7. Rheumatic heart disease control programs

Secondary prophylaxis of ARF in someone who is known to have had ARF is the only RHD control strategy shown to be effective and cost-effective at both individual and population levels. Effective RHD management involves regular clinical follow up, with specialist review and echocardiography.

This quick reference guide is derived from the *Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease* (2nd edn).

What is acute rheumatic fever?

Acute rheumatic fever (ARF) is an illness caused by a reaction to a bacterial infection with group A streptococcus. It causes an acute, generalised inflammatory response and an illness that targets specific parts of the body, including the heart, joints, brain and skin. Individuals with ARF are often unwell, have significant joint pain and require hospitalisation. Despite the dramatic nature of the acute episode, ARF typically leaves no lasting damage to the brain, joints or skin, but can cause persisting heart damage, termed 'rheumatic heart disease' (RHD). Recurrences of ARF may cause further cardiac valve damage. Hence, RHD steadily worsens in people who have multiple episodes of ARF.

What is RHD?

RHD is damage to the heart that remains after the acute ARF episode has resolved. It is caused by an episode or recurrent episodes of ARF, where the heart has become inflamed; the heart valves remain stretched and/or scarred, and normal blood flow is interrupted. Recurrences of ARF may cause further valve damage, leading to steady worsening of RHD. Preventing recurrences of ARF by using secondary prophylaxis treatment with penicillin is therefore of great importance.

Who gets RHD?

In Australia, the vast majority of people with RHD are Aboriginal people and Torres Strait Islanders, many of whom live in remote areas of central and northern Australia. Pacific Islanders, and migrants from high-prevalence countries, are also at high risk.

How RHD can be controlled

Secondary prophylaxis of ARF in someone who is known to have had ARF is the only RHD control strategy shown to be effective and cost-effective at both individual and population levels. The recommended method is a four-weekly benzathine penicillin G (BPG) injection.

The appropriate duration of secondary prophylaxis is determined by a number of factors, including age, time since the last episode of ARF and potential harm from recurrent ARF. For most individuals, the duration of secondary prophylaxis is at least 10 years.

Effective RHD management involves regular clinical follow up, with specialist review and echocardiography.

Problems with the control of RHD

While strategies for controlling RHD have been proven to be simple, cheap and cost-effective, they must be adequately implemented in the populations at highest risk of the disease.

Persistent high rates of recurrent ARF in high-risk populations highlight the continued barriers to secondary prevention.

Organised approaches are needed to increase the effectiveness of the secondary prevention of ARF and management of RHD. This should include strategies aimed at improving the delivery of secondary prophylaxis and patient care, the provision of education, coordinating available health services and advocacy for necessary and appropriate resources.

Organisational approaches to RHD control

A coordinated control program is the most effective approach in improving adherence to secondary prophylaxis of ARF and the clinical follow up of people with RHD.

Central to coordinated control programs at individual, community and national levels are registers of people with RHD or a history of ARF. Register-based programs improve case detection, increase adherence to secondary prophylaxis, reduce recurrences of ARF and decrease hospitalisations from ARF/RHD.

Registers also provide a mechanism for monitoring patient movements, orienting staff to ongoing care requirements, identifying individuals with poor adherence to long-term therapy and monitoring the success of programs and changes in disease epidemiology.

RHD control programs aim to:

- identify and register new cases of ARF and RHD

- improve the uptake of and adherence to secondary prophylaxis

- increase awareness of the diagnosis and management among healthcare providers

- improve clinical care and follow up in line with best practice

- support education and health promotion for individuals, families and the community

- promote primary prevention, aimed at preventing initial episodes of ARF

- use data to monitor patient outcomes and improve program strategies.

Recommended elements of RHD control programs

- Commitment from national, regional and local services, particularly to ensure long-term funding and governance support.

- An effective advisory committee that includes cardiologists, paediatricians, general practitioners, physicians, epidemiologists, nurses, public health practitioners, Aboriginal health service organisations and relevant community representatives.

- A dedicated coordinating team.

- An electronic patient register that contains data elements that support quality patient management, as well as any internal and external reporting requirements.

- A commitment to partnerships between clinicians and public health services in order to support the needs of people with ARF/RHD and the community.

- Prioritisation of primary and secondary antibiotic prophylaxis delivered within the framework of primary healthcare.

- Planning and advocacy for a stable supply of BPG, and established plans for established secondary prophylaxis in the event of supply reductions.

- A commitment to partnerships between clinicians and public health services in order to support the needs of people with ARF/RHD and the community.

- Education for health practitioners and health workers, and supported education for the community, those with the disease and their families.

- Activities guided by locally-relevant, evidence-based guidelines.

- Legislation and/or regulations warranting the notification of ARF/RHD, which is supported by public health surveillance activities at the state or territory level.

- A priority system that ensures that services are delivered to those at highest risk.

- A mechanism for monitoring the delivery of secondary prophylaxis and ongoing care.

- Evaluation of patient management and program activities.

Improving the uptake of and adherence to secondary prohylaxis in primary care

Evaluate the local health service environment to identify specific barriers to injection delivery. Based on the outcome of the evaluation, the following strategies may be useful:

- identify local, dedicated staff members responsible for the delivery of secondary prevention and the coordination of routine care

- focus on improving relationships between health staff and patients/families

- support and utilise the expertise, experience, community knowledge and language skills of Aboriginal health workers

- develop and implement recall and reminder systems (based on a local ARF/RHD register, where established) to accommodate the high mobility of individuals and groups

- ensure that recall systems extend beyond community boundaries

- establish networks for timely communication between health clinics

- use a centralised coordinator and register to assist in monitoring movement

- minimise staff turnover in remote and rural primary healthcare centres and regional hospitals, or minimise the impact of staff turnover, where possible

- promote the importance of secondary prophylaxis in preventing recurrent ARF, and the development or worsening of RHD

- improve the quality and delivery of ongoing health education and support for staff, patients and families

- implement measures to reduce pain of injections, where indicated

- base routine care on standardised evidence-based guidelines.

Surveillance

Passive surveillance of ARF usually depends on case identification from healthcare providers. Historically, this has underestimated the burden of disease, due to inaccuracies and incompleteness. In under resourced settings, problems with passive surveillance are exacerbated by the high turnover of staff, and lack of awareness of ARF and RHD.

Ideally, active surveillance should be used to augment passive surveillance. This entails establishing mechanisms to identify new cases of ARF and RHD, and to update information about existing cases. This could include mechanisms allowing access to hospital separation data, echocardiography reports, specialist review correspondence, primary healthcare clinic information and notifiable disease databases.

Where possible, these processes should be automated (e.g. with regular downloads of information regarding patients admitted to hospital with a diagnosis of ARF or RHD).

When active surveillance is established, an initial apparent increase in the prevalence of RHD is expected, primarily due to the detection and recording of existing cases, rather than the appearance of new cases. Similarly, improved access to specialist care may also result in greater rates of valvular surgery in the initial years after commencing a program.

When establishing surveillance systems for ARF/RHD control, a range of issues should be considered, including:

- defining the target population and high-risk groups requiring surveillance

- establishing a process for information flow from a range of potential data sources (e.g. case reporting, data collection instruments, data transmission and handling)

- formulating the essential data elements to be collected

- ethical and privacy legislation requirements, including consent

- data management (e.g. the most appropriate format for storing the data)

- proposed process and timeliness of data analysis

- dissemination and targets for the feedback of results

- needs of healthcare providers for individual patient and epidemiological information

- continuing refinement and evaluation of the surveillance system.

RHD registers

Register-based programs:

- improve case detection

- increase adherence to secondary prophylaxis

- reduce recurrences of ARF

- decrease hospitalisations from ARF/RHD.

Some programs have all-relevant patient data entered into a centralised register. Others choose to have a subset of data (e.g. recording of individual doses for secondary prophylaxis) entered only into the local register.

Where provision of secondary prophylaxis is not entered into a central register, local health staff should have clear guidelines on identifying and managing patients overdue for secondary prophylaxis.

Screening for RHD

RHD control programs should coordinate screening programs to detect previously undiagnosed RHD in high-risk populations, wherever possible. Ideally, screening should be undertaken at a time when the program has been established, and newly-identified cases are able to be managed within a supportive framework.

Although RHD prevalence is highest in adults, they are difficult to screen, and screening of school-aged children is therefore recommended (e.g. cardiac auscultation at school entry, and again at 10 years of age).

If time and other resources allow, consideration should be given to conducting more intensive screening programs, in which children of all ages are reviewed, and attempts are also made to examine children who miss school-based screening.

Legislated notification of ARF/RHD

ARF is a notifiable condition in Western Australia, the Northern Territory and Queensland.

RHD is not currently notifiable anywhere in Australia.

Indicators for evaluating ARF/RHD control programs

Control programs for ARF/RHD should be evaluated against criteria for routine care and key epidemiological objectives (see below).

Consideration should be given to assessing the delivery of specialist cardiology services, the availability and accessibility of echocardiography, referral practices and structures, transportation for patients and support and follow up processes.

Key Performance Indicators

1	Epidemiology		
	1.1	Yearly age-specific incidence rates of all episodes, and of first episodes of ARF according to sex (refer to 1.1.1) and ethnicity (refer to 1.1.2)	• 0-4 • 5-14 • 15-24 • 25-34 • 35-44 • >44yrs
		1.1.1 sex	• Male • Female • Indeterminate • Not stated/inadequately described
		1.1.2 ethnicity	• Aboriginal but not Torres Strait Islander origin • Torres Strait Islander, but not Aboriginal origin • both Aboriginal and Torres Strait Islander origins • Maori • other Pacific Islanders • other • unknown
	1.2	Proportion of all recorded ARF episodes classified as recurrences	
	1.3	Rates of ARF recurrences per 100 patient-years	
	1.4	Number of deaths and age-standardised rates of mortality due to ARF and RHD in the previous calendar year by ethnicity (refer to 1.1.2)	
	1.5	Yearly age-specific (refer to 1.1) and overall incidence of RHD by ethnicity (refer to 1.1.2) and broken down by method found and presented	• all recorded RHD cases • cases classified as mild • cases classified as moderate • cases classified as severe
	1.6	Yearly age-specific (refer to 1.1) prevalence of RHD, by ethnicity (refer to 1.1.2)	• all recorded RHD cases • cases classified as mild • cases classified as moderate • cases classified as severe
	1.7	Proportion of newly registered cases of ARF or RHD with an initial recorded diagnosis being established RHD (rather than ARF)	

2	**Requirement and uptake of secondary prophylaxis**	
	2.1	Proportion of all people indicated for secondary prophylaxis* who are registered to receive benzathine penicillin G (BPG)
	2.2	Median percentage of all scheduled BPG doses actually delivered
	2.3	Proportion of people indicated for BPG secondary prophylaxis who received <50%, 50-79%, and ≥80% of scheduled doses in the previous calendar year
3	**Quality of management**	
	3.1	Proportion of all registered ARF and RHD cases classified as mild, moderate, severe and inactive
	3.2	Proportion of people classified as moderate or severe RHD who had an echocardiogram within the previous 6 months, 1 year, and 1-2 years
	3.3	Number of cases, and proportion of total cases indicated for cardiac surgery, who have been waiting <6 months, 6-11 months, 12-23 months, or 24+ months
	3.4	Number and type of surgical procedures performed during the previous calendar year by the following:	
		3.4.1 age group	(refer to 1.1)
		3.4.2 ethnicity	(refer to 1.1.2)
	3.5	Number (and proportion) of people who died in the previous calendar year within 28 days and 1 year of undergoing rheumatic cardiac surgery by the following:	
		3.5.1 age group	(refer to 1.1)
		3.5.2 ethnicity	(refer to 1.1.2)

* If denominator of those indicated for prophylaxis not known, use people with a history of ARF within the last 10 years OR ARF and RHD and aged < 21 years OR aged ≥21 years with moderate or severe RHD

RHD Australia

The Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease (2nd edition)

Quick reference guides include:

- Primary prevention of ARF
- Diagnosis of ARF
- Management of ARF
- Secondary prevention of ARF
- Management of RHD
- RHD in pregnancy
- RHD control programs

RHDAustralia
Ph: 08 8922 8196
Email: info@rhdaustralia.org.au
www.rhdaustralia.org.au