevention activities

The current cornerstone of ARF/RHD preventive activities in Australia currently is secondary prophylaxis with an antimicrobial agent effective against GAS for those with diagnosed prior ARF/RHD

Levels of prevention for ARF/RHD and its sequelae, shown in Table 3, include:

- Primordial – improved social determinants of health
- Primary – treatment of GAS infection (no vaccination currently available)
- Secondary - antimicrobial prophylaxis after ARF e.g., intramuscular benzathine penicillin G every 21 – 28 days
- Tertiary – medical and surgical management of RHD, and consideration of echocardiographic screening in selected populations such as in pregnant women.

Primordial and primary preventive strategies classically refer to disease prevention in unaffected hosts and the general community however, these same messages are equally important to provide to individuals after ARF/RHD diagnosis. To potentially reduce their risk of recurrences; a new episode of ARF is estimated to be 10 times more common in an individual with past ARF than in those of similar age and living in the same community/circumstances, but without prior ARF.

Table 3 Preventive strategies targeting individuals with known ARF/RHD to prevent future ARF recurrence

Level of prevention	Background	Recommendations
Primordial	<ul style="list-style-type: none"> • ADF is very uncommon among population in developed countries due to high standards of living and access to improve health services • It is difficult to separate out the specific factors that have likely contributed to this elimination of ARF. Most of the apparent determinants (e.g., housing, education, poverty) lie outside the conventional health sector and there is a lack of interventional studies testing their relationship to ARF. • Observational studies provide evidence of the association between social and environmental factors with increased ARF risk over many decades.²⁵ • Crowding both at a bedroom and household level has been repeatedly associated with greater risks of ARF. A study of over 1000 ARF cases in New Zealand (NZ) demonstrated an incremental association of cases and index of household crowding^{**25} A similar relationship has been shown between pyoderma and number of people per bedroom in Australia's far north (pyoderma being chiefly attributable to GAS in the setting).⁸ • Military studies demonstrated that the frequency of GAS infections was directly related to cases' proximity to a known GAS carrier in their sleeping quarters, providing a biological basis for a relationship of crowding and ARF.²⁷ • Studies looking at income together with other factors suggest that low income in itself is a less important risk factor for ARF than its indirect impacts on other factors such as crowding, housing quality, nutrition and access to health care. <small>25, 26, 28-30</small> • GAS has been isolated from the environment of carriers, however the role of fomites in transmission remains unclear.^{31.32.33} • A 10-year evaluation of an Aboriginal Community Housing improvement program (Housing for health) across 71 communities in NSW to improve safety and healthy living practices (e.g., ability to wash people, clothes and bedding), noted a reduction of hospitalisation for respiratory illnesses, skin infections and intestinal infections of 40% compared to the rural. NSW communities that did not receive the Housing for health intervention 	<ul style="list-style-type: none"> • Provide education to housing providers and funding agencies, household members, communities, and local government about limited number of people per bed or bedroom** where possible and emphasising the importance of skin hygiene (i.e., frequent washing) to prevent transmission of GAS from potentially infected individuals. • Assist and support individuals diagnosed with ARF to improve their living conditions where required • Facilitate contact with the responsible agency (such as the Environmental Health Unit or relevant Housing Department) to ensure that malfunctioning toilets and taps (and/or hot water) are fixed and where necessary facilitate application for priority housing through Department of Housing. • Liaise with community environmental health units to ensure there is adequate and timely rubbish removal and that there are functioning public and private toilet facilities. • Ensure that the individual has access to functioning facilities for personal washing (including a basin for young children), and washing of clothes and bedding, through supply, installation and/or repair of running (hot) water soap, and a washing machine. • Facilitate application for priority housing through the Department of Housing. • There is no single standard measure of housing overcrowding in Australia. The Canadian National Occupancy Standard and the Proxy Occupancy Standard

Level of prevention	Background	Recommendations
		<p>are commonly used to measure overcrowding and recommend; (AIHW 2005).</p> <ul style="list-style-type: none"> ○ There should be no more than 2 persons per bedroom ○ Parents or couples may share a bedroom ○ Children <5 years of age of different sexes may reasonably share a bedroom ○ Children 5 years of age or over of the opposite sex should not share a bedroom ○ Children <18 years of age and of the same sex may reasonably share a bedroom ○ Single household members aged 18 years or over should have a separate bedroom <p>While this is difficult to achieve for a variety of reasons it should be acknowledged that this is an important precursor to GAS and other infectious disease transmission and that appropriate steps are taken to limit the number of people living in households.</p>
Primary	<ul style="list-style-type: none"> ● Primary prevention using school-based sore throat clinics is an important cornerstone of ARF prevention in NZ.³⁴ ● Treatment of GAS infection has been modelled as being cost effective in the South African settings.³⁵ ● GAS pharyngitis has been assessed as being rare in Aboriginal communities with high ARF rates making this approach challenging.⁸ Sore throat presentations and appropriate management needs to be maintained. The hypothesis has arisen in Aboriginal settings that GAS-associated skin infection may be a precursor for ARF in communities,³⁵ suggesting that treatment of skin infection could offer another primary prevention strategy, but the evidence is lacking. <p>Refer to endorsed therapeutic guidelines - Antibiotic</p>	<p>Management of streptococcal pharyngitis:</p> <ul style="list-style-type: none"> ● Treat with intramuscular BPG 900mg (450mg if <20kg) single dose or oral phenoxymethylpenicillin 500mg twice daily (250 mg twice if <34kg) for 10 days¹ ● Exclude from school* until the person has received antibiotic treatment for ≥24 hours and feels well²⁴ ● Encourage cough and sneeze etiquette and hand hygiene (for details see³⁷) <p>Management of streptococcal impetigo in high-risk communities:</p>

Level of prevention	Background	Recommendations																															
		<ul style="list-style-type: none"> • Treat with intramuscular BPG single dose or trimethoprim-sulphathiazole (see Therapeutic Guidelines for dosing regimen options)³⁸ • Exclude from school* until appropriate antibiotic treatment has started ³⁷ • Cover sores on exposed skin with watertight dressing, dispose of contaminated dressing hygienically and encourage hand hygiene (for details see ³⁷ <p>Encourage regular handwashing to reduce risk of transmission of GAS</p>																															
Secondary	<p>This is the most studied and effective preventive measure, with level 1a evidence</p> <ul style="list-style-type: none"> • Secondary prophylaxis with intramuscular injections of benzathine penicillin G every 28 days significantly reduces ARF recurrence rates compared with placebo, ³⁹ or oral penicillin ³⁰ and is the treatment of choice. This strategy forms the basis of the WHO's ARF recommendations⁴¹ and the Australian's National Guidelines.¹ 	<table border="1"> <thead> <tr> <th data-bbox="1328 671 1637 730">Agent</th> <th data-bbox="1637 671 1809 730">Dose</th> <th data-bbox="1809 671 1912 730">Route</th> <th data-bbox="1912 671 2051 730">Frequency</th> </tr> </thead> <tbody> <tr> <td colspan="4" data-bbox="1328 730 2051 767">First line</td> </tr> <tr> <td data-bbox="1328 767 1637 1082">Benzathine penicillin G (BR LAB/Bicillin)</td> <td data-bbox="1637 767 1809 1082">900mg (≥20kgs) =1.200.000 units 450mg (<20kgs) =600.000 units</td> <td data-bbox="1809 767 1912 1082">IM</td> <td data-bbox="1912 767 2051 1082">Every 21-28 days§</td> </tr> <tr> <td colspan="4" data-bbox="1328 1082 2051 1137">Second line</td> </tr> <tr> <td data-bbox="1328 1137 1637 1233">Phenoxyethylpenicillin (Penicillin V)</td> <td data-bbox="1637 1137 1809 1233">250 mg</td> <td data-bbox="1809 1137 1912 1233">oral</td> <td data-bbox="1912 1137 2051 1233">Twice daily</td> </tr> <tr> <td colspan="4" data-bbox="1328 1233 2051 1289">Following documented</td> </tr> <tr> <td data-bbox="1328 1289 1637 1380">Penicillin-allergy Erythromycin</td> <td data-bbox="1637 1289 1809 1380">250 mg</td> <td data-bbox="1809 1289 1912 1380">oral</td> <td data-bbox="1912 1289 2051 1380">Twice daily</td> </tr> </tbody> </table>				Agent	Dose	Route	Frequency	First line				Benzathine penicillin G (BR LAB/Bicillin)	900mg (≥20kgs) =1.200.000 units 450mg (<20kgs) =600.000 units	IM	Every 21-28 days§	Second line				Phenoxyethylpenicillin (Penicillin V)	250 mg	oral	Twice daily	Following documented				Penicillin-allergy Erythromycin	250 mg	oral	Twice daily
Agent	Dose	Route	Frequency																														
First line																																	
Benzathine penicillin G (BR LAB/Bicillin)	900mg (≥20kgs) =1.200.000 units 450mg (<20kgs) =600.000 units	IM	Every 21-28 days§																														
Second line																																	
Phenoxyethylpenicillin (Penicillin V)	250 mg	oral	Twice daily																														
Following documented																																	
Penicillin-allergy Erythromycin	250 mg	oral	Twice daily																														

Level of prevention	Background	Recommendations
	Duration of secondary prophylaxis: Minimum 10 years after most recent episode of ARF or until age 21 years (whichever is longer), then re-evaluate. If echocardiogram then show no or mild RHD, cease secondary prophylaxis. If echo shows moderate RHD, continue until age 35. RHD or valve surgery has been performed, continue until age 40, or longer. ¹	
Tertiary	This comprises medical and surgical management of RHD which is outside the scope of this document.	

‡ BPG = benzathine benzylpenicillin

IMI – intramuscular

*Recommendation about school exclusion need to be considered carefully in communities characterised by low school attendance. Exclusion should also incorporate avoiding contact with at-risk individuals e.g., people with prior § or every 21 days for selected individuals (uncommon)

§§ only in exceptional circumstances less effective than 1st line ** Based on the Canadian National Occupancy Standard which provides definitions of household crowding based on age and sex of occupants. In general, they consider >2 people per bedroom to represent crowding⁴²

4. Surveillance objectives

To identify cases of ARF and RHD in a timely manner:

- To notify cases to jurisdictional public health units in accordance with [Table 4](#) for notification purposes to monitor trends, detect outbreaks and potentially trigger contact tracing/screening.
- To report cases to jurisdictional disease registers in accordance with [Table 4](#) for oversight of delivery of ARF and RHD care coordination, in particular commencement and maintenance of secondary prophylaxis, and timeliness of echocardiographic, medical and dental reviews.

Data management

Depending on State/Territory requirements (see [Table 4](#)), cases should be notified to the jurisdictional notifiable diseases database and/or the RHD disease register (control program), in a timely fashion.

Reporting requirements

ARF is notifiable in QLD, NT, WA (2015), NSW (2015) and SA (2016). Possible cases are not notifiable.

RHD is notifiable in NSW (2015), WA (2015) and SA (2016).

ARF/RHD control programs

Jurisdictional ARF/RHD control programs exist in QLD, NT, SA, NSW and WA. Each of these jurisdictions maintains a register of ARF/RHD cases.

Table 4 Australian notification requirements *

	Notifiable to jurisdictional Health Department or Public Health Unit					Included in jurisdictional register				
	QLD	NT	WA	SA	NSW	QLD	NT	WA	SA	NSW
Definite ARF	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Probable (highly suspected) ARF	No	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
RHD	No	No	Yes	Yes	Yes<35 yrs	Yes	Yes	Yes	Yes	Yes

* Nil in Vic/ACT or Tas

5. Communications

Communicable Disease Directors (or their nominated delegate) should be notified if an ARF outbreak is suspected (see Section 12).

6. Case definitions

Case definition	
ARF Definite (confirmed) case	<p>A confirmed case requires clinical evidence AND laboratory suggestive evidence.^c</p> <p>Clinical evidence</p> <p>For initial episode of ARF</p> <ol style="list-style-type: none"> 1. Two major manifestations <p>OR</p> <ol style="list-style-type: none"> 2. One major and two minor manifestations. <p>OR</p> <ol style="list-style-type: none"> 3. Rheumatic Sydenham's chorea alone^c AND other forms of chorea excluded. <p>For recurrent episode in a patient with known past ARF or RHD</p> <ol style="list-style-type: none"> 1. Two major manifestations <p>OR</p> <ol style="list-style-type: none"> 2. One major and one minor manifestation <p>OR</p> <ol style="list-style-type: none"> 3. Three minor manifestations <p>OR</p> <ol style="list-style-type: none"> 4. Rheumatic Sydenham's chorea alone^c AND other forms of chorea excluded <p>Laboratory suggestive evidence</p> <ol style="list-style-type: none"> 5. Elevated or rising antistreptolysin-O or anti-DNase B or other streptococcal antibody <p>OR</p> <ol style="list-style-type: none"> 6. Positive group A streptococcal (GAS) throat culture <p>OR</p> <ol style="list-style-type: none"> 7. Positive rapid antigen test for group A streptococci.
ARF Probable case (first episode or recurrence)	<ol style="list-style-type: none"> 1. A clinical presentation that falls short by either one major or one minor manifestation <p>OR</p> <ol style="list-style-type: none"> 2. Clinical evidence (as above) without laboratory suggestive evidence AND <p>where ARF is considered the most likely diagnosis by the treating physician.</p>
ARF Possible case	As for probable case, where the treating clinician has less confidence about ARF as the correct diagnosis, but other differential diagnoses have been excluded.
RHD	A diagnosis of RHD is made by a cardiologist based on echocardiographic findings. RHD is defined as the presence of specific echocardiographic

^c Rheumatic Sydenham's chorea may occur alone without other manifestations or laboratory suggestive evidence, provided other causes of chorea are ruled out. Therefore Sydenham's chorea alone, without laboratory suggestive evidence, is sufficient evidence for a confirmed case.

Case definition

Definite and probable	features summarised in Appendix 1. We provide the definition as an appendix. Refer to Appendix 3: World Heart Federation Criteria for the Echocardiographic Diagnosis of RHD in Individuals Aged ≤20 Years.
------------------------------	---

Example: A high-risk child with no prior ARF/RHD with monoarthralgia, fever, P-R prolongation (all minor criteria) and positive streptococcal serology where other causes have been excluded, may be deemed a probable case. A high-risk child with monoarthralgia, fever and elevated CRP only, where other causes have been excluded, may be deemed to be a possible case. The decision is at the discretion of the ARF specialist consulted, based on history, examination findings, history etc.

7. Laboratory testing

There is no diagnostic test for ARF. Required laboratory testing includes:

- Tests to exclude differential diagnoses
- Full blood count, C-reactive protein (CRP) and/or erythrocyte sedimentation rate (ESR)
- Supporting evidence of preceding Group A streptococcal infection as per Table 5

Numerous laboratory methods for testing ASOT exist, and results can vary significantly between test types. Laboratories may establish locally appropriate reference ranges for a given ASOT test type and for their population, but if such data are unavailable, the cut-offs provided below are recommended, with advice to practitioners to be mindful of variability between test types. A rise in titre between paired acute and 14-28-day convalescent serum samples may help but also has limitations.⁴

Table 5: Evidence of antecedent group A streptococcal infection

Sample	Description	Test characteristics	Upper limits of normal	
			Age	UKN U/mL
Blood	Antistreptolysin O titre (ASOT)	<ul style="list-style-type: none"> • Positive in 75-80% of pharyngeal streptococcal infections • Typically follows throat streptococcal infection • Can be positive after group A, C or G streptococcal infection⁴³ • Peak in titre ~3-6 weeks post infection • Return to normal range may take 6-12 months 	1-4 5-14 15-24 25-34 ≥35	170 276 238 177 127
Blood	AntiDNaseB titre	<ul style="list-style-type: none"> • Typically follows throat and skin streptococcal infection • More specific than ASOT for Group A streptococcal infection 	1-4 5-14 15-24 25-34	366 499 473 390 265

Sample	Description	Test characteristics	Upper limits of normal	
			Age	UKN U/mL)
		<ul style="list-style-type: none"> Peak in titre ~6-8 weeks post infection Return to normal range may take 6-12 months 	>=35	
Pharyngeal swab (bacterial in transport medium)	Group A-haemolytic streptococcus cultured from pharyngeal swab	Often culture-negative by the time symptomatic ARF is evident; however, a positive result provides the required proof to make the ARF diagnosis even if serological titres for ASOT/ anti-DNaseB are below cut-off.		

8. Case management

Response times

People with suspected ARF should be admitted to hospital within 24 hours or as soon as feasible for people living in remote communities. Admission to hospital is advised to:

- help facilitate the diagnosis including undertaking an echocardiogram
- ensure that adequate information and support is provided to patient and family, given the serious nature of the diagnosis, and need for long-term adherence to treatment and high-level engagement with health services

Response procedure

Case investigation

Examples of an ARF Case notification form and an Enhanced Surveillance form for person diagnosed with ARF is provided in Appendices 1 and 2.

Case treatment

Cases should be managed by a doctor with expertise in the management of ARF/RHD. Treating doctors seeking specialist advice can be referred through the jurisdictional RHD control programs to an ARF/RHD specialist. Management is guided by the Australian National Guidelines which provide care plans based on the individuals priority status (P1, P2, P3).¹ Some jurisdictional control programs provide education and adherence support directly to patients and their families; elsewhere this responsibility lies with the primary care team.

Admission to hospital for further investigation and follow-up is highly recommended.

Important components of case management include:

1. Appropriate specialist treatment of the arthritis, carditis and/or chorea¹
 - e.g. appropriate dosing and duration of salicylate therapy for arthritis¹
2. Education for the patient, family and carers – see below
3. Commence secondary antibiotic prophylaxis preferably with BPG injections every 21- 28 days
4. Ensure pain minimisation techniques are used for delivery of BPG injections¹

5. Acknowledge the special needs of adolescents newly diagnosed with a chronic condition
 - Although individual ARF episodes are acute, the management is akin to chronic disease management with a requirement for regular medication and regular medical care to prevent the long-term complication of rheumatic heart disease
6. Appropriate discharge * planning that includes:
 - communication with primary health care team
 - setting up chronic disease care plan/recalls

* Discharge should be delayed until resolution of carditis, and inflammatory markers as early discharge has led to fatal decompensation in the community setting. Resolution of these markers should be determined and managed by an ARF/ RHD specialist clinician

Education

Ensure the patient, their family and primary carers have access to culture and language-appropriate resources, and have been provided with culturally appropriate face to face educational sessions, using an interpreter where required to cover areas including: diagnosis and cause, increased potential risk to family members of ARF, long-term medical management and pharmacological and non-pharmacological secondary prophylaxis options (see 'Additional sources of information' for web addresses for patient resources.)

Simple explanations about the non-pharmacological measures associated with reduced risk of ARF episodes should be provided. While randomised trial evidence is lacking, observational studies have repeatedly indicated that household overcrowding correlates with ARF incidence,^{25, 26} and therefore strategies to mitigate this need to be developed with families. The definition of crowding used by the Australian Institute of Health and Welfare (no more than 2 people per bedroom⁴²) may appear excessively cautious and unfeasible, but nevertheless provides a target towards which to work. Randomised trials of the effect of washing children's hands and bodies with soap and water have demonstrated a significant decrease in impetigo rates in the groups assigned to washing.^{44, 45} While a link between pyoderma and ARF has never definitively been established, washing with soap and water is an appropriate primordial preventive message to promote for families and communities affected by ARF.

Isolation and restriction

Not required

Active case finding

Not required

9. Environmental evaluation

An Environmental health assessment may be a useful adjuvant to treatment, to address social/environmental issues where this is appropriate and reasonable to do so. The capacity to conduct environmental evaluation varies by region. The appropriate response depends on the geographical setting and the nature of the affected person's housing.

To assist in advising about strategies to avoid future ARF recurrences, the living conditions experienced by the patient could be assessed by an Environmental Health Officer/Unit and in conjunction with the relevant public housing government department or housing provider/funding agency for private housing. Consent should be provided by the family affected and where possible a community member should be present with the family prior to assessment. The following describes a suggested response for remote-dwelling Indigenous Australians living as tenants in public or private housing.

Issues to assess include:

- Number of people in the house / per bedroom / per bed (see above and reference⁴² for acceptable numbers of bedroom occupants)
- All functioning health hardware which enables people to wash adequately, especially children, the ability to wash clothing and bedding
 - Access to hot and cold running water
 - Access to a functional toilet
 - Access to a hot water washing machine
 - Access to appropriate community hygiene/human-waste removal (safe rubbish removal and disposal, sewage)

Deficiencies in the health hardware which are beyond the capacity of the patient's family to manage should be referred to the jurisdictional government housing authority if the patient lives in public housing or the housing provider if the patient lives in private housing.

10. Contact management

Not routine. Contact management in potential outbreak scenarios is addressed in Section 11 – Special situations. The secondary attack rate for sporadic ARF is well below the threshold required for instigation of contact treatment (e.g. see reference regarding transmission rates³). ARF occurs in only a small proportion of individuals infected with GAS, perhaps as few as 3-5%.

Identification of contacts

Not required.

Contact definition

Not applicable.

Management of identified contacts

Not applicable.

Education

Contacts living with the case person should be included in education about ARF/RHD, in particular the association of GAS transmission with over-crowding and the importance of skin health to reduce GAS transmission for all those living in a household with a person with ARF/RHD. Additionally, for family contacts, the possibility of genetic susceptibility can be raised.

Isolation and restriction

Not required.

11. Special situations

ARF possible outbreak/clustering response

ARF has been considered to be a sporadic condition in Australia. However, in recent years, a small number of situations consistent with possible outbreaks have been observed in parts of northern Australia with already underlying high rates of ARF. This section therefore provides guidance on management of such possible ARF outbreaks, with the caveats that:

- There is a current lack of evidence to provide clear guidance on ARF outbreak response strategies
- The appropriate definition of an ARF outbreak (specifically, number of cases or type of clustering) is unclear and may evolve over time, or different definitions may be selected in different States/Territories
- The time lag between GAS exposure and onset of ARF means that the opportunities for effective outbreak responses are limited
- Where research laboratory capacity exists to undertake Streptococcal *emm*-typing of any GAS isolates obtained from pharyngeal or skin swabs, this would be a valuable adjunct to identify whether dominant rheumatogenic GAS strain(s) is/are circulating. This research aspect may help distinguish between an increase in endemic ARF and a true outbreak due to a newly introduced “rheumatogenic” GAS and add to the limited evidence in this area; it is not an essential component of the public health response.

Components of outbreak response

1. Case definition for ARF
 - a. As per definition provided above, [Section 7](#) Laboratory testing
2. Outbreak definition
 - a. A greater than expected number of confirmed or probable ARF cases occurring during an approximately 4-week period within a defined region. The threshold number of cases and timing needs to be defined by Public Health Units at regional levels and will vary by background rate of ARF and community population size. For example, two cases in a community, especially if related (e.g. in same family, household or class) may be enough to trigger scrutiny
3. Provision of information to community
 - a. Be on the alert for cases of ARF/RHD
 - b. Be on the alert for known or suspected antecedents of ARF, i.e. pharyngitis or skin sores. Develop a ‘zero tolerance’ approach to these conditions – i.e. ensure all cases are managed appropriately, and education about preventing spread is provided to affected households
 - c. Ensure staff, especially new or locum staff, know about ARF/RHD (e.g. have undertaken the online training modules – see Resources)
 - d. Ensure staff have access to the range of resources available to assist clinicians, individuals, families and communities regarding ARF/RHD, including the [public service announcements](#) available in English and Aboriginal languages (www.takeheart.tv/films/important-health-message/)
4. Control and prevention of further disease
 - a. The highest priority is to ensure all known people with ARF/RHD currently receiving penicillin prophylaxis in the affected location are up to date with their benzathine penicillin G injections as they are most at risk
 - b. Contact identification and management. The extent of contact tracing and who will be treated will vary by scenario and should be decided in consultation between the community primary care staff, regional Public Health Unit, relevant hospital specialists (paediatricians, infectious disease physicians) and the regional ARF/RHD Control Program.
 - i. Identify contacts: family, household and close contacts, defined as those staying in the same house as a case in the 4 weeks preceding onset of the ARF in the index case(s)
 - ii. Examine contacts for skin sores or pharyngitis/tonsillitis and provide appropriate management for these conditions (including covering skin sores see Table 3 above)

- iii. Treatment of all contacts aged >12 months and <50 years not already treated with anti-GAS antibiotic. If prior treatment was with an oral antibiotic more than 7 days prior, re-treat. If prior treatment was with benzathine penicillin G more than 21 days ago, re-treat. The purpose of treating contacts is to prevent further GAS transmission by removing potential sources of GAS infection and to prevent sequelae of GAS infection such as ARF in those recently infected.
5. Active case finding
 - a. Examine at-risk contacts (e.g. aged <21 years, depending on resources) for features of ARF. Cardiac auscultation by an experienced clinician but where possible use of a portable echocardiogram will provide best sensitivity for diagnosis⁴⁶
 6. Collect samples for bacterial culture +/- typing of Group A Streptococcus from all cases (already routine practice), but additionally from all contacts; ideally both throat swabs and swabs of any skin sores should be collected in this setting.
 7. Provide community education about ARF and ARF prevention strategies

Table 6: Dosage table for benzathine penicillin G for treatment of ARF contacts†‡*

Weight	Dose of benzathine penicillin G	Volume‡
3kg to <6kg	225mg	0.6mL
6 to <10kg	337.5mg	0.86mL
10 to <15 kg	450mg	1.15mL
15 to <20 kg	675mg	1.73mL
20 kg or more	900mg	2.3mL

† Note that doses differ from doses required for ARF secondary prophylaxis, since the latter requires a large depot dose with longer half-life for prevention of future exposures, whereas the strategy for contact treatment is to eradicate any current GAS infection or carriage.

‡ Note that doses in mL differ from the CARPA manual as the 2015 preparation of Bicillin LA comes as 900mg in 2.3mL. This may well change again, so check current formulation being used.

*Those allergic to penicillin should receive azithromycin 12mg/kg up to 500mg orally, daily for 5 days.

12. References and additional sources of information

Resources

[ARF fact sheet](http://www.rhdaustralia.org.au/resources/10-things-you-should-know-about-rheumatic-fever): (www.rhdaustralia.org.au/resources/10-things-you-should-know-about-rheumatic-fever)

Control Program Contact numbers for provision of patient education and clinician advice:

- NT Top End 08 89228454
- NT Central 08 89516909
- Queensland 1300135854
- Western Australia 1300622745
- South Australia 08 74257146
- New South Wales 1300 066 055
- Tasmania 1800 671 738 (No Control program contact Communicable Disease Prevention Unit for suspected cases)

[Diagnosis calculator](http://www.rhdaustralia.org.au/apps) App: www.rhdaustralia.org.au/apps

[Treatment tracker](http://www.rhdaustralia.org.au/treatment-tracker-app) App (staying on track with penicillin): www.rhdaustralia.org.au/treatment-tracker-app

RHD in [Pregnancy resources](http://www.rhdaustralia.org.au/pregnant): www.rhdaustralia.org.au/pregnant

Educational resources for patients:

- [Videos](http://www.rhdaustralia.org.au/resources/search/advanced): www.rhdaustralia.org.au/resources/search/advanced
- [Written materials](http://www.rhdaustralia.org.au/resources/what-rheumatic-fever): www.rhdaustralia.org.au/resources/what-rheumatic-fever
- [Educational resources](http://www.rhdaustralia.org.au/e-learning-discussion-forum) for staff: www.rhdaustralia.org.au/e-learning-discussion-forum

References

1. RHD Australia (ARF/RHD writing group). The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition). National Heart Foundation of Australia and the Cardiac Society of Australia and New Zealand. [www.rhdaustralia.org.au/arf-rhd-guideline (www.rhdaustralia.org.au/sites/default/files/guideline_0.pdf). 2012; (October 2012)].
2. Gewitz MH, Baltimore RS, Tani LY, et al. Revision of the Jones Criteria for the diagnosis of acute rheumatic fever in the era of Doppler echocardiography: a scientific statement from the American Heart Association. *Circulation* 2015; 131(20): 1806-18.
3. Health Protection Agency Group A Streptococcus Working Group. Interim UK guidelines for management of close community contacts of invasive group A streptococcal disease. *Communicable disease and public health / PHLS* 2004; 7(4): 354-61.
4. Haidan A, Talay SR, Rohde M, Sriprakash KS, Currie BJ and Chhatwal GS. Pharyngeal carriage of group C and group G streptococci and acute rheumatic fever in an Aboriginal population. *Lancet* 2000; 356(9236): 1167-9.
5. McDonald M, Towers RJ, Andrews RM, Carapetis JR and Currie BJ. Epidemiology of *Streptococcus dysgalactiae* subsp. *equisimilis* in tropical communities, Northern Australia. *Emerg Infect Dis* 2007; 13(11): 1694-700.
6. Carapetis JR and Currie BJ. Group A streptococcus, pyoderma, and rheumatic fever. *Lancet* 1996; 347(9010): 1271-2.
7. McDonald M, Brown A, Edwards T, et al. Apparent contrasting rates of pharyngitis and pyoderma in regions where rheumatic heart disease is highly prevalent. *Heart, lung & circulation* 2007; 16(4): 254-9.
8. McDonald MI, Towers RJ, Andrews RM, Bengner N, Currie BJ and Carapetis JR. Low rates of streptococcal pharyngitis and high rates of pyoderma in Australian aboriginal communities where acute rheumatic fever is hyperendemic. *Clin Infect Dis* 2006; 43(6): 683-9.
9. Bryant PA, Robins-Browne R, Carapetis JR and Curtis N. Some of the people, some of the time: susceptibility to acute rheumatic fever. *Circulation* 2009; 119(5): 742-53.
10. de Dassel JL, Ralph AP and Carapetis JR. Controlling acute rheumatic fever and rheumatic heart disease in developing countries: are we getting closer? *Current opinion in pediatrics* 2015; 27(1): 116-23.
11. Ralph AP and Carapetis JR. Group A streptococcal diseases and their global burden. *Current topics in microbiology and immunology* 2013; 368: 1-27.
12. Lennon D, Stewart J, Farrell E, Palmer A and Mason H. School-based prevention of acute rheumatic fever: a group randomized trial in New Zealand. *Pediatr Infect Dis J* 2009; 28(9): 787-94.
13. Remenyi B, Wilson N, Steer A, et al. World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease--an evidence-based guideline. *Nature reviews Cardiology* 2012; 9(5): 297-309.
14. Carapetis J and Currie, BJ. Rheumatic fever in a high incidence population: The importance of mono-arthritis and low grade fever. *Arch Dis Child* 2001; 85: 223-37.
15. Carapetis JR and Currie BJ. Rheumatic chorea in northern Australia: a clinical and epidemiological study. *Archives of disease in childhood* 1999; 80(4): 353-8.
16. Lawrence JG, Carapetis JR, Griffiths K, Edwards K and Condon JR. Acute Rheumatic Fever and Rheumatic Heart Disease: Incidence and Progression in the Northern Territory of Australia 1997-2010. *Circulation* 2013.
17. Watkins DA, Sebitloane M, Engel ME and Mayosi BM. The burden of antenatal heart disease in South Africa: a systematic review. *BMC cardiovascular disorders* 2012; 12: 23.

18. Olivier C. Rheumatic fever - is it still a problem? . *J Antimicrob Chemother* 2000; 45: 13-21.
19. Australian Institute of Health and Welfare. Cardiovascular disease: Australian facts. Cardiovascular disease series. Cat. no. CVD 53. Canberra: AIHW. (www.aihw.gov.au/WorkArea/DownloadAsset.aspx?id=10737418530), 2011.
20. Noonan S, Zurynski YA, Currie BJ, et al. A National Prospective Surveillance Study of Acute Rheumatic Fever in Australian Children. *Pediatr Infect Dis J* 2012.
21. Brennan RE and Patel MS. Acute rheumatic fever and rheumatic heart disease in a rural central Australian aboriginal community. *Med J Aust* 1990; 153(6): 335, 8-9.
22. Richmond P and Harris L. Rheumatic fever in the Kimberley region of Western Australia. *Journal of tropical pediatrics* 1998; 44(3): 148-52.
23. Hanna JN and Clark MF. Acute rheumatic fever in Indigenous people in North Queensland: some good news at last? *Med J Aust* 2010; 192(10): 581-4.
24. Chidzey PJ. Applied epidemiology in the Kimberley: acute rheumatic fever increase in Western Australia [master's thesis]. Canberra: Australian National University; 2015
25. Kerdemelidis M, Lennon DR, Arroll B, Peat B and Jarman J. The primary prevention of rheumatic fever. *J Paediatr Child Health* 2010; 46(9): 534-48.
26. Jaine R, Baker M and Venugopal K. Acute rheumatic fever associated with household crowding in a developed country. *Pediatr Infect Dis J* 2011; 30(4): 315-9.
27. Wannamaker LW. The epidemiology of streptococcal infections. New York, NY: Columbia University Press; 1954.
28. Adanja B, Vlajinac H and Jarebinski M. Socioeconomic factors in the etiology of rheumatic fever. *J Hyg Epidemiol Microbiol Immunol* 1988; 32(3): 329-35.
29. Okello E, Kakande B, Sebatta E, et al. Socioeconomic and environmental risk factors among rheumatic heart disease patients in Uganda. *PLoS One* 2012; 7(8): e43917.
30. Zaman MM, Yoshiike N, Chowdhury AH, et al. Socio-economic deprivation associated with acute rheumatic fever. A hospital-based case-control study in Bangladesh. *Paediatr Perinat Epidemiol* 1997; 11(3): 322-32.
31. Lenhart MK. *Recruit Medicine*. USA: Office of the Surgeon General, Department of the Army; 2006.
32. Perry W, Siegel A, Rammelkamp C and Wannamaker L. Transmission of Group A Streptococci I. The Role of Contaminated Bedding. *American Journal of Hygiene* 1957; 66: 85-95.
33. Falck G, Kjellander J and Schwan A. Recurrence rate of streptococcal pharyngitis related to hygienic measures. *Scand J Prim Health Care* 1998; 16(1): 8-12.
34. Aboriginal Environmental Health Unit. Closing the gap: 10 years of Housing for Health in NSW An evaluation of a healthy housing intervention. Sydney: NSW Health, 2010.
35. Lennon D and Stewart J. An important investment to control Acute Rheumatic Fever needs to run its course. *The New Zealand medical journal* 2015; 128(1416): 6-9.
36. Irlam J, Mayosi BM, Engel M and Gaziano TA. Primary prevention of acute rheumatic fever and rheumatic heart disease with penicillin in South African children with pharyngitis: a cost-effectiveness analysis. *Circulation Cardiovascular quality and outcomes* 2013; 6(3): 343-51.
37. McDonald M, Currie BJ and Carapetis JR. Acute rheumatic fever: a chink in the chain that links the heart to the throat? *The Lancet infectious diseases* 2004; 4(4): 240-5.
38. Australian Government National Health and Medical Research Council. Staying Healthy - Preventing infectious diseases in early childhood education and care services FIFTH EDITION (Updated June 2013)

(www.nhmrc.gov.au/_files_nhmrc/publications/attachments/ch55_staying_healthy_5th_edition_150602.pdf), 2012.

39. Antibiotic Expert Groups. Therapeutic Guidelines: Antibiotic. Version 15. Melbourne, Australia: Therapeutic Guidelines Limited; 2014.
40. Padmavati S, Sharma KB and Jayaram O. Epidemiology and prophylaxis of rheumatic fever in Delhi--a five year follow-up. Singapore medical journal 1973; 14(3): 457-61.
41. Manyemba J and Mayosi BM. Penicillin for secondary prevention of rheumatic fever. Cochrane database of systematic reviews 2002; (3): CD002227.
42. WHO Expert Consultation on Rheumatic Fever and Rheumatic Heart Disease. Rheumatic fever and rheumatic heart disease: report of a WHO Expert Consultation, Geneva, 29 October - 1 November 2001. WHO Technical Report Series 923. 2004. whqlibdoc.who.int/trs/WHO_TRS_923.pdf (accessed October 2012).
43. Australian Institute of Health and Welfare. Canadian National Occupancy Standard. 2012. (www.meteor.aihw.gov.au/content/index.phtml/itemId/386254) (accessed 23/10/2015).
44. Johnson DR, Kurlan R, Leckman J and Kaplan EL. The human immune response to streptococcal extracellular antigens: clinical, diagnostic, and potential pathogenetic implications. Clin Infect Dis 2010; 50(4): 481-90.
45. Luby S, Agboatwalla M, Schnell BM, Hoekstra RM, Rahbar MH and Keswick BH. The effect of antibacterial soap on impetigo incidence, Karachi, Pakistan. Am J Trop Med Hyg 2002; 67(4): 430-5.
46. Luby SP, Agboatwalla M, Feikin DR, et al. Effect of handwashing on child health: a randomised controlled trial. Lancet 2005; 366(9481): 225-33.

Appendices

Appendix 1: Sample Acute Rheumatic Fever Notification Form for jurisdictions where ARF is notifiable and an ARF/RHD register exists

“ARF is a notifiable condition. Report all confirmed and suspected cases to your nearest Public Health Unit, who will inform the ARF/RHD Register & Control Program”

Patient details

Hospital/Clinic UR no _____

Surname _____

Given names _____

Date of birth ____/____/____

Sex _____

Address: _____

Suburb/Town _____

Postcode _____

Phone number(s): _____

Ethnicity

Aboriginal

Torres Strait Islander

Aboriginal **and** Torres Strait Islander

Pacific Islander - Maori

Pacific Islander - Other

Other. Please state _____

Not stated

Country of birth _____

Notification Date ____/____/____

Notifying clinician

Clinician's name _____

Clinician's signature _____

Hospital/Clinic name _____

Contact email and phone _____

Current ARF episode This episode is:

Select 1: Initial ARF /Recurrent ARF/ Unknown

Select 1: Confirmed / Probable/ Possible

ARF symptom onset date: ____/____/____

RHD identified

YES / NO / Echo not performed

If YES: date RHD identified: ____/____/____

Severity: Priority 1 (severe) / Priority 2 (moderate) / Priority 3 (mild)

Date Bicillin given: ____/____/____

Hospitalisation: YES / NO

If YES: date ____/____ Hospital _____

ARF diagnostic process

Differs depending on risk level. Groups recognised as high risk or potentially high risk include Aboriginal and Torres Strait Islander Australians, Maori and Pacific Islander people, and migrants from developing countries.

MAJOR manifestations (tick all that apply)

Poly-arthralgia (high risk groups)

Aseptic mono-arthritis (high risk groups)

Poly-arthritis

Carditis

Sub-clinical carditis (high risk groups)

Erythema marginatum

Sydenham's chorea

Subcutaneous nodules

MINOR manifestations (tick all that apply)

Poly-arthralgia (low risk groups)

Aseptic mono-arthritis (low risk groups)

Mono-arthralgia (high risk groups)

Fever (≥ 38 °C): ____ °C

Prolonged P-R interval on ECG

ESR (≥ 30 mm/hr): ____ Date ____/____/____

CRP (≥ 30 mg/L): ____ Date ____/____/____

Evidence of preceding Group A Strep (GAS) infections 3

Elevated ASOT: Yes / No

result ____ IU/mL Date ____/____/____

Elevated Anti-DNaseB: Yes / No

result ____ IU/mL Date ____/____/____

Positive throat swab culture: Yes / No

Positive skin swab culture: Yes / No

History of URTI / Strep throat: Yes / No

Modified JONES CRITERIA for ARF diagnosis.

Initial episode of ARF

2 Major manifestations

or

1 Major and 2 Minor manifestations

plus evidence of a preceding GAS infection

Recurrent episode:

2 Major manifestations

or

1 Major and 1 Minor manifestations

or

3 Minor Manifestations

plus evidence of a preceding GAS infection

Sydenham's chorea alone is enough to confirm ARF

Appendix 2: Example of an Enhanced Surveillance form for person diagnosed with ARF

1. Date completed: Click or tap to enter a date. Name of person completing form:
: Contact email
2. Case Code: 3. Date of Birth: Click or tap to enter a date.
4. Sex: M F 5. Post code: 6. Country of birth:
7. Place of birth: 8. Usual place of residence: Capital city/Large town/Small town /Remote area
9. Ethnicity: Aboriginal and/or Torres Strait Islander /Caucasian /Asian /Pacific Islander/Middle Eastern /African/Other
10. Number of other children in the family: 0 1 2 3 4 5 >5 unknown
11. How many people usually sleep in the dwelling (approx)? 1-4 5-9 >9 unknown
12. How many of these people are children aged <15yrs 1-4 5-9 >9 unknown
13. Do other family members/residents in same dwelling have Hx of ARF or RHD? Yes No unknown
14. Date of diagnosis for current ARF episode: Click or tap to enter a date.
15. Date of symptom onset for current ARF episode: Click or tap to enter a date.
16. Has this child been previously diagnosed with ARF? Yes No unknown
- Months/years for previous diagnoses: Click or tap to enter a date.
17. Please select all diagnostic criteria present in the current episode of ARF:
- MAJOR criteria: Carditis Polyarthritis Sydenham's Chorea
Erythema marginatum Subcutaneous nodules
Polyarthralgia (high risk groups) Aseptic mono-arthritis (high risk groups)
- MINOR criteria: Fever (max temp__ oC) ESR ≥30mm/hr (highest ES_mm/hr)
Prolonged PR interval CRP ≥30mg/L (highest
CRP_____mg/L)
Aseptic mono-arthritis (low risk groups) Polyarthralgia (low risk groups)
18. Diagnosis Definite ARF
 Probable (highly suspected) ARF
 Possible (uncertain) ARF
19. Were any cardiac valve lesions present? Yes No unknown
- If yes: (tick all that apply)
- 19a. Mitral valve regurgitation: Severity: None Mild Moderate Severe unknown
- 19b. Mitral stenosis Severity: None Mild Moderate Severe unknown
- 19c. Aortic valve regurgitation Severity: None Mild Moderate Severe unknown
- 19d. Aortic stenosis Severity: None Mild Moderate Severe unknown
- 19e. Tricuspid valve lesion (specify) unknown
- 19f. Pulmonary valve lesion (specify) unknown
20. Evidence of valve lesions is based on Echocardiogram Clinical Assessment

21. Was there a sore throat within 3 weeks of ARF symptoms? Yes No unknown

22. Was there evidence of skin sores within 3 weeks of ARF? Yes No unknown

23. Was there evidence of Group A streptococcal (GAS) infection? Yes No unknown

If yes, please provide the following details:

Culture: Yes No unknown If yes, identify site: throat / skin / other

M type if GAS isolate typed

ASOT titre Result Date Click or tap to enter a date.

Anti DNase titre Result Date Click or tap to enter a date.

Appendix 3: World Heart Federation Criteria for the Echocardiographic Diagnosis of RHD in Individuals Aged ≤20 Years

Echocardiographic Criteria for RHD*	
Definite RHD (A, B, C, or D)	
<ul style="list-style-type: none"> A. Pathologic MR and at least 2 morphologic features of RHD of the MV B. MS mean gradient ≥4 mm Hg C. Pathological AR and at least 2 morphological features of RHD of the AV D. Borderline disease of both the AV and MV 	
Borderline RHD (A, B, or C)	
<ul style="list-style-type: none"> A. At least 2 morphologic features of RHD of the MV without pathologic MR or MS B. Pathologic MR C. Pathologic AR 	
Echocardiographic criteria for pathologic regurgitation (all 4 Doppler criteria must be met)	
Pathologic MR	Pathologic AR
1. Seen in 2 views	1. Seen in 2 views
2. In at least 1 view jet length ≥2 cm†	2. In at least 1 view jet length ≥1 cm†
3. Peak velocity ≥3 m/s for 1 complete envelope	3. Peak velocity ≥3 m/s in early diastole
4. Pansystolic jet in at least 1 envelope	4. Pandiastolic jet in at least 1 envelope
Morphologic features of RHD	
Features in the MV	Features in the AV
1. AMVL thickening ≥3 mm‡	1. Irregular or focal thickening
2. Chordal thickening	2. Coaptation defect
3. Restricted leaflet motion	3. Restricted leaflet motion
4. Excessive leaflet tip motion during systole	4. Prolapse

Abbreviations: AMVL, anterior mitral valve leaflet; AR, aortic regurgitation; AV, aortic valve; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve; and RHD indicates rheumatic heart disease.

* Congenital anomalies must be excluded.

† A regurgitant jet length should be measured from the vena contracta to the last pixel of regurgitant color (blue or red) on nonmagnified (nonzoomed) images.

‡ AMVL thickness should be measured during diastole at full excursion. Measurement should be taken at the thickest portion of the leaflet and should be performed on a frame with maximal separation of chordae from the leaflet tissue.

Appendix 4: Public Health Unit checklist (details will vary significantly according to jurisdiction). Ensure all criteria are marked

Contact the patient's doctor to:

Patient ID number: [Click or tap here to enter text.](#)

Yes	No	Not done	Public Health Unit checklist
Yes	No	Not done	Commenced ARF Case notification form and an Enhanced Surveillance form (Appendix 1 & 2)
Yes	No	Not done	Commenced jurisdictional notification form
Yes	No	Not done	Obtained necessary history from notes or interview of patient or clinician
Yes	No	Not done	Confirmed results of relevant pathology tests or recommend that the tests be done
Yes	No	Not done	Ensure the patient diagnostic workup is completed including ECG, CRP or ESR, serology and/or culture. * Follow-up serology may be required if negative or potentially mistimed
Yes	No	Not done	Recommended follow-up echocardiograms, specialist and dental reviews, in accordance with established care plans based on priority status
Yes	No	Not done	Commenced secondary prevention with regular benzathine penicillin G after ARF diagnosis
Yes	No	Not done	Ensured treating team is aware of local resources for patient and family education (including information on primordial and primary prevention strategies as well as the details of secondary prophylaxis provision locally)
Yes	No	Not done	Ensured the diagnosis is recorded clearly in the patients file
Yes	No	Not done	Provided feedback to clinicians if prior missed episodes or inadequate workup is identified

Contact the patient (or caregiver) to:

Yes	No	Not done	Public Health Unit checklist
Yes	No	Not done	Confirmed onset date and symptoms of the illness
Yes	No	Not done	Ensured patient is consented for inclusion on the jurisdictional register according to local eligibility criteria
Yes	No	Not done	Provided education for case and household/family about primordial and primary preventive strategies to reduce future GAS infections and the subsequent impacts
Yes	No	Not done	Ensured the patient is referred to the best available RHD Control program or service for continuity of education, appropriate pain relief and ability to recall for follow up
Yes	No	Not done	

Contact laboratory to:

Yes	No	Not done	Public Health Unit checklist
Yes	No	Not done	Obtain any outstanding results for supporting evidence of preceding Group A streptococcal infection as per Table 5 <ul style="list-style-type: none"> • antistreptolysin-O or anti-DNase B or other streptococcal antibody OR • Positive group A streptococcal (GAS) throat culture OR • Positive rapid antigen test for group A streptococci.

Contact local Housing Authority to:

Yes	No	Not done	
			Inform them of case details and environmental/housing information

Other issues:

Yes.No Not done	Used the RHD Australia Diagnosis Calculator App to assist early detection and diagnosis of ARF
Yes No Not done	Assessed information against case definition to confirm case
Yes No Not done	Entered case data onto notifiable diseases database (process varies in each jurisdiction)
Yes No Not done	Consider referral for a housing assessment according to local protocol
Yes No Not done	Maintained awareness of the possibility of ARF clusters/outbreaks and have a protocol in place to investigate and respond
Yes No Not done	Consider alerting local RHD program if there are more cases than usual
Yes No Not done	Consider active case finding where appropriate (refer Section 12)

Jurisdictional specific issues

ARF is not currently notifiable in ACT, Victoria or Tasmania. See Table 4.

Cross jurisdictional/border issues: Control programs or treating clinicians should ensure that for patients transferred from one jurisdiction to another, all relevant details, especially benzathine penicillin dosing dates, are provided. No formal transfer of care form is currently in use. This process is guided by the CDNA Cross-border NNDSS Notification Protocol.

There is no National register. For [information](#) on RHD programs and notification process in each jurisdiction visit: www.rhdaustralia.org.au.